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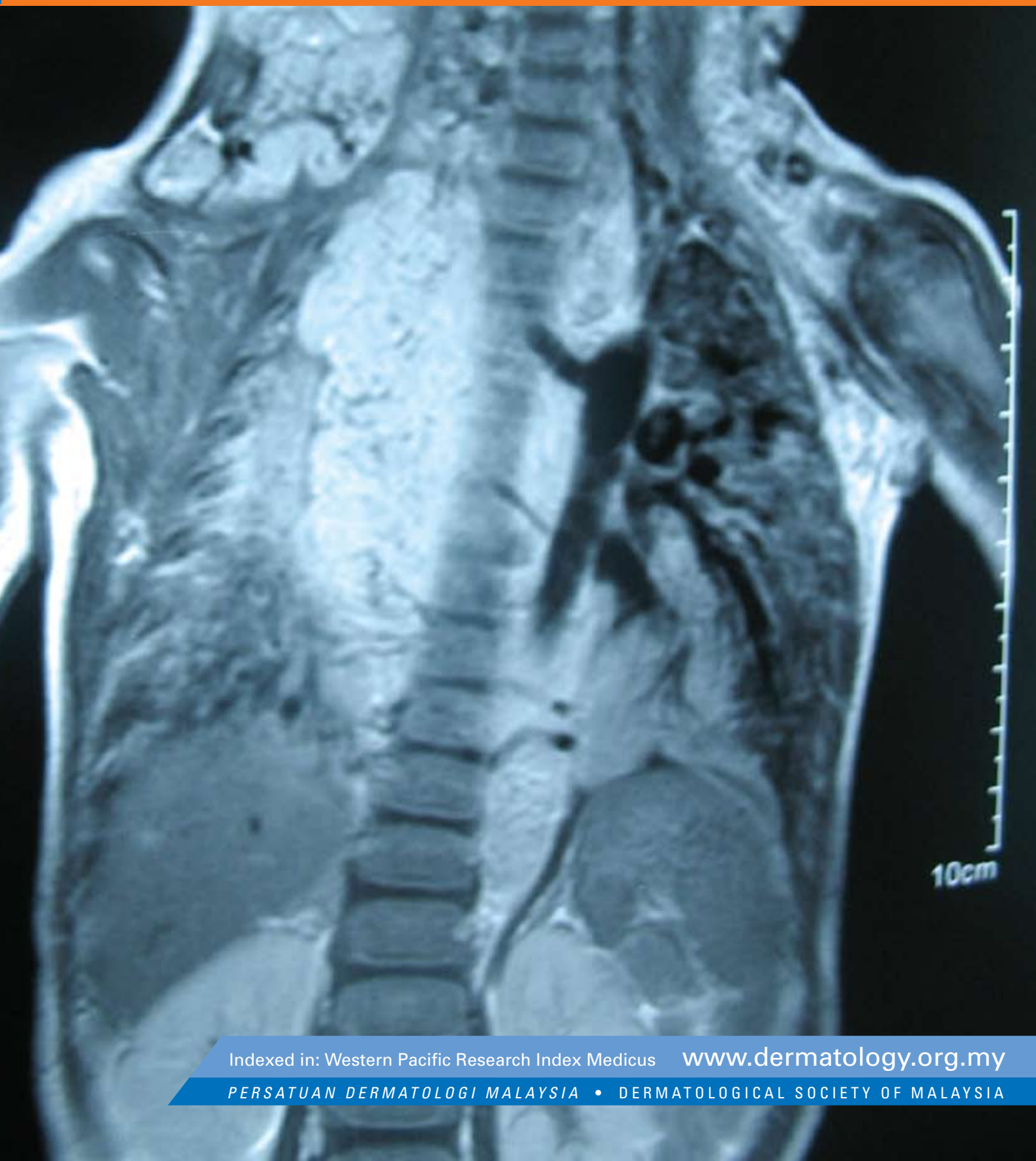

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editorial

Dermatology Practice in the 21st Century

Introduction of internet world wide has increased the patients' awareness of their own illness and this has led to an increase in their curiosity in the way doctors manage them. Providing immediate therapy is insufficient to the present patients who aspect their frontline dermatology care clinician to investigate and ascertain the cause of their illness especially those with eczema. In today's review article, we have a Polish dermatologist Dr. Radoslaw Spiewak discussing on epidemiology of eczema. We welcome you to share your views and feedback on the articles in this journal.

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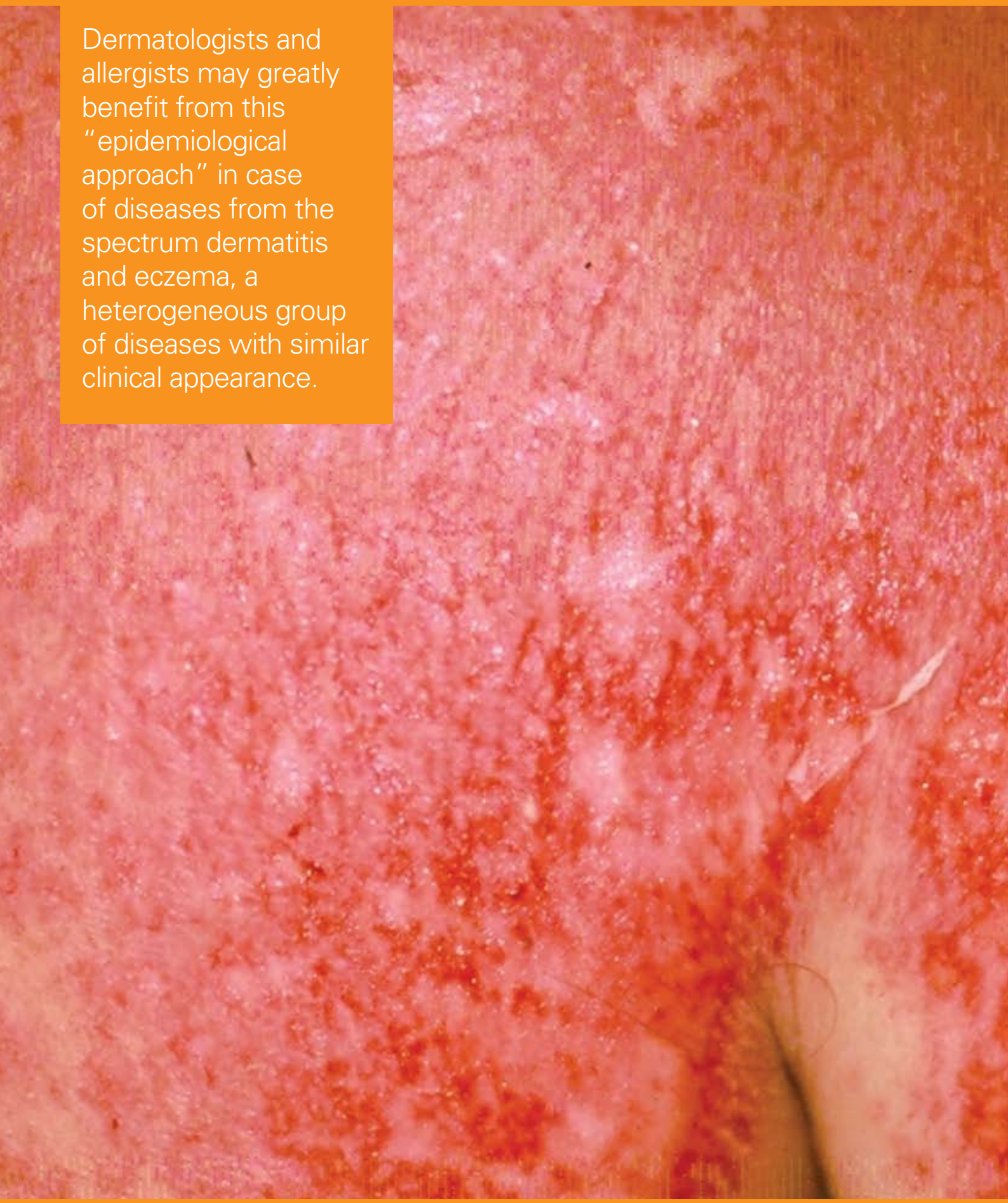
Dermatologists and allergists may greatly benefit from this "epidemiological approach" in case of diseases from the spectrum dermatitis and eczema, a heterogeneous group of diseases with similar clinical appearance.



Epidemiology of Skin Diseases from the Spectrum of Dermatitis and Eczema

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Abstract

Particular types of eczema may affect up to 29% individuals in certain populations (lifetime prevalence), thus placing the diseases among most frequent clinical problems. Nevertheless, diseases from the spectrum of dermatitis and eczema are poorly defined and frequently misdiagnosed; they also frequently overlap, making the diagnostic process even more difficult. In doubtful cases, where no further means of clinical or laboratory differentiation are available, reliable epidemiological data may provide relevant help in the diagnostic process, as the best candidate for a tentative diagnosis seems the most frequent among diseases in question, which can be verified later by the effectiveness of respective treatment regimen. However, results of epidemiological studies in the field of eczema and dermatitis may be strikingly contradictory, one of the possible reasons being definitions of various types of eczema/dermatitis that leave too much space for individual decision and thus seem hardly suitable for epidemiological research. Better studies based on unequivocal definitions of various types of eczema are necessary to achieve the quality of epidemiological data that would ensure the level of certainty expected from a diagnostic tool. The present paper collates results from available epidemiological data on various types of eczema: atopic eczema, allergic and irritant contact dermatitis, protein contact dermatitis, seborrhoeic dermatitis, asteatotic dermatitis, stasis dermatitis, nummular eczema, dyshidrotic eczema (pompholyx), hand dermatitis and occupational dermatitis. Problems and possible sources of bias in available studies are addressed and discussed along with the results from the studies.

Keywords: *epidemiology; atopic eczema; allergic and irritant contact dermatitis; protein contact dermatitis; seborrhoeic dermatitis; asteatotic dermatitis; stasis dermatitis; nummular eczema; dyshidrotic eczema; pompholyx; hand dermatitis; occupational dermatitis*

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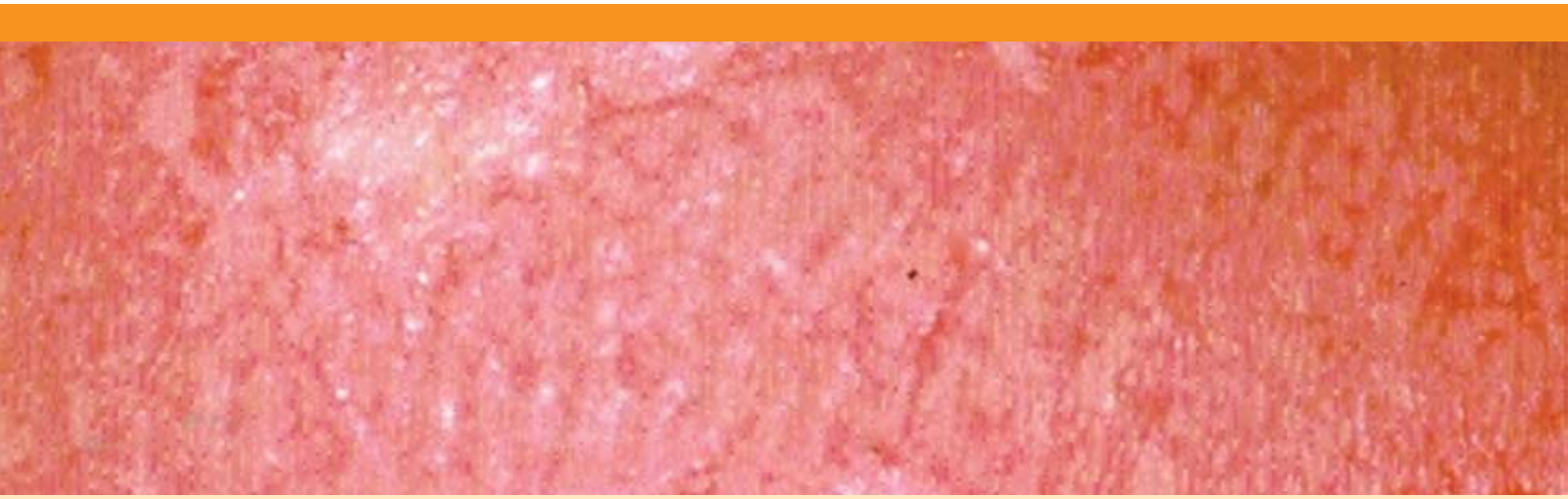

Introduction

The knowledge of the frequency of diseases is important for policy makers, insurers, but it is also a very important diagnostic tool in the hand of a clinician. In a considerable group of patients the clinical picture does not allow for a clear-cut diagnosis, and after exhausting all available possibilities of differential diagnosis the clinician is stuck with two or more possible diagnoses.

In such cases, the knowledge of epidemiology may be resorted to as the ultimate instance of clinical decision. Knowing the prevalence rates of otherwise equally possible diseases that come in question, it seems rational to pick the more prevalent disease as the tentative diagnosis, with a possible revision if appropriate treatment turns out ineffective.

Eczema

(synonym: dermatitis) is noncontagious inflammation of the epidermis and dermis with characteristic clinical features (itch, erythema, papule, seropapule, vesicle, scale, squame, crust or lichenification that emerge simultaneously or evolve from one another) and distinct histological picture (spongiosis, acanthosis, parakeratosis, lymphocytic and granulocytic infiltrates)[1, 2].

The debate on the differences between the terms "eczema" and "dermatitis" has been ongoing for many decades, with no definite conclusion[3, 4]. Therefore, in the present article these terms will be considered as synonyms. The clinical spectrum of dermatitis/eczema diseases includes an array of diseases that sometimes are depicted as mutual opposites, however, their clinical features and pathomechanisms overlap to an extent making any clear-cut differentiation virtually impossible.

In epidemiological studies of the various dermatitides, most striking is the difficulty of drawing general conclusions, mainly due to imprecise definitions and incompatible outcome measures. This must be born in mind when looking at the epidemiological data discussed below.

In the analysis of diseases frequency, it is crucial to remember that different methods of collecting epidemiological data may give different outcomes. The most popular method to obtain epidemiological data on diseases is self-administered questionnaire. This method has some advantages, which are very important when conducting an epidemiological research: it is inexpensive and easy to use, so it can be applied in large populations.

Disadvantages of the questionnaire-based method are also very significant, especially the possibility of misunderstanding the questions which may lead to the probability of overestimation of the obtained results[5].

Another method for assessing the frequency of diseases is medical examination. This method seems more objective and thus more reliable because it allows for verification of symptoms by a specialist. In comparison to the questionnaire-based method, medical examination requires much more costs and time for performing[6]. Moreover, when comparing these two methods of collecting epidemiological data, it is important to remember that questionnaire-based method is more suitable for collecting information about prevalence of diseases over a period of time (e.g., lifetime prevalence or one-year prevalence), while medical examination is more appropriate for assessing the presence of the disease at a particular point in time (point prevalence)[7]. Thus these methods should be regarded as complementary.

Some estimates about the frequency of diseases come from various registers, such as hospital records, national or local statistics (e.g. occupational diseases statistics). This "ecological" (i.e. not consuming new resources) method has its advantages, for example it allows for comparison of trends at different time points. However, discrepancies may arise due to different classifications of diseases used in various data collecting systems, or in various periods of time.

A major possible disadvantage of using the "epidemiological approach" in clinical diagnosis is that of a "self-fulfilling prophecy": With poor-quality epidemiological data at the start, one may classify unclear cases of eczema for this type that is believed to be most frequent, which may not necessarily be the truth, however, by doing so the statistics are biased toward the tentative diagnosis, thus reinforcing one's beliefs into seemingly "scientific proof". It seems that this is especially true for the diseases from the spectrum of dermatitis and eczema.

Therefore, it is extremely important to be critical when looking at the frequency rates of diseases from the spectrum of dermatitis and eczema. The differences in definitions of the diseases in various studies or sometimes lack of any definitions, strongly supports this attitude. In this article, in order to be able to collate available epidemiological data, we have adopted a simplistic attitude that the diagnosis of a given disease is defined by the authors' declaration (i.e. belief) that they studied this particular disease. The following data, therefore, give us some idea about possible prevalence rates, however, due caution is recommended while using them for "epidemiological" diagnosis.

Atopic eczema

(AE, synonym: atopic dermatitis) is a chronic inflammatory skin disease that commonly begins in early infancy, runs a course of exacerbations and remissions, and is associated with a characteristic distribution and morphology of skin lesions. Furthermore, pruritus and subsequent sleeplessness are hallmarks of this disease[8]. This "minimalist" definition seems most acceptable for the time being, as it puts forward the common clinical characteristics while avoiding references to pathomechanisms, which are still subject to controversy (see below).

Prevalence of atopic dermatitis/eczema in children has been widely assessed. The most known epidemiological study on atopic eczema (AE) in children is the ISAAC Study[9]. This questionnaire-based study allows estimating one-year and lifetime prevalence rates of AE among children. Table 1 presents prevalence rates of atopic eczema according to studies based on the ISAAC questionnaire. Both indices of the disease frequency (one-year prevalence, and lifetime prevalence) showed great variability in the estimations among countries ranging from 4.5% to 20.2% (1-year prevalence) and from 2.4% to 28.7% (lifetime prevalence) [10-12].

However, there has been a heated discussion on how reliable is the ISAAC questionnaire in detecting AE[13,14], with recent data showing that up to 50% of children with 'ISAAC eczema' may in fact be ill with allergic contact dermatitis (ACD)[15]. Flexural eczema - almost a "diagnostic fetish" in past epidemiological studies of AE has turned out less specific to AE than previously believed[16], not least so because this clinical feature is also common in ACD[17-21], and cases of ACD-related flexural eczema have been misdiagnosed as AE for decades[22, 23]. With this respect, ISAAC studies may be looked at as an example of the "self-fulfilling prophecy" in the epidemiology of eczema in children. In order to overcome these limitations, other methods were also used when assessing the frequency of AE.

Detailed information on the prevalence of AE in children according to studies not based on the ISAAC questionnaire is shown in Table 2. Less is known on the prevalence of AE in adults - available data are collated in Table 3. The major problem with the epidemiological data of AE is that "atopic eczema" seems in fact to be a heterogeneous group of diseases with similar clinical appearance, rather than a single disease.

The spectrum of involved pathologies range from type I and IV allergy (possibly also types II and III), to barrier dysfunction, abnormalities of the innate immune response and autoimmunity, while it remains unclear, which of those are actual causes and which secondary phenomena[24-27]. For example, the causal role of IgE-mediated food allergy in AE seems overrated[28, 29] and the development of food-specific IgE may, in fact, be secondary to eczema[30].

