CLINICOPATHOLOGIC CHALLENGE

A lady with prickly nodules on both lower limbs

Choon SE, *FRCP*

49 year-old lady referred by nephrologist for increasing number of skin nodules on both legs for past 2 years. Systemic lupus erythematosus with lupus nephritis was diagnosed 10 years ago when she presented with Raynauld's phenomenon, hair loss and nephrotic syndrome. Patient was treated initially with monthly pulses of IV cyclophosphamide and oral prednisolone ranging between 30-10mg. Her renal function is preserved with instituted treatment. On presentation, she was still on prednisolone 7.5mg, mycophenolate mofetil 500mg bd and telmisartan for hypertension which she developed 5 years ago. Physical examination revealed non-tender, woody-hard induration of both legs with underlying firm, sharply angulated papules and nodules (Fig 1).

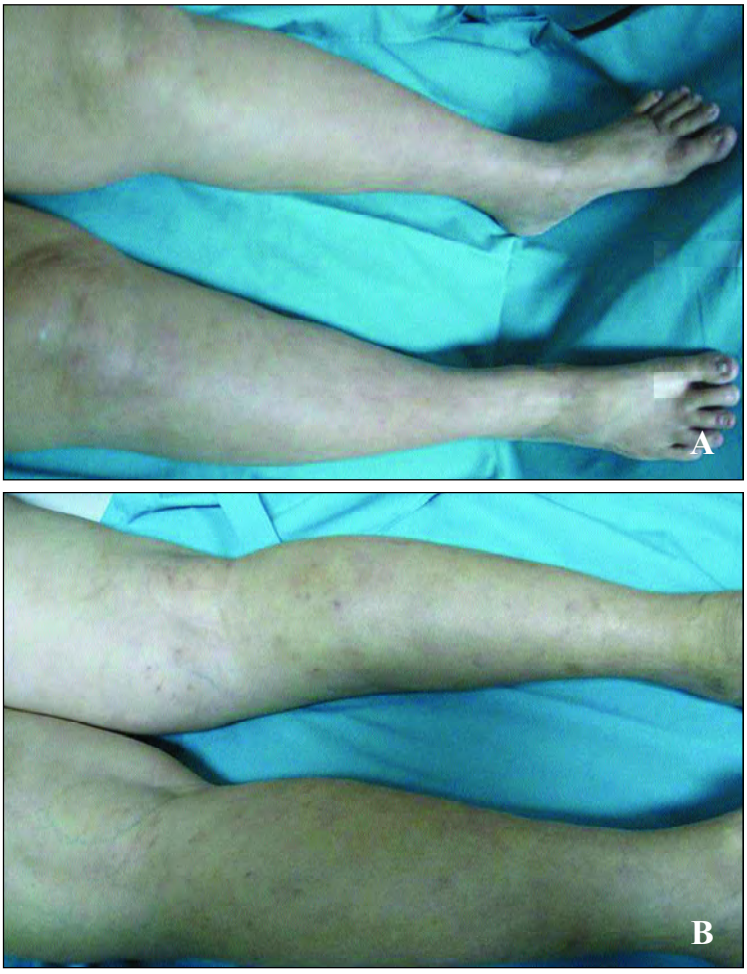

Figure 1 A & B show induration of both legs with smooth shiny, hide-bound skin.

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Histologic features are shown Fig 2.