The name "atopic dermatitis" itself was already criticized by Rajka in 1975 as an "unfortunate choice of term"[31], which is supported by the fact that a majority of AE patients show no evidence of atopy[32]. Perhaps "Hanifin-Rajka Syndrome" would be a more appropriate name for this entity, avoiding the reference to questionable aetiology and focusing instead on the common clinical picture first compiled by the authors.

Table 1 Prevalence rates of atopic dermatitis in children according to studies based on the ISAAC questionnaire.

Country	Age of children	Gender of children	One-year prevalence	Lifetime prevalence
Austria[33*]	6-9 (1995-97)	Boys	5.0%	8.2%
		Girls	7.0%	10.2%
	6-9 (2001-03)	Boys	5.9%	10.2%
		Girls	7.6%	11.8%
Brazil[34]	13-14	Boys and girls	-	16.2%
China[35]	6-13	Boys and girls	5.5%	-
China[36^]	0-14	Boys and girls	-	14.5%
Croatia[37]	12-14	Boys and girls	5.3%	7.0%
Germany[38]	6-7	Boys	7.3%	14.3%
		Girls	6.7%	14.6%
	13-14	Boys	5.0%	8.2%
		Girls	9.4%	12.3%
	6-7	Boys	6.6%	13.6%
		Girls	9.8%	16.9%
	13-14	Boys	4.5%	10.9%
		Girls	11.1%	17.4%
Ghana[39^]	4-16	Boys	-	4.0%
		Girls		
Iran[40]	13-14	Boys and girls	10.1%	-
Italy[41]	2	Boys	16.8%	-
		Girls	18.7%	-
	3	Boys	16.2%	-
		Girls	20.2%	-
	4	Boys	19.1%	-
		Girls	17.2%	-
Korea[42]	8-11	Boys	12.7%	26.8%
		Girls	14.5%	28.7%
Malta[43]	13-15 (1995)	Boys and girls	12.8%	11.2%
	13-15 (2000)	Boys and girls	10.1%	8.5%
Mexico[44**]	6-8	Boys and girls	10.1%	15.0%
	11-14		10.5%	17.0%
	6-8	Boys and girls	5.8%	7.3%
	11-14		5.4%	7.0%
Montenegro[45]	6-7	Boys and girls	9.5%	-
	13-14		9.1%	-
Poland[46]	7	Boys and girls	-	9.4%
	16		-	3.4%
Serbia[45]	6-7	Boys and girls	11.2-17.2%	-
	13-14		8.2-16.2%	-
Spain[47]	6-7	Boys and girls	-	5.9%
Spain[48]	10-11	Boys and girls	-	11.4%
	1-2		15.0%	16.2%
Sweden[49]	2-3	Boys and girls	20.2%	23.7%
	3-4		20.7%	25.8%
	6-7	Boys and girls	17.8%	22.5%
	7-8		16.6%	21.2%
	8-9		20.7%	26.1%
United Kingdom[50]	6-7	Boys	-	27.8%
		Girls	-	27.0%

* *prevalence rates were estimated using questions about presence of an itchy rash in the past 12 months and lifetime symptoms of an itchy rash*
** *prevalence rates were estimated using questions about presence of dry itchy skin spots in the last 12 months and at any time*
^ *calculated based on the figures provided by the authors*

Table 2 Prevalence of atopic eczema in children in various studies based on different methods.

Country	Age of children	Method of assessment	Results
Denmark[51]	12-16	Q ME	*lifetime prevalence:* 21.3% (17.0% boys; 25.7% girls) *one-year prevalence:* 6.7% (5.6% boys; 7.7% girls) *point prevalence:* 3.6% (3.8% boys; 3.4% girls)
Denmark[52]	7	Q	*lifetime prevalence:* 22.9%
Gabon[39]	4-16	ME	*point prevalence:* 4.0%
Germany[53]	5-7	ME	*point prevalence:* 12.9%
Germany[54]	0-4	Q	*lifetime prevalence:* 21.4%
Germany[52]	7	Q	*lifetime prevalence:* 13.1%
Germany[55]	< 10	Q	*lifetime prevalence:* 13.0% (Leipzig), 13.9% (Munich)
Ghana[39]^	4-16	ME	*point prevalence:* 1.6%
Rwanda[39]	4-16	ME	*point prevalence::* 0.8%
Spain[48]	10-11	ME	*point prevalence:* 1.9%
Sweden[52]	7	Q	*lifetime prevalence:* 15.5%
Turkey[56]	0-16	HR	*lifetime prevalence:* 11.8%
United Kingdom[57]	1-5	Q	*one-year prevalence:* 16.5% (22% in 1-2 y.o.; 19% in 2-3 y.o.; 13% in 3-4 y.o.; 15% in 4-5 y.o.)
United States[58]	5-9	Q	17.2% (standard scoring criteria) 6.8% (highly stringent criteria)

Q - questionnaire; ME - medical examination; HR - hospital record *^ own calculations based on the figures provided by the authors*

Contact dermatitis

(Synonym: contact eczema) is a collective term for three dermatitides with various aetiologies, whose common feature is the development of skin inflammation in response to a direct contact with the provoking agent: 1) irritant contact dermatitis, 2) allergic contact dermatitis and 3) protein contact dermatitis[66].

Allergic contact dermatitis

(Synonym: allergic contact eczema) is inflammatory skin disease initiated by specific immune reaction to a hapten. It occurs in individuals with previously acquired contact allergy following re-exposure to the sensitizing hapten[67]. In contrast to ICD, only a minority of people exposed to a particular hapten will respond with dermatitis. When looking at epidemiological data, one must remember that ACD is not the same as contact allergy (CA).

Table 3 Prevalence rates of atopic eczema in adults.

Country	Age of children	Method of assessment	Results
Australia[59]	20+	ME	*point prevalence:* 5.7% in men, 8.1% in women
Denmark[60]	18-69	Q	*lifetime prevalence:* 10.0%
Germany[61]	0-99	Q ME	*lifetime prevalence:* 23.5% *point prevalence:* 16.0%
Japan[62]	20+	ME	*point prevalence:* 6.9% (participants in their 20s: 9.8%; 30s: 8.7%; 40s: 4.4%; 50/60s: 2.6%)
Norway[63]	18-69	Q	*lifetime prevalence:* 13.8% in men, 19.0% in women
Norway[64]	born in 1970, 1960, 1955, 1940-1941 and 1924-1925	Q	*lifetime prevalence:* 8.8%
Poland[65]	18-19	Q ME	*lifetime prevalence:* 5.0% *one-year prevalence:* 3.9% *point prevalence:* 2.5%
Russia[63]	18-69	Q	*lifetime prevalence:* 10.4% in men, 12.0% in women

Q - questionnaire; ME - medical examination

The term "contact allergy" refers to a state of altered response of the immune system to a specific substance, which is not synonymous with disease. Certain proportion of people with CA will never develop clinical symptoms. Among those symptomatic, vast majority will develop ACD, which is an inflammatory disease of the skin provoked by a hapten (a low molecular sensitizer), following the exposure to this hapten of a sensitized person[68]. Confusing contact allergy with allergic contact dermatitis seems a frequent mistake of doctors and authors of clinical and epidemiological studies.

Children

A very comprehensive method of establishing the prevalence of ACD in children was used in the study conducted in Denmark. ACD in the group of 12-16 years old children was defined by the co-existence of the three criteria: 1) contact allergy diagnosed by a positive patch test 2) exposure history and 3) history or present dermatitis pattern. Lifetime prevalence of ACD was 7.2%, and point prevalence 0.7% (calculated on the basis of data provided in the article)[51]. A Polish study showed that among 7-year old children the lifetime prevalence of symptoms of ACD was slightly higher than among 16-year olds (7.2% versus 6.1%)[46]. This is also reflected in higher contact hypersensitivity rates among children (67.0%) than adolescents (58.1%) seen in a similar cohort of Polish children[69], which may be explained by changing exposure patterns in the rapidly westernising country[70].

Adults

In the United States, in a study of university students, ACD was the cause of 3.1% of first-time visits to dermatologists, and 2.4% of total visits to dermatologists[71]. In Poland, prevalence of ACD was assessed among students of vocational agricultural schools. History and symptoms-based physician diagnosis estimated the frequency of ACD as: 2.0% (point prevalence), 9.3% (one-year prevalence), and 17.5% (lifetime prevalence)[65].

Irritant contact dermatitis

(ICD) is acquired inflammatory skin disease caused by chemical or physical insults leading to direct cellular injury. Most of ICD cases are associated with detergents, solvents, acids or alkali. Acute ICD (toxic dermatitis) develops rapidly (minutes to hours) after exposure to potent irritants, while chronic, cumulative variants of ICD develop gradually in response to repeated contacts with milder irritants[72]. ICD is essentially an injury, therefore, everyone will develop this disease after an individual threshold of resistance to irritants is exceeded[73].

The prevalence of irritant contact dermatitis (ICD) in general population is hard to determine, especially among children. Study conducted on a group of university students in the United States, showed that ICD was the cause of 2.3% of first-time visits to dermatologists, and 1.6% of total visits to dermatologists[71]. In Poland, estimations from the study conducted among students of a vocational school were: 0.5% (point prevalence), 4.3% (one-year prevalence), and 12.7% (lifetime prevalence)[65].

Protein contact dermatitis

(PCD) is acquired inflammatory skin disease initiated by specific immune reactions to allergens - proteins with molecular weight exceeding 10000 Daltons, usually of animal or plant origin[74, 75]. There is lack of data on the frequency of protein contact dermatitis among children. Estimates for adults are available only for work-related settings. In Finland, protein contact dermatitis (together with contact urticaria) accounted for 11.1% of all allergic occupational diseases reported in 1991[76]. Protein contact dermatitis was found in 22% of a group of 144 slaughterhouse workers in Denmark[77].

Seborrhoeic dermatitis

Seborrhoeic dermatitis is an inflammatory skin disease of the dermatitis/eczema spectrum, with a characteristic restriction to "seborrhoeic areas", i.e. areas with a high density of sebaceous glands (face, sternum, interscapular area). The aetiology remains unclear, one possibility being the excessive development of lipophilic Malassezia yeasts on the seborrheic skin with secondary development of inflammation in response to signalling molecules such as malassezin[78].

Little is known on the prevalence of seborrhoeic dermatitis. In a Turkish study of paediatric patients (0-16 years old) in a hospital registry, 4.3% children were diagnosed with seborrhoeic dermatitis[56]. The prevalence of seborrhoeic dermatitis in adults was established in an Australian study based on medical examination, was 12.3% in men, and 7.3% in women[59]. Among university students in the USA, seborrhoeic dermatitis was the cause of 3.1% of first-time visits, and of 2.4% of all dermatologist consultations[71].

In a prospective, skin examination-based study of renal transplant recipients in the UK, seborrhoeic dermatitis was found in 9.5% of the participants[79]. The prevalence of seborrhoeic dermatitis of the face and scalp diagnosed among mountain guides was 16.3%, which might hint on a role of UV irradiation in these cases[80].

Asteatotic dermatitis

Asteatotic dermatitis (dry skin dermatitis, winter itch) is an entity of unknown aetiology, characterised by the presence of dry, scaly, fissuring skin accompanied with pruritus, typically localised on the calves, with a possibility of spreading. Among exacerbating/causative factors, skin ageing with atrophy and xerosis, low humidity of ambient air, as well as frequent bathing and excessive detergent use are mentioned. Among Australian adults the prevalence of doctor-diagnosed asteatotic dermatitis was 6.6% in men, and 10.4% in women[59].

Stasis dermatitis

Stasis dermatitis is a skin manifestation of venous insufficiency and frequently is accompanied by other symptoms like the presence of varicous veins, leg oedema and ulcers, hemosiderin deposits in the skin and liposclerosis of the skin. The typical localization is calves. In the above-mentioned Australian study, the frequency of stasis dermatitis was assessed at 2.1% in men, and 1.5% in women[59].

Nummular eczema

Nummular eczema (nummular dermatitis, discoid dermatitis) is characterized by solitary or multiple, well-demarcated, round or oval-shaped itchy lesions. The typical course of the disease is chronic recurrent. The identity of this disease is built based upon the characteristic clinical appearance; however, the aetiology remains unknown. One of the more popular hypotheses considers immunological response (allergic reaction type II or IV) to circulating antigens of bacteria, fungi or parasites. On the other hand, it seems that may various types of eczema may take this clinical appearance, e.g. atopic eczema, allergic contact dermatitis (to nickel, neomycin, etc.), along with asteatotic and stasis dermatitis. In a Turkish study utilizing data of hospitalised paediatric patients, 0.4% children (0-16 years old) were diagnosed with nummular dermatitis[56].

Dyshidrotic eczema

Dyshidrotic eczema (pompholyx) is a non-infectious inflammation of the skin characterized by the appearance of pruritic vesicles on the palms and soles. The course of

Table 4 Prevalence rates of hand dermatitis/eczema.

CHILDREN

Country	Age of children	Method of assessment	Results
Denmark[51]	12-16	Q ME	*lifetime prevalence:* 9.2% (6.3% boys, 12.2% girls) *one-year prevalence:* 7.3% (4.6% boys, 10.1% girls) *point prevalence:* 3.2% (2.2% boys, 4.2% girls)
Norway[84]	7-12	Q ME	*one-year prevalence:* 6.5% *point prevalence:* 3.5%

ADULTS

Country	Age of children	Method of assessment	Results
Denmark[60]*	18-69	Q	*lifetime prevalence:* 21.8% (17.0% men, 25.7% women) *one-year prevalence:* 11.7% (8.9% men, 14.0% women)
Norway[64]	born in 1970, 1960, 1955, 1940-1941 and 1924-1925	Q	*lifetime prevalence:* 8.2%
Poland[85]	20-73	Q	*lifetime prevalence:* 17.3% *one-year prevalence:* 10.1% *point prevalence:* 1.9%
Sweden[86]	20-65	Q ME	*one-year prevalence:* 11.0% *point prevalence:* 5.4%
Sweden[87]	20-65	Q	*one-year prevalence:* 11.8% (1983) and 9.7% (1996)
Sweden[88]*	20-77	Q	*lifetime prevalence:* 11.0% (6.8% men, 14.0% women) *one-year prevalence:* 6.5% (4.5% men, 8.1% women)

Q - questionnaire; ME - medical examination ** calculated based on the figures provided by the authors*

the disease may be acute, recurrent, or chronic. The skin lesions frequently are restricted to areas with high density of sweat glands and frequently accompanied by hyperhidrosis[81]. However, it appears that the lesions are not connected with the glands.

In the above-mentioned Turkish study, dyshidrotic eczema was diagnosed in 1.0% of paediatric hospital patients (0-16 years old)[56]. In an epidemiological study of adult Dutch metalworkers, symptoms of dyshidrotic eczema were found in 7.3% of a group of metalworkers[82].

Hand dermatitis

Hand dermatitis is a very special nosological entity that refers to the clinical picture (dermatitis localized on the hands) rather, than to the cause. Hand dermatitis/eczema may be a manifestation of ACD, ICD, atopic dermatitis, or other inflammatory diseases, which in this location are very difficult to differentiate based on the clinical picture or medical tests (including histopathology). A co-existence of more than one causes of hand dermatitis (e.g. ACD + ICD + atopic hand dermatitis) is relatively common, hence it seems practical to view hand dermatitis as a distinct clinical entity[83]. Prevalence rates of hand dermatitis/eczema in children and adults are shown in Table 4.

Occupational dermatitis

Occupational contact dermatitis is neither clinical nor pathological entity; however, due to specific circumstances of appearance and special legal status in many countries, cases of such diseases are closely followed. OCD occurs mostly on hands (80% cases) and face (10% cases)[89]. The frequency of occupational contact dermatitis (OCD) in the United Kingdom is estimated as 12.9 cases per 10 thousand full-time workers each year[90]. One-year prevalence of occupational hand dermatitis, depending on the method of estimation, varies from 0.5-6.7% (medical examination) to 8.2-10.6% (questionnaire) in different populations[91].

In a study based on medical examination, 4.1% Polish farmers were diagnosed with occupational hand eczema[92]. One in three of those who stated to have hand dermatitis ever, and one in five with wrist and forearm dermatitis reported on exacerbations of dermatitis due to substances present at workplace[85]. Irritant contact dermatitis (ICD) and allergic contact dermatitis (ACD) contribute to most cases of OCD. Different proportions of ICD and ACD are reported in studied populations - frequency of ICD varies from 32% (USA) to 71% (Australia)[89]. The differences might reflect the diagnostic routines (most importantly the use and extensiveness of patch tests).

Final remarks

The major disadvantage of available epidemiological studies of diseases from eczema and dermatitis spectrum is that they depend on clinical symptoms, which are frequently difficult to properly classify even by an experienced clinician, as clinical features and pathomechanisms of various types of eczema overlap to an extent making clear-cut differentiations virtually impossible. Studies based on self-administered questionnaires, are even more susceptible to bias as conclusions are built based upon patient's own opinions and interpretations. Furthermore, various types of eczema may co-exist, while most researchers and doctors rest satisfied with a first diagnosis established. To acquire reliable data on the epidemiology of various types of dermatitides, better studies are needed in the future based on well-defined criteria that would enable accurate differentiation between analysed diseases. Specific requirements for such studies have been recently discussed elsewhere[93].

Reference

1. *Weidinger S, Ring J. Diagnosis of atopic eczema. In: Ring J, Przybilla B, Ruzicka T, editors. Handbook of Atopic Eczema. Second Edition. Berlin: Springer; 2006:84-99.*
2. *Darsow U, Wollenberg A, Simon D, et al. ETFAD/EADV eczema task force 2009 position paper on diagnosis and treatment of atopic dermatitis. J Eur Acad Dermatol Venereol 2009;24:317-328.*
3. *Ackerman AB, Ragaz A. A plea to expunge the word "eczema" from the lexicon of dermatology and dermatopathology. Arch Dermatol Res 1982;272:407-20.*
4. *Altekrueger I, Ackerman AB. "Eczema" revisited. A status report based upon current textbooks of dermatology. Am J Dermatopathol 1994;16:517-22.*
5. *Dotterud LK, Falk ES. Evaluation of a self-administered questionnaire on atopic diseases. Discrepancy between self-reported symptoms and objective signs. Eur J Public Health 2000;10:105-107.*
6. *Perreault N, Brisson C, Dionne CE, et al. Agreement between a self-administered questionnaire on musculoskeletal disorders of the neck-shoulder region and a physical examination. BMC Musculoskelet Disord 2008;17;9:34.*
7. *Berg M. Evaluation of a questionnaire used in dermatological epidemiology. Discrepancy between self-reported symptoms and objective signs. Acta Derm Venereol Suppl (Stockh) 1991;156:13-7.*
8. *Werfel T, Breuer K. Role of food allergy in atopic dermatitis. Curr Opin Allergy Clin Immunol 2004;4:379-385.*
9. *The ISAAC Study [electronic resource, URL: http://isaac.auckland.ac.nz/about/about.php] (accessed on 5 June 2012).*
10. *Odhiambo JA, Williams HC, Clayton TO, et al.; ISAAC Phase Three Study Group. Global variations in prevalence of eczema symptoms in children from ISAAC Phase Three. J Allergy Clin Immunol 2009;124:1251-8.e23.*

11. *Sole D, Mallol J, Wandalsen GF, Aguirre V; Latin American ISAAC Phase 3 Study Group. Prevalence of symptoms of eczema in Latin America: results of the International Study of Asthma and Allergies in Childhood (ISAAC) Phase 3. J Investig Allergol Clin Immunol 2010;20:311-23.*
12. *Flohr C. Recent perspectives on the global epidemiology of childhood eczema. Allergol Immunopathol (Madr) 2011;39:174-82.*
13. *Flohr C, Williams HC. Childhood eczema according to the International Study of Asthma and Allergies in Childhood (ISAAC) questionnaire tool - response to Czarnobilska et al. J Eur Acad Dermatol Venereol 2011;25:993-4.*
14. *Spiewak R, Czarnobilska E. Not all that looks like eczema is atopic eczema. J Eur Acad Dermatol Venereol 2011;25:992-3.*
15. *Czarnobilska E, Obtulowicz K, Dyga W, Spiewak R. A half of schoolchildren with 'ISAAC eczema' are ill with allergic contact dermatitis. J Eur Acad Dermatol Venereol 2011;25:1104-7.*
16. *Flohr C, Weiland SK, Weinmayr G, et al. The role of atopic sensitization in flexural eczema: findings from the International Study of Asthma and Allergies in Childhood Phase Two. J Allergy Clin Immunol 2008;121:141-147.*
17. *Dou X, Liu LL, Zhu XJ. Nickel-elicited systemic contact dermatitis. Contact Dermatitis 2003;48:126-129.*
18. *Oiso N, Ota T, Yoshinaga E, et al. Allergic contact dermatitis mimicking atopic dermatitis due to enoxolone in a topical medicament. Contact Dermatitis 2006;54:351.*
19. *Matiz C, Jacob SE. Systemic contact dermatitis in children: how an avoidance diet can make a difference. Pediatr Dermatol 2011;28:368-374.*
20. *innicki M, Shear NH. A systematic approach to systemic contact dermatitis and symmetric drug-related intertriginous and flexural exanthema (SDRIFE): a closer look at these conditions and an approach to intertriginous eruptions. Am J Clin Dermatol 2011;12:171-180.*
21. *Tan SC, Tan JW. Symmetrical drug-related intertriginous and flexural exanthema. Curr Opin Allergy Clin Immunol 2011;11:313-318.*
22. *Shanon J. Pseudo-atopic dermatitis. Contact dermatitis due to chrome sensitivity simulating atopic dermatitis. Dermatologica 1965;131:176-190.*
23. *Herro EM, Jacob SE. Systemic contact dermatitis - kids and ketchup. Pediatr Dermatol 2012 (Epub ahead of print, DOI: 10.1111/j.1525-1470.2011.01702.x).*
24. *Akdis M. The cellular orchestra in skin allergy; are differences to lung and nose relevant? Curr Opin Allergy Clin Immunol 2010;10:443-451.*
25. *Mittermann I, Aichberger KJ, Bunder R, et al. Autoimmunity and atopic dermatitis. Curr Opin Allergy Clin Immunol 2004;4:367-371.*
26. *Niebuhr M, Werfel T. Innate immunity, allergy and atopic dermatitis. Curr Opin Allergy Clin Immunol 2010;10:463-468.*
27. *Heratizadeh A, Wichmann K, Werfel T. Food allergy and atopic dermatitis: how are they connected? Curr Allergy Asthma Rep 2011;11:284-291.*
28. *Worth A, Sheikh A. Food allergy and atopic eczema. Curr Opin Allergy Clin Immunol 2010;10:226-230.*
29. *Spiewak R. Food-provoked eczema: A hypothesis on the possible role of systemic contact allergy to haptens present in both cosmetics and foods. Estetol Med Kosmetol 2011;1:35-40.*
30. *Hanifin JM. Atopic dermatitis nomenclature variants can impede harmonization. J Invest Dermatol 2012;132:472-473.*
31. *Rajka G. Atopic dermatitis. London: W. B. Saunders; 1975: 2.*
32. *Flohr C, Johansson SG, Wahlgren CF, Williams H. How atopic is atopic dermatitis? J Allergy Clin Immunol 2004;114:150-158.*
33. *Weber AS, Haidinger G. The prevalence of atopic dermatitis in children is influenced by their parents' education: results of two cross-sectional studies conducted in Upper Austria. Pediatr Allergy Immunol 2010;21:1028-35.*
34. *Toledo MF, Rozov T, Leone C. Prevalence of asthma and allergies in 13- to 14-year-old adolescents and the frequency of risk factors in carriers of current asthma in Taubaté, Sao Paulo, Brazil. Allergol Immunopathol (Madr) 2011;39:284-90.*
35. *Li F, Zhou Y, Li S, et al. Prevalence and risk factors of childhood allergic diseases in eight metropolitan cities in China: a multicenter study. BMC Public Health 2011;11(1):437.*
36. *Zhao J, Bai J, Shen K, et al. Self-reported prevalence of childhood allergic diseases in three cities of China: a multicenter study. BMC Public Health 2010;13;10:551.*
37. *Munivrana H, Vorko-Jovic A, Munivrana S, et al. The prevalence of allergic diseases among Croatian school children according to the ISAAC Phase One questionnaire. Med Sci Monit 2007;13:CR505-509.*
38. *Maziak W, Behrens T, Brasky TM, et al. Are asthma and allergies in children and adolescents increasing? Results from ISAAC phase I and phase III surveys in Munster, Germany. Allergy 2003:58:572-579.*
39. *Hogewoning AA, Bouwes Bavinck JN, Amoah AS, et al. Point and period prevalences of eczema in rural and urban schoolchildren in Ghana, Gabon and Rwanda. J Eur Acad Dermatol Venereol 2012;26:488-94.*
40. *Rahimi Rad MH, Hejazi ME, Behrouzian R. Asthma and other allergic diseases in 13-14-year-old schoolchildren in Urmia, Iran. East Mediterr Health J 2007;13:1005-16.*
41. *Peroni DG, Piacentini GL, Bodini A, et al. Prevalence and risk factors for atopic dermatitis in preschool children. Br J Dermatol 2008;158:539-43.*
42. *Suh M, Kim HH, Sohn MH, et al. Prevalence of allergic diseases among Korean school-age children: a nationwide cross-sectional questionnaire study. J Korean Med Sci 2011;26:332-8.*
43. *Montefort S, Ellul P, Montefort M, et al. A decrease in the prevalence and improved control of allergic conditions in 13- to 15-yr-old Maltese children (ISAAC). Pediatr Allergy Immunol 2011;22:e107-11.*
44. *Barraza-Villarreal A, Hernandez-Cadena L, Moreno-Macias H, et al. Trends in the prevalence of asthma and other allergic diseases in schoolchildren from Cuernavaca, Mexico. Allergy Asthma Proc 2007;28:368-74.*
45. *Zivkovic Z, Vukasinovic Z, Cerovic S, et al. Prevalence of childhood asthma and allergies in Serbia and Montenegro. World J Pediatr 2010;6:331-6.*
46. *Czarnobilska E, Obtulowicz K, Dyga W, et al. Contact hypersensitivity and allergic contact dermatitis among school children and teenagers with eczema. Contact Dermatitis 2009;60:264-9.*
47. *Suarez-Varela MM, Alvarez LG, Kogan MD, et al. Diet and prevalence of atopic eczema in 6 to 7-year-old schoolchildren in Spain: ISAAC phase III. J Investig Allergol Clin Immunol 2010;20:469-75.*
48. *Batlles Garrido J, Torres-Borrego J, Bonillo Perales A, et al. Prevalence and factors linked to atopic eczema in 10- and 11-year-old schoolchildren. Isaac 2 in Almeria, Spain. Allergol Immunopathol (Madr) 2010;38:174-80.*
49. *Larsson M, Hägerhed-Engman L, Sigsgaard T, et al. Incidence rates of asthma, rhinitis and eczema symptoms and influential factors in young children in Sweden. Acta Paediatr 2008;97:1210-5.*
50. *Shamssain MH, Shamsian N. Prevalence and severity of asthma, rhinitis, and atopic eczema: the North East Study. Arch Dis Child 1999;81:313-7.*

51. *Mortz CG, Lauritsen IM, Bindlsev-Jensen C, Andersen KH. Prevalence of atopic dermatitis, asthma, allergic rhinitis, and hand and contact dermatitis in adolescents. The Odense Adolescence Cohort Study on Atopic Diseases and Dermatitis. Br J Dermatol 2001;144;523-32.*
52. *Schultz Larsen F, Diepgen T, Svensson A. The occurrence of atopic dermatitis in north Europe: an international questionnaire study. J Am Acad Dermatol 1996;34:760-4.*
53. *Schafer T, Vieluf D, Behrendt H, et al. Atopic eczema and other manifestations of atopy: results of a study in East and West Germany. Allergy 1996;51(8):532-9.*
54. *Bockelbrink A, Heinrich J, Schäfer I, et al.; LISA Study Group. Atopic eczema in children: another harmful sequel of divorce. Allergy 2006;61:1397-402.*
55. *von Mutius E, Fritzsch C, Weiland SK, et al. Prevalence of asthma and allergic disorders among children in united Germany: a descriptive comparison. BMJ 1992;305(6866):1395-9.*
56. *Tamer E, Ilhan MN, Polat M, et al. Prevalence of skin diseases among pediatric patients in Turkey. J Dermatol 2008;35:413-8.*
57. *Emerson RM, Williams HC, Allen BR. Severity distribution of atopic dermatitis in the community and its relationship to secondary referral. Br J Dermatol 1998;139:73-6.*
58. *Laughter D, Istvan JA, Tofte SJ, Hanifin JM. The prevalence of atopic dermatitis in Oregon schoolchildren. J Am Acad Dermatol 2000;43:649-55.*
59. *Plunkett A, Merlin K, Gill D, et al. The frequency of common nonmalignant skin conditions in adults in central Victoria, Australia. Int J Dermatol 1999;38:901-8.*
60. *Thyssen JP, Linneberg A, Menné T, et al. The effect of tobacco smoking and alcohol consumption on the prevalence of self-reported hand eczema: a cross-sectional population-based study. Br J Dermatol 2010;162:619-26.*
61. *Worm M, Forschner K, Lee HH, et al. Frequency of atopic dermatitis and relevance of food allergy in adults in Germany. Acta Derm Venereol 2006;86:119-22.*
62. *Saeki H, Tsunemi Y, Fujita H, et al. Prevalence of atopic dermatitis determined by clinical examination in Japanese adults. J Dermatol 2006;33:817-9.*
63. *Smith-Sivertsen T, Tchachtchine V, Lund E. Atopy in Norwegian and Russian adults: a population-based study from the common border area. Allergy 2003;58:357-62.*
64. *Bo K, Thoresen M, Dalgard F. Smokers report more psoriasis, but not atopic dermatitis or hand eczema: results from a Norwegian population survey among adults. Dermatology 2008;216:40-5.*
65. *Spiewak R. Occupational Dermatoses in Agriculture: Epidemiology, Etiopathogenesis, Risk Factors. Czelej Publishers, Lublin 2002. (In Polish)*
66. *Spiewak R. Contact eczema. Post Dermatol Alergol 2009;26:375-377.*
67. *Spiewak R. Immunotherapy of allergic contact dermatitis. Immunotherapy 2011;3:979-996.*
68. *Spiewak R. Patch testing for contact allergy and allergic contact dermatitis. Open Allergy J 2008;1:42-51.*
69. *Czarnobilska E, Obtulowicz K, Dyga W, Spiewak R. The most important contact sensitizers in Polish children and adolescents with atopy and chronic recurrent eczema as detected with the extended European Baseline Series. Pediatr Allergy Immunol 2011;22:252-256.*
70. *Czarnobilska E, Dyga W, Krzystyniak D, et al. Influence of environment exposures on the frequency of contact allergies in children and adolescents. Ann Agric Environ Med 2012 23;19:11-6.*
71. *Shenefelt PD. Descriptive epidemiology of contact dermatitis in a university student population. Am J Contact Dermat 1996;7:88-93.*
72. *Akhavan A, Cohen SR. The relationship between atopic dermatitis and contact dermatitis. Clin Dermatol 2003;21:158-162.*
73. *Spiewak R. Pesticides and skin diseases in man. In: Rathore H, Nolet LM, editors. Pesticides: Evaluation of Environmental Pollution. Boca Raton: CRC Press; 2012: 525-542.*
74. *Wüthrich B. Zur Genese des Bäckerekzems. Hautarzt 1970;21:214-218.*
75. *Hjorth N, Roed-Petersen J. Occupational protein contact dermatitis in food handlers. Contact Dermatitis 1976;2:28-42.*
76. *Kanerva L, Jolanki R, Toikkanen J. Frequencies of occupational allergic diseases and gender differences in Finland. Int Arch Occup Environ Health 1994;66:111-6.*
77. *Hansen KS, Petersen HO. Protein contact dermatitis in slaughterhouse workers. Contact Dermatitis 1989;21:221-4.*
78. *Hay RJ. Malassezia, dandruff and seborrhoeic dermatitis: an overview. Br J Dermatol 2011;165 (Suppl 2): 2-8.*
79. *Lally A, Casabonne D, Newton R, Wojnarowska F. Seborrheic dermatitis among Oxford renal transplant recipients. J Eur Acad Dermatol Venereol 2010;24:561-4.*
80. *Moehrle M, Dennenmoser B, Schlagenhauff B, et al. High prevalence of seborrhoeic dermatitis on the face and scalp in mountain guides. Dermatology 2000;201:146-7.*
81. *Wollina U. Pompholyx: a review of clinical features, differential diagnosis, and management. Am J Clin Dermatol 2010;11:305-14.*
82. *de Boer EM, Bruynzeel DP, van Ketel WG. Dyshidrotic eczema as an occupational dermatitis in metal workers. Contact Dermatitis 1988;19:184-8.*
83. *Coenraads PJ. Hand eczema is common and multifactorial. J Invest Dermatol 2007;127:1568-70.*
84. *Dotterud LK, Falk ES. Contact allergy in relation to hand eczema and atopic diseases in north Norwegian schoolchildren. Acta Paediatr 1995;84:402-6.*
85. *Dorynska A, Pasich J, Lach L, et al. Self-reported hand, wrist and forearm dermatitis in a random sample of 691 Polish medical professionals, firefighters and students. Contact Dermatitis 2010;63(Suppl 1):96-7.*
86. *Meding B. Epidemiology of hand eczema in an industrial city. Acta Derm Venereol Suppl (Stockh) 1990;153:1-43.*
87. *Meding B, Järvholm B. Hand eczema in Swedish adults - changes in prevalence between 1983 and 1996. J Invest Dermatol 2002;118:719-23.*
88. *Montnemery P, Nihlen U, Lofdahl CG, et al. Prevalence of hand eczema in an adult Swedish population and the relationship to risk occupation and smoking. Acta Derm Venereol 2005;85:429-32.*
89. *Belsito DV. Occupational contact dermatitis: etiology, prevalence, and resultant impairment/disability. J Am Acad Dermatol 2005;53:303-13.*
90. *Meyer JD, Chen Y, Holt DL, et al. Occupational contact dermatitis in the UK: a surveillance report from EPIDERM and OPRA. Occup Med (Lond) 2000;50:265-73.*
91. *Diepgen TL, Coenraads PJ. The epidemiology of occupational contact dermatitis. Int Arch Occup Environ Health 1999;72:496-506.*
92. *Spiewak R. Skin diseases related to farm work - a questionnaire survey of 145 Polish farmers in Lublin Region. Post Dermatol Alergol 2001;18:194-199.*
93. *Spiewak, R. Contact dermatitis in atopic individuals. Curr Opin Allergy Clin Immunol 2012 (in press).*

A 10-Year Retrospective Review of Non-Scarring Alopecia in a Tertiary Hospital in Malaysia

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Abstract

Background Non-scarring alopecia is a common hair disorder with paucity of clinical reviews.

Objectives We aim to study the spectrum of non-scarring alopecia, its' demographic, clinical and treatment pattern among patients at University Malaya Medical Centre.

Methodology We have retrospectively reviewed the demography, clinical characteristics and treatment of non-scarring alopecia at University Malaya Medical Centre (UMMC). A total of 154 records were reviewed.

Results A majority of patients had alopecia areata (28.6%), followed by androgenetic alopecia (12.3%), telogen effluvium (3.2%), tineacapitis (2.6%) and unspecified hair loss (53.2). Treatment for alopecia areata included topical steroids (53.3%), intralesional steroids (26.7%), topical minoxidil (17.8%), oral steroids (11.1%), oral finasteride (2.2%) and oral azathiopine (2.2%). Prescribed treatment for androgenetic alopecia comprised of topical minoxidil (68.1%) or oral finasteride (10.5%).

Conclusion We concluded that alopecia areata was the most common cause of non-scarring alopecia diagnosed at UMMC and deduced that the high number of patients diagnosed with unspecified hair loss was attributed to the lack of confidence amongst out-patient physicians in diagnosing the cause of alopecia.

Keywords: *alopecia areata, androgenetic alopecia, telogen effluvium, anagen effluvium and alopecia mucinosa*

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Introduction

There is a current paucity of information in non-scarring alopecia, especially in South-east Asia. Non-scarring alopecia is more common than scarring alopecia and includes alopecia areata (AA), androgenetic alopecia (AGA), telogen effluvium, trichotillomania, tinea capitis and non-specific alopecia.

Alopecia areata (AA)

AA is relatively common in childhood[1,2], with a prevalence of 3.8% in Singapore[2]. It is linked to autoimmune endocrinopathies (e.g. type I diabetes mellitus, thyroid diseases, Addison's diseases), pernicious anemia and vitiligo[3,4].

Stress is a known factor in onset and aggravation[5]. AA also has a genetic component, with up to 20% of patients having a family history[6]. Onset of AA is also early, occurring before adulthood in 44% of patients[7].

Androgenetic alopecia (AGA)

A common cause of alopecia in adults[1], AGA has a higher frequency in Caucasians (approximately 50%)[8] than in Asians[9]. In Korea, a prevalence of approximately 14.1% (Norwood types ≥III) was observed in men and 5.6% in women (Ludwig scale)[10]. In China, this was 21.3% in men and 6.0% in women[11], and a much higher 63% in Singaporean men[12]. The severity of AGA increases with age[10] and presence of family history[13], which is also predictive of early-onset[13]. Smoking[13] and metabolic syndrome[14] have been found to play a role in the development of AGA.

With different prevalence data in Asians compared to Caucasians, a better understanding of the aetiology and current management of non-scarring alopecia in the region is needed. Hence, we aim to study the spectrum of non-scarring alopecia, its' demographic, clinical and treatment pattern among patients in University Malaya Medical Centre. A long-term retrospective study may therefore impact the treatment and diagnosis of this spectrum of conditions.

Methods

This was a retrospective review of patients diagnosed with non-scarring alopecia from January 2000 to December 2009 in a tertiary hospital in Malaysia. Patients classified as having the following conditions under the ICD-10 (International Classification of Diseases) were recalled from the dermatology and general out-patient database: alopecia areata (L63), androgenic alopecia (L64) and other non scarring hair loss (L65). This included male-pattern baldness, telogen effluvium, anagen effluvium and alopecia mucinosa but excluded trichotillomania (F63.3).

A total of 154 patients were included in the analysis. Data on demography, clinical characteristics and treatment of these patients were computed into a standard case report form in Microsoft Infopath 2007. These data were then exported to Microsoft Excel 2007 before being converted into SPSS 17.0 files. The continuous data were analyzed via means and standard deviations.

Workflow of retrospective review:

1.	Retrieval of patient cases from database (n=306)
2.	Exclusion of inconsistent diagnoses; Final patient cohort (n=154)
3.	Patients divided into subgroups for further analysis AA (n=44) AGA (n=19)

Results

A total of 154 medical records were reviewed. The mean age of patients at presentation was 24.6 ± 11.8 years (range 3-62 years). Of these, the majority were adults (70.1%), with 29.9% under the age of 16. There were slightly more females, with a male to female ratio of 1:1.03. The main ethnic groups were well-represented, with 31.8% Malays, 33.1% Indians, 27.3% Chinese and 7.8% classified as 'others'. More than a quarter of patients were diagnosed with AA (44, 28.6%), followed by AGA (19, 12.3%), telogen effluvium (5, 3.2%), tinea capitis (4, 2.6%) and unspecified hair loss (82, 53.2%). (Table I)

The co-morbidities present were anaemia (9, 5.8%), thyroid disorders (4, 2.6%), atopy (3, 1.9%) and type 2 diabetes mellitus (3, 1.9%). Stress was identified in 9 patients (5.8%) and 8 patients (5.1%) had family history of alopecia. Fourteen percent of patients received prior treatment, either from other doctors, traditional healers or over-the-counter purchase. More than a third of patients were lost to follow-up after first consultation (40.3%).

Table 1 Ethnic composition of AA and AGA.

Ethnic group	Number of patients (AA, AGA, etc)	Number with AA (28.2% of total patients)	Number with AGA (12.2% of total patients)
Chinese	42 (27.3%)	5 (26.3%)	10 (22.7%)
Malay	49 (31.8%)	8 (42.1%)	15 (34.1%)
Indian	51 (33.1%)	5 (26.3%)	14 (31.8%)
Others	12 (7.8%)	1 (5.3%)	5 (11.4%)
Total	**154**	**19**	**44**

Alopecia Areata (n=44)

The mean age of patients with AA was 24.7 ±11.6 years (range 5-62 years) with a mean age of onset at 24.2 ± 11.9 years (range 4-62 years). Nine (20.5%) were less than 16 years old. A majority of patients were Malays (34.1%), followed by Indians (31.8%), Chinese (22.7%) and 'others' (11.4%). We had three patients with type 2 diabetes, anaemia and thalassemia traits respectively. None of the patients were found to have a positive family history of alopecia, thyroid disorder, vitiligo, atopic dermatitis and connective tissue disease. Stress was identified in 3 patients (6.7%). More than half received topical steroids (53.3%). This was followed by intralesional steroids (26.7%), topical minoxidil (17.8%), oral steroids (11.1%), oral finasteride (2.2%) and azathiopine used concurrently with oral steroids (2.2%).

Androgenetic Alopecia (n=19)

The mean age of patients with AGA was 25.4 ± 2.2 years (range 17-55 years) and the mean age of onset was 23.3 ± 9.6 years (range 16-55 years). Unsurprisingly, the ethnic composition of these patients was Malays (42.1%), Indians (26.3%), Chinese (26.3%) and others (5.3%). Five (26.3%) had a positive family history of alopecia. Recommended treatment was predominantly topical minoxidil (68.1%), followed by oral finasteride (10.5%).

Discussion

The prevalence of AA and AGA was similar to other studies conducted in the region[10-12]. The most common confirmed cause of non-scarring alopecia was AA. However, the significant proportion of unspecified alopecia (51.9%) reflects inexperience and lack of confidence in primary physicians. This inability to provide a specific diagnosis may hamper management of non-scarring alopecia, thus necessitating in-depth understanding among physicians on common causes and clinical presentations.

Age groups affected by non-scarring alopecia

Non-scarring alopecia affects adults mainly in their 3rd decade of life but also affects patients below 16 years old. AA patients were found to be younger in our review, with our paediatric patients almost double that of those reported in China[15]. AGA patients were also younger compared to a Singaporean review reporting the highest age distribution at 37-46 years old[12].The mean duration to presentation was longer by about 18 months for AGA compared to AA. We postulate that this could be due to slower and more subtle presentation of AGA. Moreover, patients might have perceived AGA as part of natural aging and not a medical condition.

Autoimmune association

Autoimmune association is well recognized with AA, with a varying degree of association with vitiligo (0.4%-4.1%)[15,16]. None of our patients had vitiligo, but this could be attributed to the small sample size.

Genetic correlation

Genetic inheritance is the only significant factor leading to AGA and AA. Nevertheless, the low incidence of patients with family history (5.5%) in our data could be due to under-reporting.

Stress correlation

Similarly, we also had fewer patients identifying stress preceding the onset of hair loss, as opposed to figures ranging from 9.5%-68.9% in other reviews[17]. The high number of patients lost to follow-up may be due to certain factors such as lack of efficacy of prescribed treatment or limited treatment options. Other factors include the self-limiting nature of AA and the ambiguity of the diagnoses given. Studies showed that more than 70% of AGA patients seek non-medical sources of treatment[12]. Therefore, these patients may have sought alternative treatment after the initial counselling and management at the hospital.

Limitations

The low number of patients involved in the study may affect the statistical power of this review. However, the main limitation of our study is the retrospective design, thus, the possibility of under-documentation. Having different doctors involved in diagnosis could result in a non-standardize assessment. Patients could also under-report family history or wrongly describe their symptoms if a doctor-patient language barrier exists, given the three different predominant ethnic groups. A standardized pictorial description of alopecia eradicating the issue of comprehension or extensive interviewing of family members could address this, but may not be a viable option.

Conclusion

Our review showed the most commonly diagnosed non-scarring alopecia was AA followed by AGA. Nevertheless, the relatively high numbers of unspecified hair loss cases indicate a need for physician education to enable accurate diagnosis and optimal management of various causes of non-scarring alopecia.

Reference

1. *Tosti A, Lorizzo M, Piraccini BM. Androgenetic Alopecia in Children: Report of 20 Cases. British J Dermatol 2005; 152: 556-9.*
2. *Tan E, Tay YK, Goh CL, Chin Giam Y.The pattern and profile of alopecia areata in Singapore--a study of 219 Asians. Int J Dermatol2002 Nov;41 (11):748-53.*
3. *Puavilai S, et al. Prevalence of thyroid diseases in patients with alopecia areata. Int J Dermatol1994; 33: 632-3.*
4. *Koev DG. Diabetes mellitus. Diabetes mellitus type LADA. In: Losanov BS, ed. Endocrinology. Sofia: Academic Press Marin Drinov, 2000: 838.*
5. *Liana M, Vasile B. Stress in Patients with Alopecia Areata and Vitiligo. JEADV 2007; 21: 921-8.*
6. *McDonaugh AJG, Messenger AG. The Pathogenesis of Alopecia Areata.Dermatol.Clin1996; 14: 661-70.*
7. *Bolduc C, Elston DM, et al. Alopecia Areata – Medscape Reference.http://emedicine.medscape.com/article/1069931-overview. Accessed 7 July 2011.*
8. *Tsuboi R, Tanaka T, Nishikawa T, Ueki R, Yamada H, Katsuoka K, et al. A randomized, placebo-controlled trial of 1% topical minoxidil solution in the treatment of androgenetic alopecia in Japanese women. Eur J Dermatol2007 Jan-Feb; 17(1):37-44.*
9. *Feinstein RP, Elston DM, et al. Androgenetic Alopecia - Medscape Reference. http://emedicine.medscape.com/article/1070167-overview. Accessed 7 July 2011.*
10. *Paik JH, Yoon JB, Sim WY, Kim BS, Kim NI. The prevalence and types of androgenetic alopecia in Korean men and women. Br J Dermatol2001 Jul;145 (1):95-9.*
11. *Wang TL, Zhou C, Shen YW, Wang XY, Ding XL, Tian S, et al. Prevalence of androgenetic alopecia in China: a community-based study in six cities. Br J Dermatol2010 Apr; 162 (4):843-7.*
12. *Tang PH, Chia HP, Cheong LL. A Community Study of Male Androgenetic Alopecia in Bishan, Singapore. Singapore Med J 2000; 41(5): 202-5.*
13. *Su LH, Chen TH. Association of androgenetic alopecia with smoking and its prevalence among Asian men: a community-based survey. Arch Dermatol 2007 Nov; 143 (11):1401-6.*
14. *Su LH, Chen TH. Association of androgenetic alopecia with metabolic syndrome in men: a community-based survey. Br J Dermatol2010 Aug; 163 (2):371-7.*
15. *Xiao FL, Yang S, Liu JB, He PP, Yang J, Chui Y. The Epidemiology of Childhood Alopecia Areata in China: A Study of 226 Patients. Paeds Dermatol 2006; 23:13-18.*
16. *Tan E, Tay YK, Goh CL, Giam YC. The Pattern and Profile of Alopecia Areata in Singapore - a Study of 219 Asians. Int. J of Dermatol 2002; 41:748-53.*
17. *Kakourou T, Karachristou K, Chrousos G. A Case Series of Alopecia Areata in Children: Impact of Personal and Family History of Stress and Autoimmunity. JEADV 2007; 21:356-59.*

A 7-Year Retrospective Review of Skin Cancer at University Malaya Medical Centre: A Tertiary Centre Experience

Ch'ng CC[1], Wong SM[1], Lee YY[1], Rokiah I[1], Jayalaskmi Pailoor[2]

Abstract

Introduction Skin cancer is ranked the ninth commonest cancer among males and tenth among females in Malaysia.

Objectives To review the pattern of skin cancers at University Malaya Medical Centre (UMMC).

Methods This is a retrospective review of all histo-pathological confirmed skin cancers at UMMC from 2004 till 2010.

Results Among the 155 patients reviewed, basal cell carcinoma (BCC) was the commonest skin cancer (44.5%), followed by squamous cell carcinoma (SCC) (27.1%) and malignant melanoma (MM) (11.6%). The nodulo-ulcerative subtype made up 46% of all BCC while 50% of MM was of acral lentiginous subtype. Patients with BCC were significantly older (>60 years old), (p=0.003). A majority of skin cancers were found on the head and neck.

Conclusion BCC was the commonest skin cancer, with significantly older patients and located mainly on head and neck. MM was the least common skin cancer but associated with the highest mortality.

Keywords: *basal cell carcinoma, squamous cell carcinoma, malignant melanoma, Malaysia*

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Introduction

Skin cancer is the commonest malignancy among the Caucasians. Since the 1960s, the incidence of skin cancer among predominantly white populations has increased between 5% and 8% annually[1]. Skin cancer represents 20-30% of all neoplasms among Caucasians, but is only seen in 2-4% of Asians, and 1-2% of blacks and Asian Indians[1]. It appears that darker skin has relative protection against skin cancers.

The three main types of skin cancers are basal cell carcinoma (BCC), squamous cell carcinoma (SCC) and malignant melanoma (MM), each named after the cells from which it arises.

BCC is the commonest malignancy in Caucasians as well as Asians. In the United States, non-melanoma skin cancers constitute more than one third of all cancers with 75% made up of BCC[2]. MM, though not as common, accounts for 75% of deaths due to skin cancers[2]. A Japanese survey of skin cancers in 101 institutes from 1987 to1996 showed that 47% were BCC, 30% were SCC and 19% were MM[3].

A review of skin cancers by our neighbouring country Singapore from 2003 till 2007 showed similar results where BCC accounted for 54.5% of all skin cancers whilst SCC and MM accounted for 25.3% and 5.8% respectively[4-5].

Malaysia is a multiracial country, at latitude 2° 30' North of the equator, with a population of 28.31 million people, comprising Malays (51%), Chinese (23%), natives (11%), Indians (7%) and others (1%)[6]. A majority of this population have Fitzpatrick skin phototype III to V[7]. In 2003, the Malaysian National Cancer Registry (NCR) ranked skin cancer as the 9th commonest cancer among males and 10th among females[8]. However, to date, data on the demography and clinical characteristics of skin cancer is still lacking.

Aim

To analyse the demography and spectrum of histo-pathologically confirmed skin cancers at University Malaya Medical Centre (UMMC) from January 2004 to December 2010.

Table 1 Type and Subtypes of Skin Cancer.

Types of skin cancers		Number	Total
BC	Nodular/noduloulcerative	54	
	Superficial	2	
	Cystic	5	
	Unspecified	8	69* (44.5%)
SMC		42	42* (27.1%)
MM		18	18* (11.6%)
Others	Dermatofibrosarcoma protuberans	8	
	Extramammary Paget Disease	5	
	Kaposi Sarcoma	4	
	Verrucous Carcinoma	2	
	Malignant cutaneous adnexal tumor	1	
	Malignant eccrine cylindroma	1	
	Sebaceous carcinoma	1	
	Cutaneous lymphoma	8	30 (19.4%)

** Three patients had multiple skin cancers and one patient had concurrent pigmented and nodulo-ulcerative BCC.*

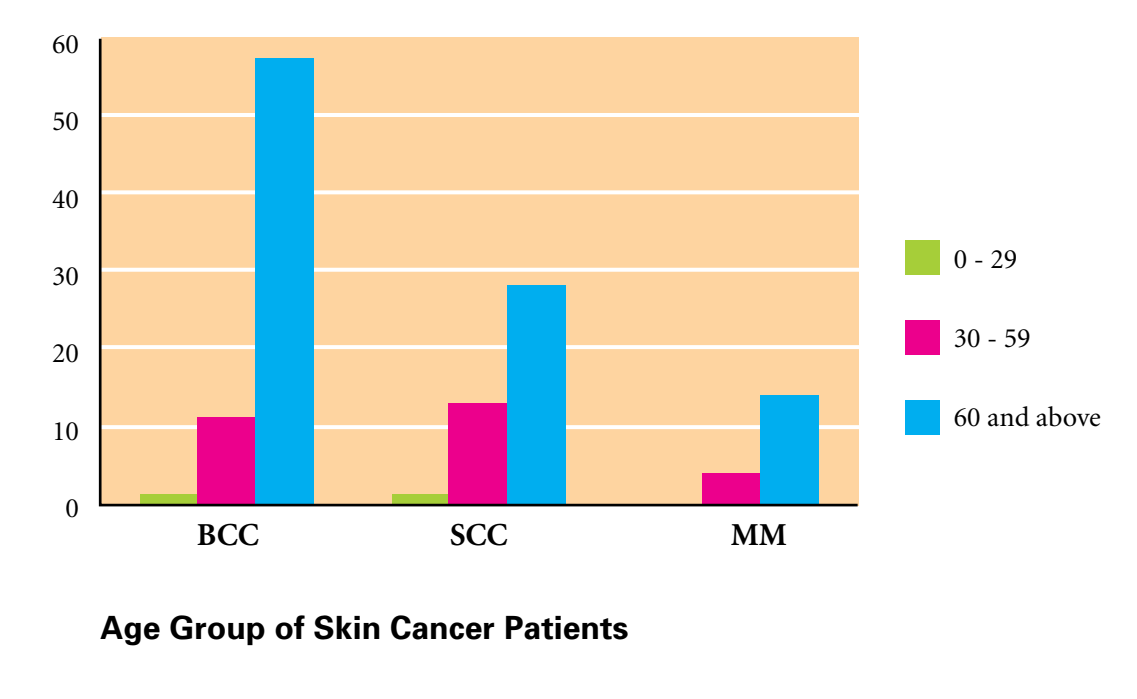

Fig 1 Skin Cancer, Distribution According to Age Group

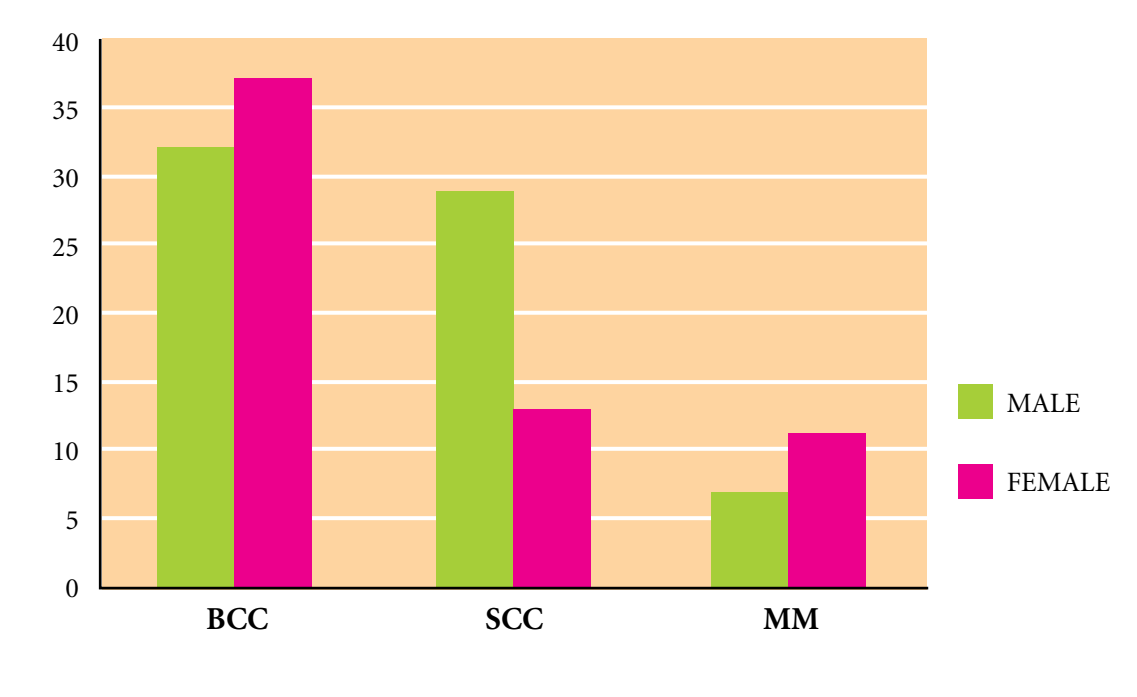

Fig 2 Skin Cancer, Gender Distribution.

Methodology

This is a retrospective study conducted at the Dermatology Unit, Department of Medicine and the Pathology Department at UMMC. Skin cancers were identified from the skin biopsy histopathology recording book and medical files were retrieved from our hospital medical records. All patients diagnosed with skin cancers from 1st January 2004 to 31st December 2010 were included. Patients were excluded if there was no confirmatory skin biopsy performed or there were missing or incomplete data.

Demographic data and clinical characteristics of patients were collected. These data were analysed using Statistical Package for Social Sciences (SPSS) version 18.0. Continuous variables were expressed as mean ± standard deviation (SD) and analysed using 2 sample independent t-tests. Categorical variables were described as frequencies and were analysed using Chi-Square test. Analysis was done for comparison of clinical characteristics based on regions of involvement and gender. The level of significance was set at p-value less than 0.05.

Results

A total of 172 histo-pathologically confirmed skin cancers were recorded between 2004 and 2010. Due to missing and incomplete medical records, only 155 patients were analysed. There were 80 males and 75 females. The mean age of lesion onset and diagnosis were 59.1± 16.7 years and 63.4 ± 15.3 years, respectively. The mean duration from onset of lesions until definite diagnosis was 3.5 ± 5.9 years.

BCC, SCC and MM were the three major skin cancers in our cohort followed by other rarer skin malignancies (Table 1). The mean age at diagnosis was the highest for BCC (68.0 ± 11.0 years), followed by MM (65.1 ± 11.1 years) and SCC (63.4 ± 15.9 years). Patients with BCC were significantly older (p=0.003) but this was not seen in patients with SCC (p=0.63) and MM (p=0.506) (Fig. 1).

Male to female ratio was 0.86:1 for BCC, 3:1 for SCC and 0.64:1 for MM (Fig. 2). There were significantly more male patients suffering from SCC (p=0.006). However, there was no significant gender preponderance among patients with BCC and MM (p=0.261, p=0.318).

Chinese patients made up the highest ethnic group for all types of skin cancers (Table 2). This was a significant finding as a review into UMMC 2004-2007 outpatient census revealed that Malay (44%) was the major ethnic group that frequents our outpatient clinic, followed by Chinese (31%), Indian (24%) and others (2%).

BCC was significantly more common on the head and neck (p<0.001) compared to other skin cancers, whilst MM was significantly commoner in the lower limbs (p<0.001). (Fig. 3)

A large proportion of patients with BCC had the nodulo-ulcerative variant (n=54, 78%) while the superficial variant made up the smallest proportion (n=2, 3%) (Fig. 4). In addition, a majority of the nodulo-ulcerative variant were pigmented (21/53, 38%).

Fifty percent (nine out of eighteen) of our cohort with a malignant melanoma had the acral lentiginous (ALM) type, although the number of patients with ALM was too small for statistical analysis. (Fig. 5) There were only two Indians in our study with malignant melanoma and both had the ALM subtypes. Among the five Malays with malignant melanoma, four had the ALM subtype. This trend was not seen among the Chinese, as only two out of ten patients had the ALM subtypes, seven had nodular melanoma and one had superficial spreading melanoma.

An analysis of the follow-up rate after skin biopsy showed a remarkably high defaulter rate of 25.8% in our cohort of patients.

Table 2 Racial distribution of Skin Cancer Patients.

Ethnicity	BCC	SCC	MM
Malay	13 (18.8%)	8 (19.0%)	5 (27.8%)
Chinese	51 (73.9%)	25 (59.5%)	10 (55.6%)
Indian	2 (2.9%)	7 (16.7%)	2 (11.1%)
Others	3 (4.4%)	2 (4.8%)	1 (5.6%)

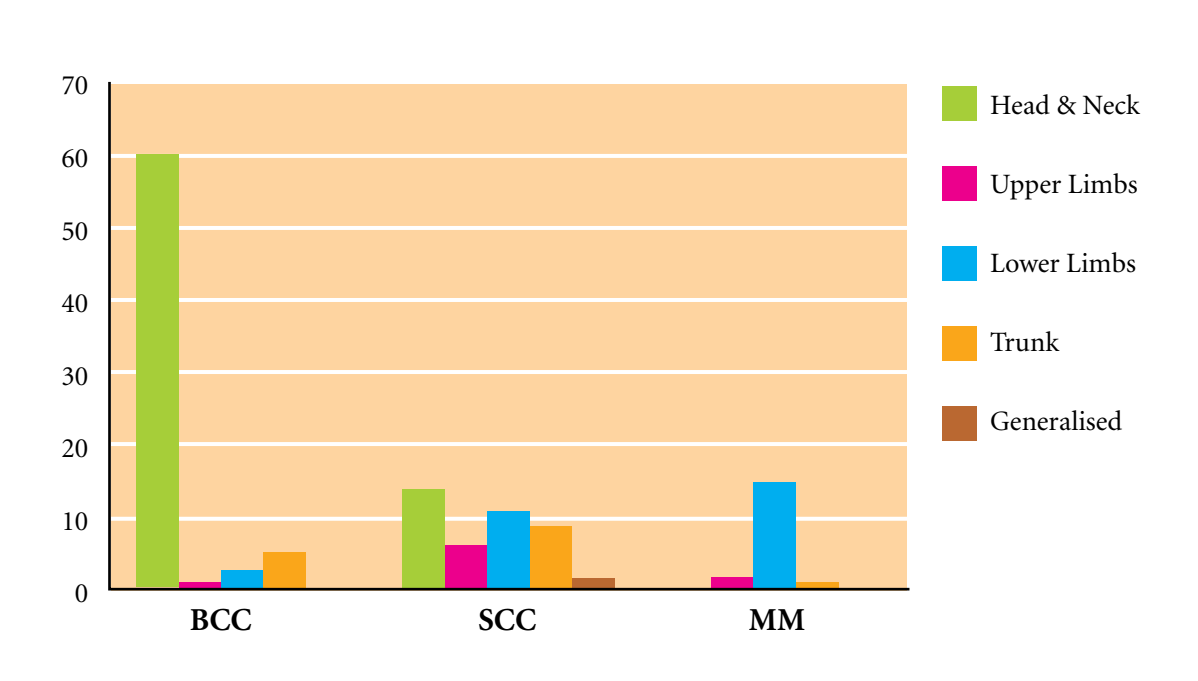

Fig 3 Site of Skin Cancers.

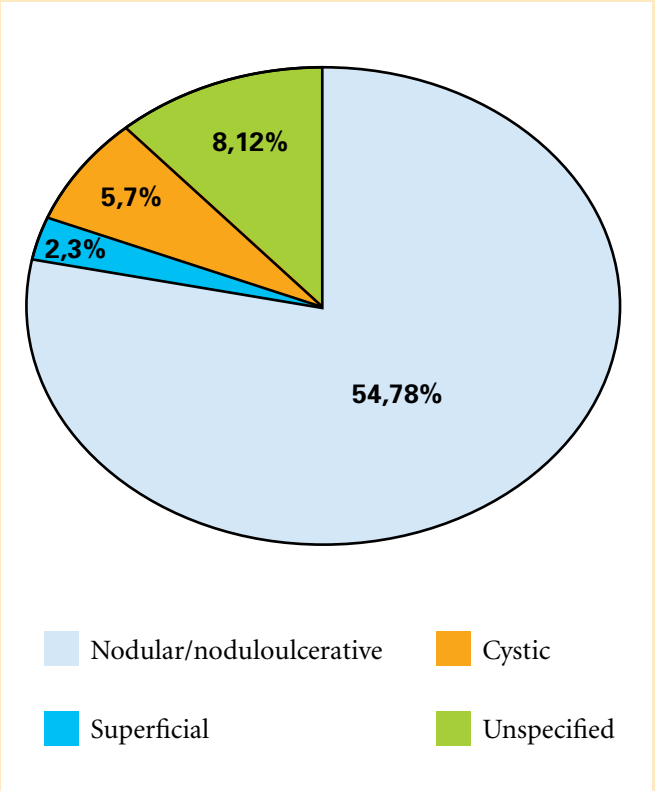

Fig 4 Basal Cell Carcinoma, Clinical Subtypes.

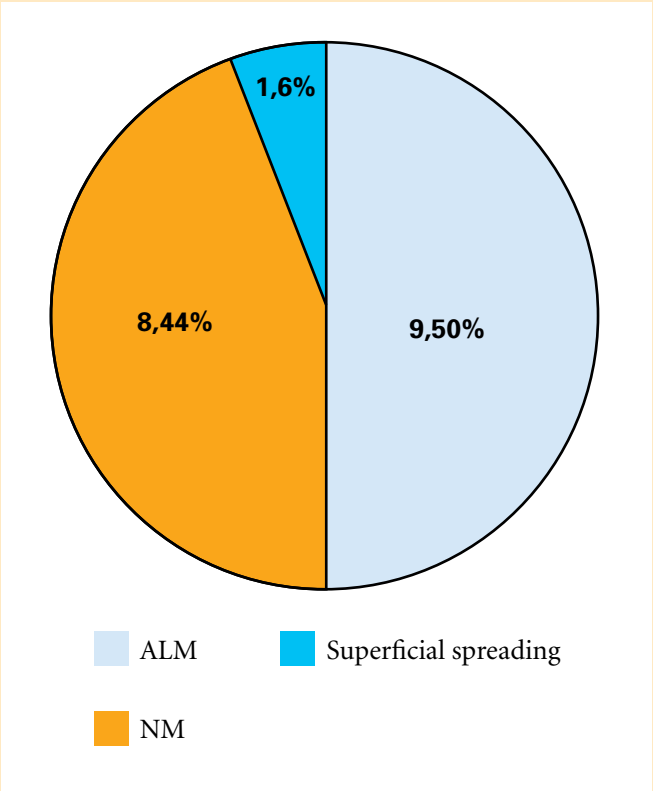

Fig 5 Malignant Melanoma, Subtypes.

Discussion

This study aims at gaining an insight into the pattern of skin cancers in our local population. BCC is the most common skin cancer, followed by SCC and melanoma. This is consistent with other studies in European and Asian populations[3-6, 9-17]. However, in comparison with a previous local study conducted in Sarawak, we had more cases of melanomas and less cutaneous lymphomas[16]. In addition, the patients with BCC in our cohort were from an older age group and were significantly older compared to other skin cancers. This is not surprising as BCC is well known for affecting the older population, whereas SCC and malignant melanoma are generally seen in the younger and middle-age groups.

In general, BCC has a male preponderance in the Caucasian population. Male to female ratio for BCC ranges from 1.4-1.9:1 in United States[18,19,20], 1.3-1.7:1 in Australia[12,21,22], and 1.1-1.4:1 in Europe[11,23,24]. Our study showed a slightly higher female preponderance with a male to female ratio of 0.86:1. This also corresponds with other Asian countries such as Singapore (male:female ratio 0.81:1)[3], Hong Kong (1:1.32-1.46)[25, 26] and Korea (0.9:1)[15]. Asian females are at least as likely as males to develop BCC despite the general greater awareness of sun protection. We postulate that this higher number of female patients may be because of higher detection rate due to a heightened tendency to seek medical treatment especially when most BCCs are found on the head and neck and pigmented lesions are more common in the Asian population.

Our study showed a male preponderance with a male:female ratio of 3:1 for SCC. This data is similar with other Asian and Western studies (Australia 3:112, US 2.4:1.017 and Singapore 1.29 to 2.4:13,10).

For MM, gender preponderance seems to vary with different geographical locations. An Australian study reported a male preponderance with a male to female ratio of 1.3:1.0[12], in the US, the ratio was 1:117, while in Europe, a slight female preponderance was reported[4]. Closer to our country, the male:female ratio was 0.93:1 in Japan[5] and 0.81:1 in Singapore[3]. Our study with a ratio of 0.64:1 concurred with our neighbouring countries which could indicate that Asian females may be more susceptible to malignant melanoma.

The Chinese ethnicity made up the highest number of patients for all types of skin cancers. This was significant as a review of our outpatient census from 2004-2007 revealed that the Malays were the major ethnic group that frequented our outpatient clinic (44%), followed by Chinese (31%), Indians (24%) and others (2%). These findings are not surprising as most Chinese have a lower Fitzpatrick skin phototype compared to the Malays and Indians, leading to lower protection from photodamage and subsequently, a higher risk of skin cancer.

In our study, most of the skin cancers were found on head and neck except for MM which is consistent with other studies[16-17]. The head and neck regions are sun-exposed areas and are more susceptible to skin cancers. Not surprisingly, the majority of BCC were of noduloulcerative subtype. But there is a difference as compared with Caucasian population[9-12] where a large portion of our cohort had the pigmented variant, perhaps related to increased epidermal melanin. Pigmented BCC can be easily confused with other benign pigmented lesions such as seborrhoeic keratosis, dermatosis papulosa nigra, dermatofibroma or nevus. Hence, it is important for clinician to have a high index of suspicion. On the other hand, the high frequency of acral lentiginous melanoma among patients with darker Fitzpatrick skin type, warrants further study to gain insight into the aetiology of malignant melanoma in Asian.

It is alarming that the defaulter rate for follow-up in our cohort of patients was as high as 25.8%. A third of patients with potentially fatal SCC did not return for follow-up. The lack of symptoms or knowledge about the disease may contribute to this high defaulter rate. Although skin cancers in the Asian population is generally lower than in the West, perhaps we should be spending more time educating our patients and increasing public awareness of skin cancers among the general population.

Conclusion

BCC was the commonest skin cancer in our cohort and found mainly on the head and neck regions. Patients with BCC were also older and mainly of Chinese ethnicity with a slight female preponderance. A majority of malignant melanomas in our centre were of the acral lentiginous melanoma subtype. Defaulter rate of skin cancer was high in our centre which demands urgent attention. There is a need for public education to increase awareness of skin cancer in our country. Health campaigns to educate the public on self skin examination and the importance of photoprotection may be beneficial especially in high risk individuals.

Reference

1. 1. *Gloster H, Neal K. Skin cancer in skin of color. J Am Acad Dermatol. 2006;55:741–60.*
2. 2. *Gloster HM, Brodland DG. The epidemiology of skin cancer. Dermatol Surg 1996: 22: 217-26.*
3. 3. *Ishihara K, Saida T, Yamamato A. Updated statistical data for malignant melanoma in Japan. Int J Clin Oncol 2001; 6: 109–16.*
4. 4. *Koh D, Wang H, Lee J, et al. Basal cell carcinoma, squamous cell carcinoma and melanoma of the skin: analysis of Singapore Cancer Registry data 1968–97. British Association of Dermatologists. 2003;148:1161–66.*
5. 5. *Lee HP, Ang PT, Chow KY et al. Cancer Incidence in Singapore 1968-2007. Singapore Cancer Registry Report No 7, 2000.*
6. 6. *Department of Statistics. Yearbook of Statistics, Malaysia 2009. March 2010: 2-33*
7. 7. *Fitzpatrick TB. The validity and practicality of sun reactive skin types I through VI. Arch Dermatol 1988;124: 869–71*
8. 8. *Gerard Lim Chin Chye, Halimah Yahay. Second Report of National Cancer Registry. Malaysia Cancer Registry. 2003.*
9. 9. *Halder RM, Bridgeman-Shah S. Skin cancer in African Americans. Cancer 1995;75:667-73.*
10. 10. *S L brown CJ Thirlaway, CM Lawrence. A 6 months prospective study of the incidence and management of dermatological malignancies in an NHS teaching hospital. Br J Dermatol 2005;153:74*
11. 11. *Holme SA, Malinovszky K, Roberts DL. Changing trends in non-melanoma skin cancer in South Wales 1988-98. Br J Dermatol 2000: 143: 1224-9*
12. 12. *Green A, Battistutta D, Hart V, et al. Skin cancer in a subtropical Australian population: incidence and lack of association with occupation. The Nambour Study Group. Am J Epidemiol. Dec 1 1996;144(11):1034-40*
13. 13. *Nakjang Y, Kullavanija P. Basal cell carcinoma: seven years' experience at the Institute of Dermatology in Bangkok. J Dermatol 1994; 21: 660–63*
14. 14. *Diepgen TL, Mahler V. The epidemiology of skin cancer. Br J Dermatol. Apr 2002;146 Suppl 61:1-6*
15. 15. *Cho S, Kim MH, Whang KK, Hahm JH. Clinical and histopathological characteristics of basal cell carcinoma in Korean patients. J Dermatol 1999; 26: 494–501.*
16. 16. *Yap FB. Clinical characteristics of basal cell carcinoma in a tertiary hospital in Sarawak, Malaysia. Int J Dermatol. 2010 Feb;49(2):176-9.*
17. 17. *Raasch BA, Buettner PG, Garbe C. Basal cell carcinoma: histological classification and body site distribution. Br J Dermatol. 2006;155(2):401-7.*
18. 18. *Miller DL, Weinstock MA. Nonmelanoma skin cancer in the United States: Incidence. J Am Acad Dermatol 1994; 30: 774–8.*
19. 19. *Serrano H, Scotto J, Shornick G et al. Incidence of nonmelanoma skin cancer in New Hampshire and Vermont. J Am Acad Dermatol 1991; 24: 574–9.*
20. 20. *Gray DT, Suman VJ, Su WPD et al. Trends in the population-based incidence of squamous cell carcinoma of the skin first diagnosed between 1984 and 1992. Arch Dermatol 1997; 133: 735–40.*
21. 21. *Buettner PG, Raasch BA. Incidence rates of skin Cancer in Townsville, Australia. Int J Cancer 1998; 78: 587–93.*
22. 22. *Kaldor J, Shugg D, Young B et al. Non-melanoma skin cancer: ten years of cancer-registry-based surveillance. Int J Cancer 1993; 53: 886–91.*
23. 23. *Ko CB, Walton S, Keczkes HPR et al. The emerging epidemic of skin cancer. Br J Dermatol 1994; 130: 269–72.*
24. 24. *Scottish Cancer Intelligence Unit ISD, National Health Service in Scotland. Cancer Registration Statistics Scotland 1990–95. Edinburgh: National Health Service, 1998.*
25. 25. *Cheng SY, Luk NM, Chong LY. Special features of non-melanoma skin cancer in Hong Kong Chinese patients: 10-year retrospective study. Hong Kong Medical Journal 2001;7: 22-28.*
26. 26. *Cheng SY. Non-melanoma skin cancer: A five-year retrospective study in Social Hygiene Service of Hong Kong. Hong Kong Journal of Dermatology and Vereneology. 1999; 7: 162-69.*

Squamous Cell Carcinoma Arising from Linear Porokeratosis in a Young Chinese Man

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Keywords: *skin cancer, extensive plaques, autosomal dominant*

Background

Porokeratosis is a specific disorder of epidermal keratinization, characterised histologically by the presence of cornoid lamella[1]. Linear is a distinct, mosaic variant of this autosomal dominant condition. There is a well recognized association between porokeratosis and malignancy, especially the linear variant which has the highest malignant potential[2, 3].

Case Report

A 25-year old gentleman presented to our dermatology clinic in 2004 with non-healing erosions within a pre-existing plaque on his left calf of one-year duration. The erosions were not preceded by any trauma. On further questioning, he claimed to have multiple asymptomatic cutaneous plaques over his face, trunk, upper and lower extremities since he was 3-months old.

The first lesion as noted by his mother was over his left calf, which subsequently increased in size and number and spreading in a linear pattern.

Apart from cosmetic disfigurement, he developed multiple nodular lesions on his left calf and thigh.These were not painful, itchy, did not ulcerate and were not associated with discharge. There was no family history of similar skin lesions or skin malignancy.

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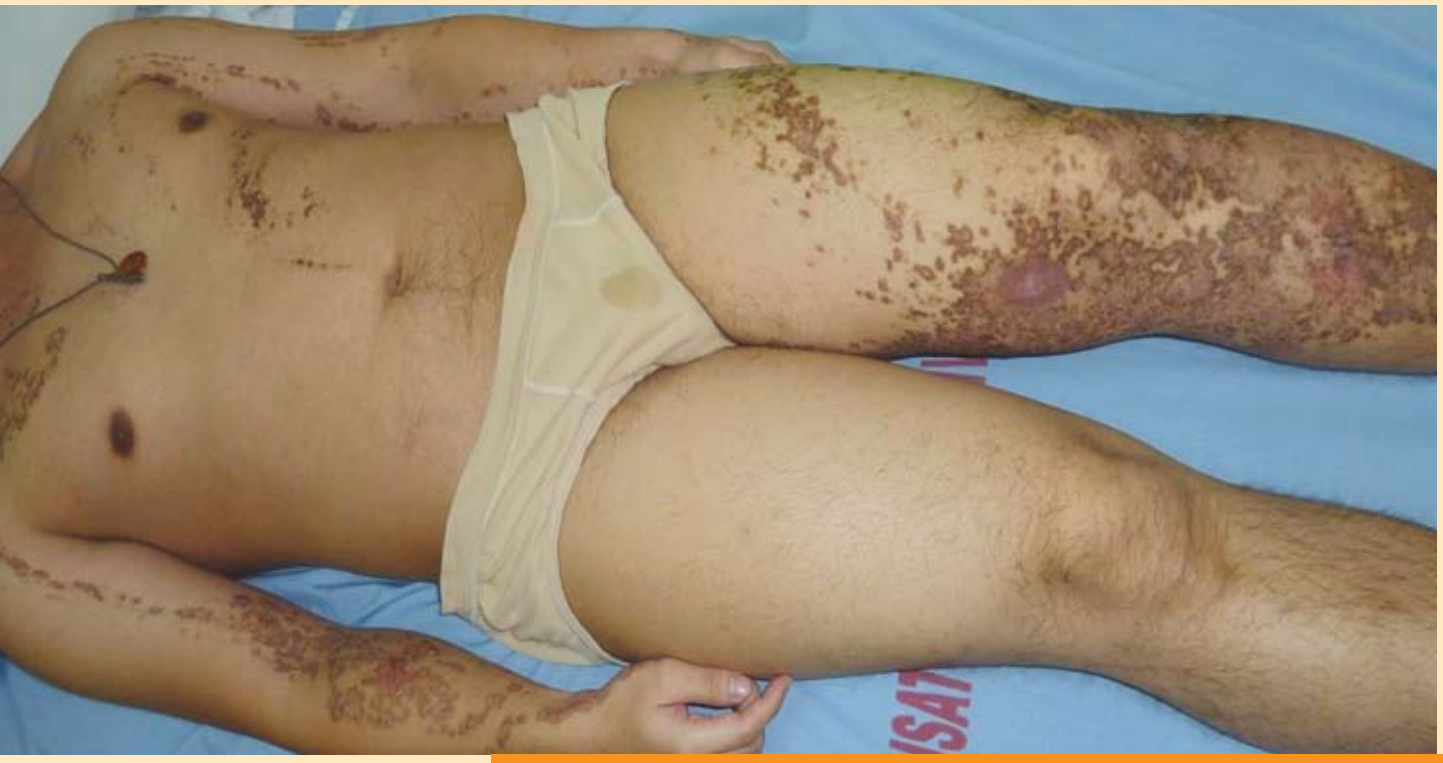

Fig 1
Linear cutaneous plaques on the anterior trunk, both upper and lower extremities in a Blaskho's distribution.

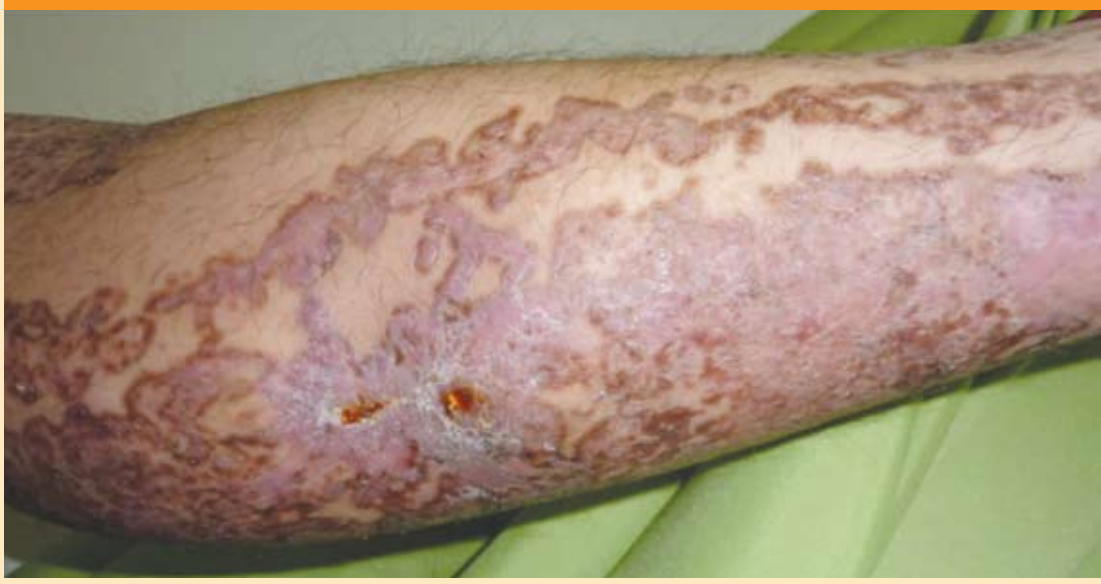

Fig 2 Excised nodule on his left calf leaving a crusted plaque.

Clinically, there were multiple hyperpigmented scaly plaques arranged in linear fashion along the Blaschko distribution. The plaques were atrophied with raised, well-defined, irregular margins and appeared more pigmented at the periphery than centrally. Over the medial aspect of his left calf and thigh, there were also several plaques, which are thicker and more hyperkeratotic (Figure 1 & 2).

Skin biopsies were taken from the margin of one lesion and another at a hyperkeratotic plaque over his left calf. The margin of the lesion showed features of porokeratosis such as focal areas of acanthosis, parakeratosis and foci of cornoid lamella. The hyperkeratotic plaque showed moderate to severe dysplasia.

The thickened plaques were treated with multiple sessions of cryotherapy. Concurrently, oral acitretin was commenced. Unfortunately, he did not notice any improvement despite taking acitretin. Hence, he defaulted on his follow-up and treatment after a year of acitretin.

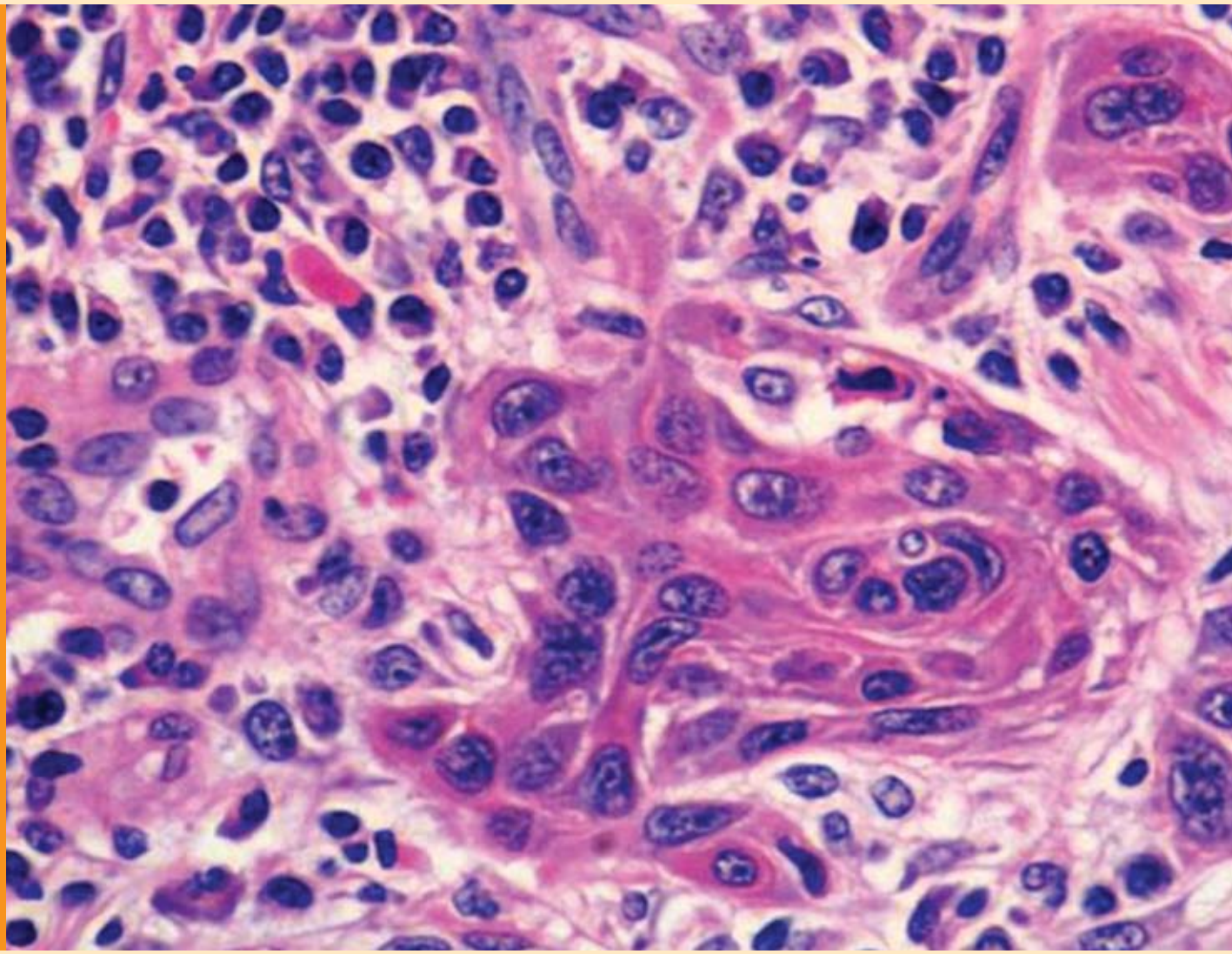

Fig 3 Hematoxylin-eosin stain. Original magnification X100. Dense inflammatory infiltrate with squamous cells arranged in discrete nests, cords and trabeculae extending into the underlying dermis and subcutis. Presence of mature and well developed keratin pearl.

We recently saw him, after a 4-year hiatus, with history of painful, gradually enlarging left calf nodule of 2-year duration. There was no associated discharge or bleeding. Clinically, there was a 0.5 - 2.0 cm diameter flesh-coloured firm nodule with superficial erosion seen. There was no regional lymphadenopathy. Incisional skin biopsy well differentiated squamous cell carcinoma with keratin pearls and intercellular bridging (Figure 3). He was subsequently referred to a plastic surgeon for total excision and skin grafting.

It typically presents as sharply demarcated hyperkeratotic plaques (annular, central atrophy, linear, punctate) surrounded by a thread-like elevated border that expands centrifugally.

There are a few clinical variants of porokeratosis, e.g. porokeratosis of Mibelli, disseminated superficial porokeratosis (DSP), disseminated superficial actinic porokeratosis (DSAP), porokeratosis palmaris et plantaris disseminata (PPPD), linear porokeratosis and punctate porokeratosis.

Our patient was diagnosed with disseminated linear porokeratosis, a rare autosomal dominant keratinization disorder that frequently presents unilaterally in a linear pattern and bears a close resemblance to linear verrucous epidermal neavus. However, our patient has a far more extensive presentation with the involvement of his trunk, upper and lower extremities in a Blashko distribution bilaterally as opposed to the commonly described unilateral distribution.

The pathogenesis of this condition is attributed to clones of abnormal epithelial cells, resulting in keratotic lesions that can progress to cutaneous neoplasia. The high rate of abnormal DNA and malignant features, e.g. p53 over-expression suggest an increased proliferative potential in affected keratinocytes[4]. The incidence of malignant degeneration in porokeratosis can be as high as 10%[4].

Otsuka et al had reported malignant degeneration and metastasis in this condition, which can be either be a basal cell carcinoma, Bowen's disease or squamous cell carcinoma, and most likely to occur in older adults[5]. A genetic mechanism of allelic loss may also be a representation in the initial step leading towards the development of cancer[3].

The chronicity of the plague and extensive involvement predisposes our patient to malignant transformation even though he did not have other frequently reported predisposition such as previous treatment with radiotherapy[6,7]. Being immunocompetent with no co-morbidities, the squamous cell carcinoma was found at the calf, a non-traumatic site consistent with the finding of Girla and Bhattacharya[8]. Other reported risk factors for the development of squamous cell carcinoma includes patients on immunosuppressive therapy, recipients of organ transplant, trauma sites, burn sites, Crohn's disease and end-stage liver disease[9, 10].

The currently available topical treatments for porokeratosis include 5-fluorouracil, imiquimod, and retinoids, as well as more destructive modalities such as cryotherapy, CO2 laser, curettage, excision, and dermabrasion. Oral acitretin has also been found to be useful in reducing the magnitude of disease, but there is usually recurrence of lesions upon its discontinuation[11]. Our patient showed no response to oral acitretin. Nevertheless, cryotherapy with liquid nitrogen appeared to reduce the thickness of the hyperkeratotic plaques.

Due to the localized nature of the squamous cell carcinoma, it was excised locally with a wide margin of clearance. We are monitoring for any possible transformation at the same site, or other new areas. It is prudent for dermatologists managing patients with linear porokeratosis to follow up with the patient over a period of time, and perform biopsies in the event of any cutaneous changes due to its preponderance for malignant degeneration, especially at the distal extremities.

Conclusion

Dermatologists managing patients with porokeratosis should follow them up closely and practice a low threshold for biopsies of any suspicious lesions to detect early dysplasia or malignancy.

Reference

1. *I.M. Friedberg, A.Z. Eisen, K. Wolff, Editors, Fitzpatrick's dermatology in general medicine (6th ed.), McGraw-Hill, New York (2003).*
2. *Otsuka F, Iwata M, Watanabe R, Chi HI, Ishibashi Y. Porokeratosis: clinical and cellular characterization of its cancer-prone nature. J Dermatol 1992; 19:702-706.*
3. *Happle R. Cancer proneness of linear porokeratosis may be explained by allelic loss. Dermatology 1997; 195:20-25.*
4. *Rongioletti F, Rebora A. Disseminated porokeratosis with fatal metastatic squamous cell carcinoma, an additional case of 'malignant disseminated porokeratosis.' Am J Dermatopathol 2002; 24:144-8.*
5. *Otsuka F, Someya T, Ishibashi Y. Porokeratosis and malignant skin tumours. J cancer Res Clin Oncol 1991; 117:55-60.*
6. *Sasson M, Krain AD. Porokeratosis and cutaneous malignancy: a review. Dermatol Surg 1996; 22: 339–342.*
7. *Cort DF, Abdel-Aziz AM. Epithelioma arising in porokeratosis of Mibelli. Br J Plast Surg 1972; 25:318-328.*
8. *Girla VS and Bhattacharya SK: Clinical study of Porokeratosis. Report of 10 cases. Interna J Dermatol 1976; 15:43-51.*
9. *Shane G. Silver, Richard I. Crawford. Fatal squamous cell carcinoma arising from transplant-associated porokeratosis. J Am Acad Dermatol 2003 Nov; 49(5):931-3.*
10. *Morton CA, et al. Porokeratosis and Crohn's disease. J Am Acad Dermatol 1995; 32:894.*
11. *Goldman, GD, Milstone LM. Generalized linear porokeratosis treated with etretinate. Arch Dermatol 1995; 131:496*

Pseudocyst of Ear

Kar, Atul Mohankar, Ajay Krishnan, Nitin Gangane

Keywords: *intracartilagenous swelling, auricle*

Introduction

Pseudocyst of the auricle is a rare, asymptomatic, cystic, swelling of the upper portion of the auricle. It results from spontaneous collection of an oily, serous fluid within an unlined intracartilaginous cavity[1]. The aetiology and pathogenesis of this condition is not known. Although various medical and surgical therapeutic approaches have been described, the treatment of pseudocyst of auricle is difficult and recurrences are frequent[2].

Case report

An 18 year old young boy presented with asymptomatic swellings in both the pinna of the ear since 15 years. There was no preceding history of trauma to the ear and his general condition was fair. On examination, there were 2 cystic, non-tender 1.5 x 2cm sized swellings over each of the ear lobule. A clinical diagnosis of pseudocyst of the auricle was made. The cyst was excised under all aseptic precautions and sent for histopathological examination which confirmed a keratin inclusion cyst.

Discussion

Pseudocyst of the auricle is characterized by a unilateral, asymptomatic, cystic swelling of the helix or the antihelix, most often located in the scaphoid fossa. Engel in 1966 first reported the pseudocyst of auricle in the Chinese[3]. This rare disorder results from spontaneous accumulation of a sterile, oily yellowish fluid, resembling olive oil. It is mostly observed in young adult males and presents clinically as a solitary, fluctuant, non-inflammatory swelling of the upper portion of the auricle with normal overlying skin.

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Fig 1 cyst in left ear lobule.

Fig 2 cyst in right ear lobule.

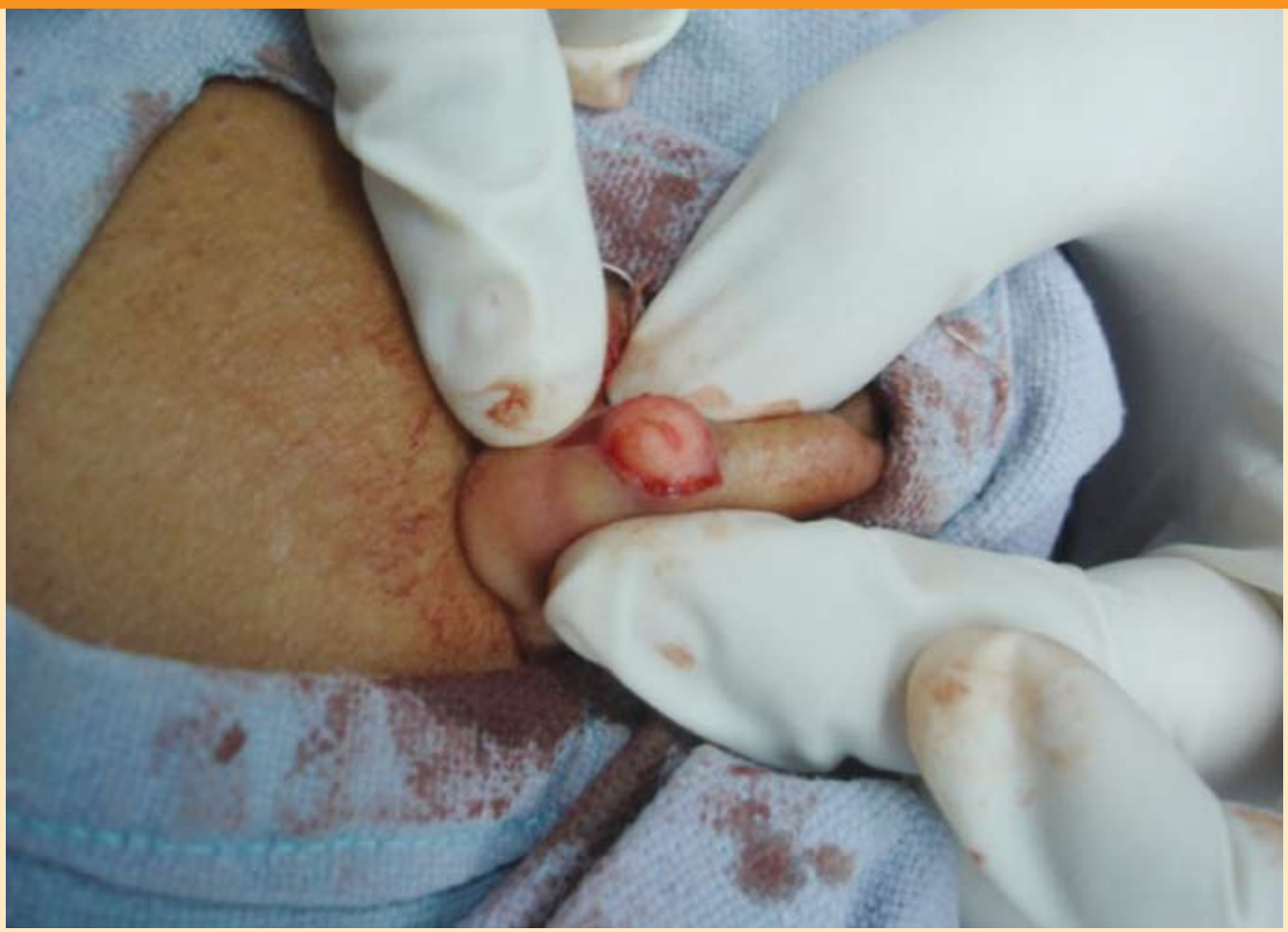

Fig 3 removal of cyst.

Histopathology reveals an intracartilaginous accumulation of fluid without an epithelial lining. The lack of epithelial lining led to the term 'pseudocyst'. In early lesions, the cystic space is surrounded by fibrosed cartilage while in some areas necrosis and total dissolution of the cartilage may be present. In later stages, intracavity foci of granulation tissue and more extensive intracartilaginous fibrosis is present[4]. The aim of treatment is to ensure successful resolution of the pseudocyst without damage to the healthy cartilage and to prevent its recurrences. Various treatments reported in literature include simple aspiration, intralesional injection of corticosteroids, and aspiration in combination with bolstered pressure sutures or plaster of Paris cast. More invasive techniques like incision and drainage of the cavity followed by its obliteration by curettage, sclerosing agent and pressure dressing; open deroofing that involves removal of the anterior cartilaginous leaflet of pseudocyst with repositioning of the overlying flap of skin have also been recommended. However, the invasive treatment modalities carry the risk of perichondritis complicated by formation of floppy ear or cauliflower deformity and may be followed by recurrences[5].

In our case, we removed the cyst by simple excision under local anaesthesia as an outpatient procedure. The patient has been followed up for next 6 months with no recurrence.

Conclusions

We report this case due to its rare site, and bilateral presentation.

Reference

1. *Cohen PR, Grossman ME. Pseudocyst of the auricle:case report and world literature reviwe. Arch Otolaryngol Head Neck Surg 1990; 116:1202-1204.*
2. *Harder M, Zachary C. Pseudocyst of the ear: Surgical treatment. J Dermatol Surg Oncol 1993; 19:585-588.*
3. *Engel D. Pseudocyst of the auricle in the Chinese. Arch Otolaryngol 1966; 83;197-202.*
4. *Heffner DK, Hyamo VJ. Cystic chondromalacia (endochondral pseudocyst) of the auricle. Arch Pathol Lab Med 1986; 110:740-743.*
5. *Schulte KW, Neumann NJ, Ruzicka T. Surgical pearl: The close fitting ear cover cast-a noninvasive treatment for pseudocyst of the ear. J Am Acad Dermatol 2001; 44:285-287*

A Case of Cutaneous and Paravertebral Infantile Haemangioma

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Keywords: *mediastinal mass, vascular tumour, seizures*

Introduction

Haemangiomas are common during infancy, affecting as many as 10% of infants. However their occurrence in the mediastinum is rare with an incidence of less than 0.5% of all mediastinal masses[1]. We present a case of mediastinal haemangioma with paravertebral extension complicated by neurological sequelae.

Case report

A 7 month-old infant presented with seizures and fever of one week duration and mild left hemiparesis. She was ventilated for status epilepticus and was treated with anticonvulsants and antibiotics for presumed meningitis. She was noted to have a mixed haemangioma measuring 7 by 4 cm over the back which appeared soon after birth and rapidly increased in size (Figure 1 and 2) There are areas of healed ulceration. There was another deep haemangioma over the upper right shoulder measuring 3 by 2 cm. Blood investigations showed no evidence of consumptive coagulopathy. Treatment with prednisolone 3 mg/kg/day, intravenous(IV) methylprednisolone, 8 courses of IV Vincristine 0.05mg/kg and propranolol 2 mg/kg/day was instituted as high output cardiac failure complicated her condition.

Magnetic resonance imaging (MRI) of the spine done at 8 and a half months of life showed an extensive vascular tumour from the level of skull base extending down to the cervical space and further down to bilateral retroperitoneal paravertebral spaces. Co-existence right subcutaneous posterior trunk mass is present communicating with intrathoracic mass and overall appearances are typical for haemangioma. (Figure 1 and 2)

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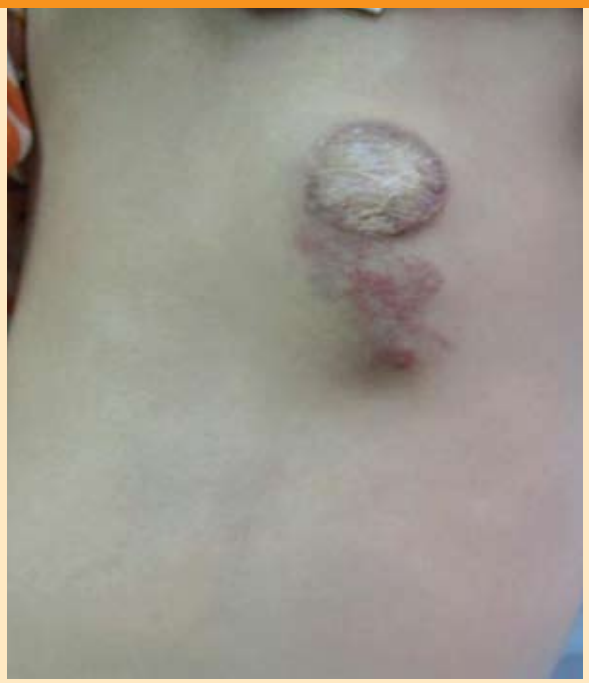

Fig 1 Right subcutaneous posterior trunk mass

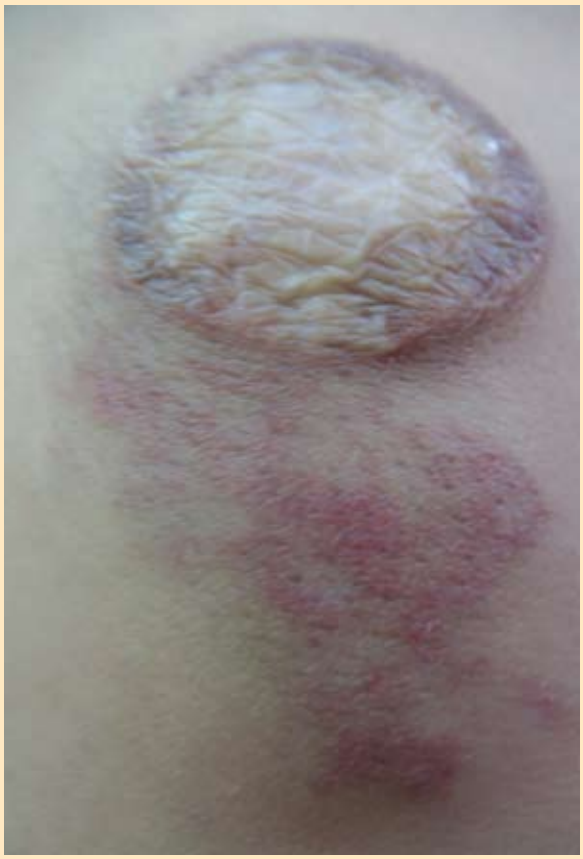

Fig 2 Close-up view of right subcutaneous posterior trunk mass.

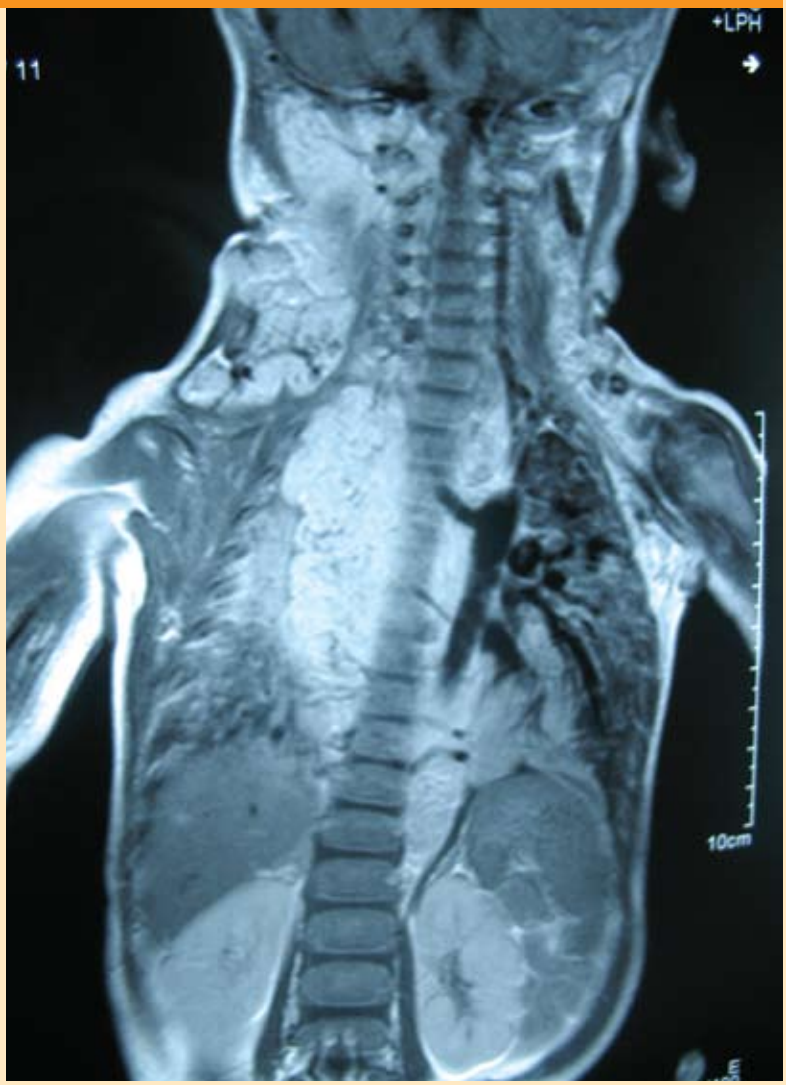

Fig 3 The prominent cerebral vascularization and pacchymeningeal enhancement is likely attributable to hyperdynamic circulation.

MRI brain shows right subdural collection with cortical laminar infraction in both parietal lobes. The prominent cerebral vascularization and pacchymeningeal enhancement is likely attributable to hyperdynamic circulation. (Figure 3)

After a prolonged stay in the ward for 3 months, she was discharged well. Unfortunately she was admitted a week later with seizures and complete right hemiparesis. Repeat CT brain showed massive left cerebral infarction at middle cerebral artery territory. Conservative management was opted after discussion with parents and she succumbed to her illness soon after.

Discussion

Haemangiomas are the most common tumours of childhood with most lesions (>60%) found in the head and neck[2]. The diagnosis of haemangioma was made clinically in our patient based on physical examination, absence of consumptive coagulopathy and supported by MRI findings. It is usually not present at birth as in our patient in contrast to vascular malformations.

Mediastinal extension of haemangiomas and paravertebral and intraspinal involvement are exceptional but potentially harmful because of the risk of neurological impairment[3]. Chiller et al demonstrated that in a series of 327 patients, isolated cutaneous infantile hemangioma associated with hemangioma of the mediastinum were found in only 3 patients (<1%)[4].

Our patient presented with neurological impairment due to rapid proliferation of her haemangioma which extended from the level of skull base down to bilateral retroperitoneal paravertebral spaces, also extending into the anterior mediastinum. She was treated aggressively with prednisolone, methylprednisolone, vincristine and propranolol but unfortunately did not respond to treatment. She developed multiple episodes of ischaemic stroke possibly due to steal phenomenon, compression effect or cerebrovasculature anomalies. It is unfortunate that we could not do a magnetic resonance angiogram (MRA) to delineate the cerebral vessels clearly as patient had already succumbed to her illness. Neurological complications are rare in true infantile haemangiomas unlike PHACES syndrome[5]. In PHACES syndrome neurologic complications arise mainly due to abnormality of cerebral vessels like progressive stenosis and occlusion of principal cerebral arteries. Our patient did not fulfill criteria for PHACES as she did not have a large cervicofacial haemangioma. However the presence of possible abnormal vessels between anterior and posterior cerebral vessels as seen in the MRI brain of the patient indicate that patient may have a variant of PHACES.

Conclusions

Cutaneous haemangiomas over the spine should be studied with ultrasound then MRI with gadolinium (if abnormalities are present on ultrasound) as they can be associated with intraspinal extension and dysraphic lesions[6]. Paravertebral extension of haemangioma and mediastinal haemangioma are rare but associated with risk of neurological complications. Our case highlights this potential complication and acts as reminder to physicians that imaging is essential in certain cases of infantile haemangioma.

Reference

1. *Sabharwal GK, Strouse PJ. Posterior mediastinal hemangioma. Pediatr Radiol 2005;35:1263-1266.*
2. *Metry D. Update on hemangiomas of infancy. Curr Opin Pediatr 2004;16:373-377.*
3. *Herna´ndez-Martin A, Torrelo A. Cutaneous and Paravertebral Infantile Hemangioma: Report of Two Cases. Pediatric Dermatology 2008;25:193-195.*
4. *Chiller KG, Passaro D, Frieden IJ. Hemangiomas of Infancy. Clinical Characteristics, Morphologic Subtypes, and Their Relationship to Race, Ethnicity, and Sex. Arch Dermatol 2002;138:1567-1576.*
5. *Heyer GL, Millar WS, Ghatan S, Garzon MC. The Neurologic Aspects of PHACE: Case Report and Review of the Literature. Pediatric Neurology 2006;35:419-424.*
6. *Frieden IJ, Haggstrom AN, Drolet BA et al. Infantile Hemangiomas – Current Knowledge, Future Directions. Pediatric Dermatology 2005;22:383-406.*

A Rare Case of Suspected Dystrophic Epidermolysis Bullosa Pruriginosa

Satya Wydya Yenny, Yuanita, Rina Gustia

Keywords: *: congenital skin anomaly, skin ulceration, skin atrophy*

Introduction

Inherited epidermolysis bullosa (EB) encompasses over 30 phenotypes or genotypes. A characteristic feature of all types of EB is the presence of recurrent blistering or erosions, the result of even minor traction to this tissues.[1, 2]

There are four major types of inherited EB: EB simplex (EBS), junctional EB (JEB), dystrophic EB (DEB), and Kindler syndrome. These differ not only phenotypically and genotypically but more importantly by the site of ultrastructural disruption or cleavage.[1, 2]

Dystrophic epidermolysis bullosa (DEB) is a rare mechanobullous genodermatosis inherited either with autosomal dominant or recessive pattern and characterized by fragility, blistering and scarring of the skin and mucous membranes. Blistering is due to abnormalities in anchoring fibrils (AF), microstructures mainly composed of type VII collagen (COLLVII), which contributes to the maintaining of dermal-epidermal adhesion.[3]

Most cases are sporadic, but a few show autosomal dominant or autosomal recessive pattern of inheritance. Microscopic studies of EB pruriginosa show typical findings of dystrophic EB, and it has been postulated that itching lesions of EB pruriginosa could represent an abnormal dermal reactivity of some subjects to their inherited bullous disorder.

The study of the molecular basis of dominant dystrophic EB (classical) and EB pruriginosa shows that both diseases are caused by a missense glycine substitution mutation by different amino acids in the same codon of COL 7A (G2028R and G2028A).[4]

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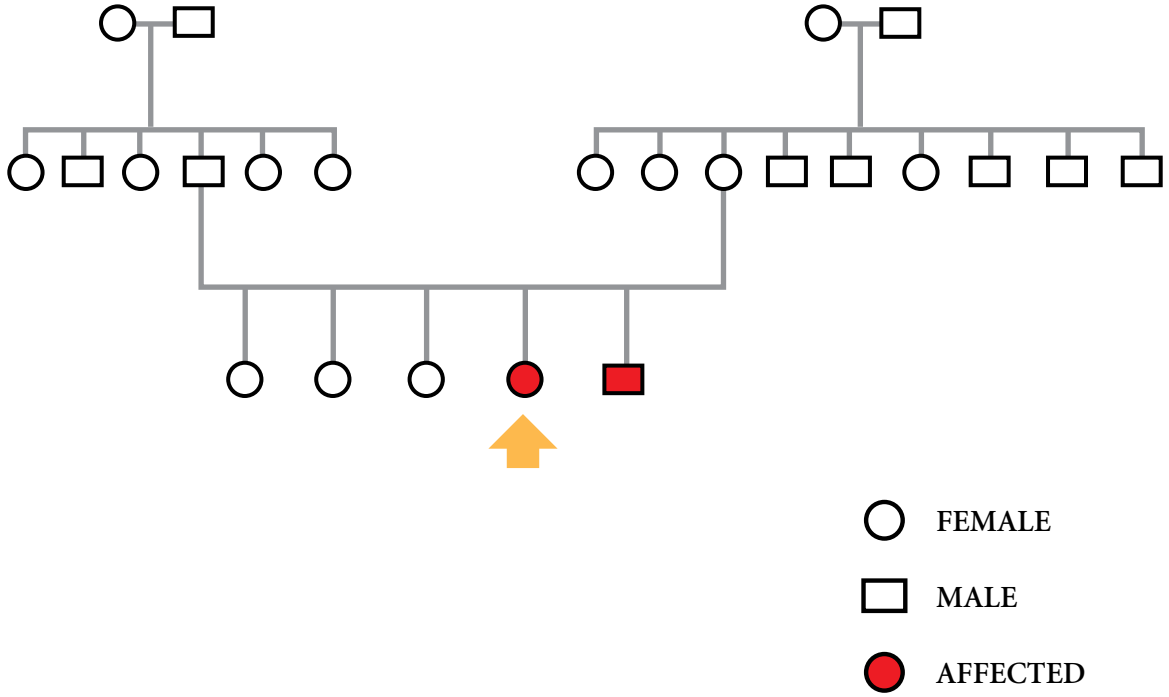

Dystrophic epidermolysis bullosa puriginosa (DEB-Pr) is a distinctive clinical subtype of dystrophic EB. In DEB-Pr patients, autosomal dominant and autosomal recessive inheritance and sporadic inheritance patterns have been recognized. DEB-Pr presents either at birth with mild acral blistering and erosions, or during infancy or childhood. It is clinically characterized by pruritus lichenified plaques or nodular prurigo-like lesions, violaceous linear scarring, occasional trauma induced blistering, excoriations, milia, nail dystrophy and, in some cases, albopapuloid lesions on the trunk. The scarring is most evident on the limbs, particularly on the shins, with relative sparing elsewhere. Intact blisters are rarely seen. The diagnosis of EB in these patients may therefore be difficult, particularly as the condition may only manifest itself some years after birth. Scars frequently have a lichenoid appearance which may cause confusion with non-EB dermatoses, particularly hypertrophic lichen planus, lichen simplex, cutaneous amyloidosis and Nekam's disease.[5]

Case report

A 23-year-old unmarried woman, student, Batak ethnic, referred from dermato-venereologist, came to the Dermato-Venereology Policlinic of Dr. M.D Jamil Hospital Padang complaining of itchy reddish patches with blackish crusts and small blisters containing clear fluid in most part of the body of one year duration.

About 10 years ago, she complained of itchy reddish patches on the folds of the breast and the abdomen followed by the appearance of tiny blisters containing clear fluid that broke easily and leaved black crusts which felt sore. Those reddish patches persisted and never healed completely as whitish papules and plaques.

For the past year, itchy reddish patches gradually appeared on her trunk and lower extremities followed by the appearance of small blisters containing clear liquid that broke easily and leaved blisters and reddish- black crusts. There was no history of reddish patches or blisters triggered by consumption of wheat-made foods (bread, biscuits, noodles or cakes). There was no history of diarrhoea after consumption of wheat-made foods. There was no history of hair loss, brittle or discoloured nails. She lost 8 kg over the past one year. There was no history of herbal medicine and long-term medication. There was no history of having blisters before 10 years of age.

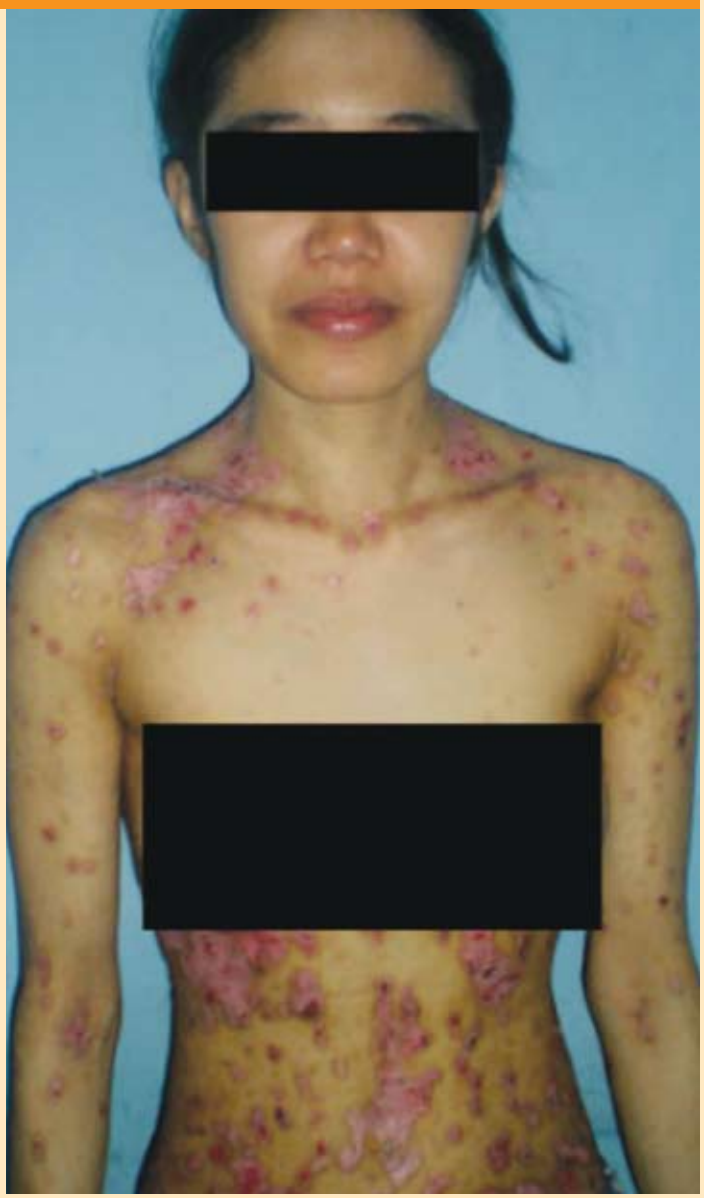

Fig 1 Image of patient.

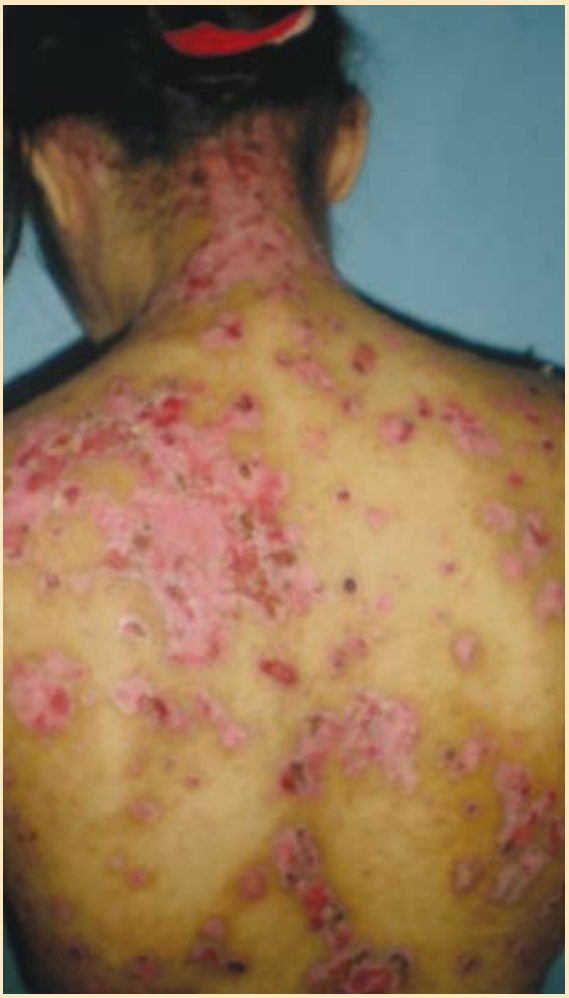

Fig 2 Image of patient.

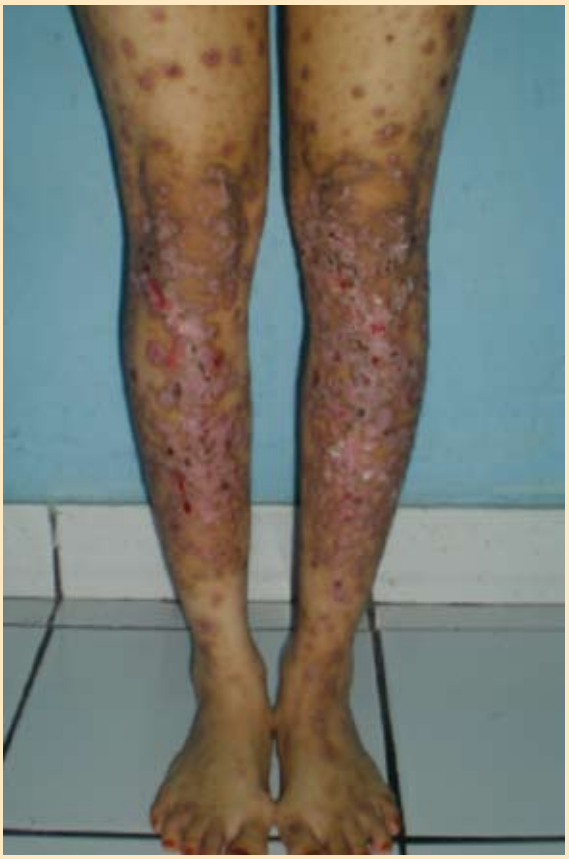

Fig 3 Image of patient.

She is the fourth child in her family. Her younger brother, who is a 19 year-old Batak student, had similar itchy reddish patches with black crusts in his left neck, and both lower limbs for the past year. There was no history of atopy in her family.

Clinically patient was underweight with a BMI of 14.45 and anaemic. Other examinations were in normal limits. There was no hair loss. On extremities there was no oedema.

There were erythematous plaques, black-reddish crusts, vesicles, reddish papules, hyperpigmented plaques, excoriations, hypertrophic scars, atrophic scars, whitish/albupapuloid papules on most of her body. The size of the blisters ranges from pin-point to large plaques. Koebner's phenomenon was positive. Nikolsky sign was negative. Tzanck test was negative. Mucous membranes were normal. Hair and nails were normal. There were no enlargements of regional lymph nodes.

The provisional diagnosis was linear IgA dermatoses with differential diagnosis of dermatitis herpetiformis, epidermolysis bullous acquisita and dystrophic epidermolysis bullous pruriginosa,

From laboratory findings we found anemia and hypereosinophilia. Result of histopathology examination revealed mild acanthosis in epidermis, there were debris with fibrocollagen tissue with perivascular lymphocyte and eosinophils infiltrate noted in dermis. These feature supported diagnosis of non-specific chronic dermatitis. Result of direct immunofluorescence from skin biopsy did not show deposits of immunoglobulins (IgG, IgA, IgM), C3 and fibrinogen and from indirect immunofluorescence antibody against epidermal's component was negative, this result indicate this case was not caused by immunological factor, thus supporting the diagnosis of dystrophic epidermolysis bullosa.

Our provisional diagnosis was dystrophic epidermolysis bullosa pruriginosa (DEB-Pr) with a differential diagnosis of generalized dystrophic epidermolysis bullosa. We suggest the patient to do transmission electron microscope examination, salt split technique and gene mutation analysis examination to find mutation in COL7A1 gene encoding type VII collagen, but these examinations could not be done in Indonesia.

Patient was treated with oral methylprednisolone 20 mg and loratadine 10 mg daily. She was also prescribed with topical desoximetasone 0.25% ointment twice daily for the red patches.

Discussion

Dystrophic epidermolysis bullosa pruriginosa (DEB-Pr) was first described by McGrath et al. in 1994. It is a rare clinical variant of DEB,[3, 7] characterized by marked pruritus, trauma-induced blistering, especially on the extensor aspect of the leg, nail dystrophy, prurigo-like lesions and multiple milia.

DEB is caused by mutations in the COL7A1 gene encoding type VII collagen, resulting in a reduced number or disorganization of anchoring fibrils. In DEB-Pr, mainly glycine substitutions have been reported. The onset of clinical symptoms of DEB-Pr is typically during the first decade or even in infancy; however, in some cases clinical onset may be delayed until later in life.[7]

Pruritus is a well-known accompanying symptom in many epidermolysis bullosa (EB) subtypes but also in a number of other dermatological conditions and systemic diseases such an atopic eczema, chronic renal failure, cholestasis, iron deficiency and thyroid malfunction. The itch may be induced by a specific cause or occur spontaneously. Repeated scratching can eventually lead to a prurigo-like clinical picture, similar to the skin manifestations in DEB-Pr.[8] Our patient complaint of pruritus which accompanies the skin disorders without any history of atopy from patient and family.

The aims of treatment for DEB-Pr are to ease the pruritus and to suppress the scratching activity that leads to the formation of blisters and or prurigo-like lesions; however, no universally successful treatment has been established.

Recent studies have described the efficacy of topical tacrolimus, systemic cyclosporine and thalidomide.[6,10,11] Banky et al. advocate use of topical tacrolimus[6] and Hayashi found that a higher dose of prednisolone (10-30 mg/day) as potentially useful treatment options for DEB-Pr.[7]

Meymandi SS et al. (Iran, 2010) reported a case of epidermolysis bullosa pruriginosa in a 15-year-old Iranian girl who improve after four months therapy with topical clobetasol propionate 0.05% ointment twice daily with oral vitamin E 400 u/daily and oral antihistamine (loratadine 10 mg/daily).[11]

We treated the patient with methylprednisolone 20 mg, loratadine 10 mg, and dexamethasone 0.25% ointment twice daily. We gave the treatment for two weeks which resulted in decrease in pruritus and absence of new lesion.

Reference

1. *Fine JD. Inherited epidermolysis bullosa. Fine Orphanet Journal of Rare Diseases 2010, 5:12*
2. *Marinkovich MP, Bauer EA. Inherited epidermolysis bullosa. In: Freedberg IM, Eisen AZ, Wolff AK, Austen KF, Goldsmith LA, Katz SI, Fitzpatrick TB, editors. Dermatology in general medicine. 7th Edition. New York: Mc.Graw-Hill; 2008:505-15*
3. *Drera B, Castiglia D, Zoppi N, Gardella R, Tadini G, Floriddia G, De Luca N, Pedicelli C, Barlati S, Zambruno G, Colombi M. Dystrophic epidermolysis bullosa pruriginosa in Italy: clinical and molecular characterization. Clin Genet 2006;70: 339–47.*
4. *Das JK, Sengupta S, Gangopadhyay AK. Epidermolysis bullosa pruriginosa – Report of three cases. Indian J DermatolnVenereol Leprol 2005;71:109-11.*
5. *Meymandi SS, Dabiri S, Shafiei H. Epidermolysis Bullosa Puriginosa: report of a case. Iranian Journal of Dermatology 2010;13:20-3.*
6. *Banky JP, Sheridan AT, Storer AL, Marshman G. Successful Treatment of Epidermolysis Bullosa Pruriginosa With Topical Tacrolimus. Arch dermatol 2004;140:794-6*
7. *Hayashi M, Kawaguchi M, Hozumi Y, Nakano H, Sawamura D, Suzuki T. Dystrophic epidermolysis bullosa pruriginosa of elderly onset. Journal of Dermatology 2011;38: 173–8*
8. *Schumann H, Has C, Kohlhase J, Bruckner-Tuderman L. Dystrophic epidermolysis bullosa pruriginosa is not associated with frequent FLG gene mutations. British Journal of Dermatology 2008,159:464–9*
9. *amasaki H, Tada J, Yoshioka T, Arata J. Epidermolysis bullosa pruriginosa successfully controlled by oral cyclosporin. Br J Dermatol 1997;137: 308–10.*
10. *Ozanic Bulic S, Fassihi H, Mellerio JE, McGrath JA, Atherton DJ. Thalidomide in the management of epidermolysis bullosa pruriginosa. Br J Dermatol 2005; 152: 1332–4.*
11. *Meymandi SS, Dabiri S, Shafiei H. Epidermolysis bullosa pruriginosa: report of a case. Iran J Dermatol 2010;13:20-23 .*