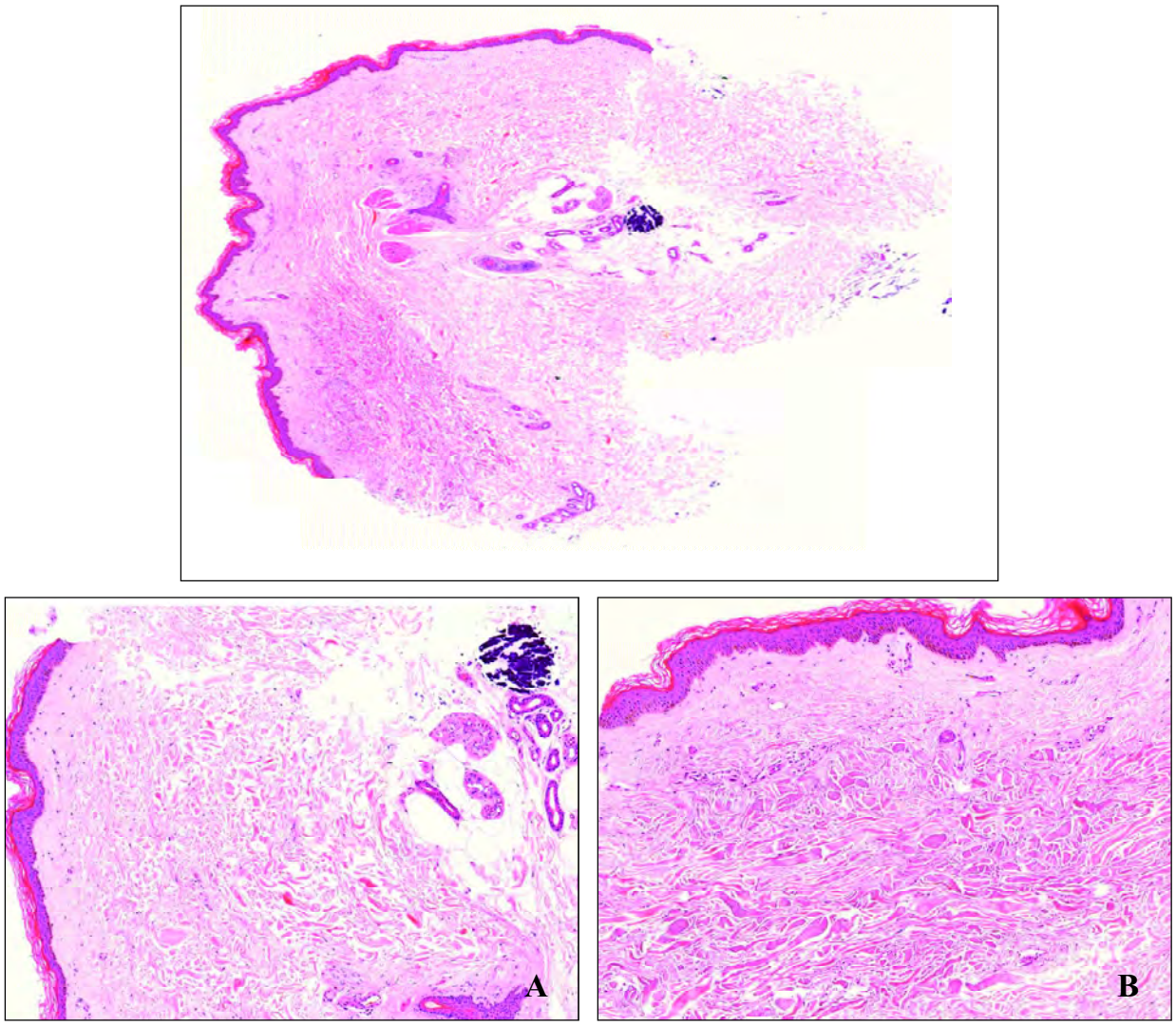

Fig. 2 shows epidermal atrophy with loss of rete pegs, atrophic eccrine glands, haphazardly arranged thickened collagen, scanty inflammatory infiltrates and calcium deposits in dermis and subcutis. 2A shows close-up of calcinosis cutis, 2B shows abnormal thick, homogenous and haphazardly-arranged collagen fibre.

What is your diagnosis?

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Aesthetic Medical Practice Guidelines for Medical Specialists

1. PREREQUISITES FOR MEDICAL SPECIALISTS PERFORMING AESTHETIC MEDICAL PROCEDURES

1.1 A medical practitioner who wishes to perform aesthetic/cosmetic medical procedures must be fully registered with the Malaysian Medical Council.

1.2 He/she must possess a current and valid Annual Practicing Certificate.

1.3 He/she is required to possess a higher qualification in dermatology with full dermatological training; or alternatively, is registered on the National Specialist Register in a medical related field in order to be regarded as medical Specialist.

1.4 He/she must possess experience through recognised practical training courses conducted by bona-fide professional bodies specialising in aesthetic medical practice.

1.5 He/she must exercise strict patient selection criteria, must communicate to the potential client/patient the risks involved, the possible outcome, obtain valid consent for the aesthetic medical procedure planned, and generally observe all aspects of the Code of Professional Conduct of the Malaysian Medical Council.

1.6 He/she must place client/patient safety as the primary concern and should provide aesthetic medical services in a healthcare facility licensed or registered under the Private Healthcare Facilities and Services Act 1998 and regulations 2006.

1.7 He/she is required to obtain a Letter of Credentialing and Privileging (LCP) for the aesthetic/cosmetic medical procedure(s) which he/she intends to perform. The LCP shall be issued by the Cosmetic Dermatology and Laser Medicine (CDLM) Board under the Dermatological Society, Malaysia, Academy of Medicine, Malaysia.

1.8 With the LCP, he/she is eligible for registration with the Registry of Aesthetic Medical Practice which shall be maintained by the Medical Practice Division, Ministry of Health (MOH), Malaysia.

2. SCOPE OF PRACTICE

The basic considerations for the scope of practice in aesthetic medical practice by medical specialists are whether they are core medical specialists or non-core medical specialists (refer Table 1).

A. Core Medical Specialists

This consists of dermatologists performing aesthetic/cosmetic surgery within their core curriculum and core competency.

The core specialist society will submit a list of their specialists to the CDLM Board for inclusion in the National Registry of Aesthetic Medical Practice.

Specialists may also apply directly to the CDLM Board.

B. Non-Core Medical Specialists

This refers to medical specialists whose routine areas of practice are completely unrelated to dermatology e.g. anaesthetists, pathologists, radiologists etc.

These specialists may be subjected to similar requirements for privileging of a general practitioner practising aesthetic medical practice.

If possible they should be sanctioned by their own professional peers before application to the CDLM Board; alternatively they may apply directly to the Board with the necessary documentation.

Table 1 Scope of practice for Medical Specialists.

PROCEDURES	CORE SPECIALISTS	NON-CORE SPECIALISTS	APPROPRIATE PREMISES NEEDED	PROCEDURES PERFORMED FOR NON-CORE SPECIALISTS
A. NON INVASIVE				
Chemical peels (superficial)	Dermatologists	Case by case basis	Clinic	25
Microdermabrasion	Dermatologists	Case by case basis	Clinic	20
Intense pulse light (IPL)	Dermatologists	Case by case basis	Clinic	40
B. MINIMALLY INVASIVE				
Chemical peel (medium)	Dermatologists	Case by case basis	Clinic	25
Botulinum toxin injection	Dermatologists	Case by case basis	Clinic	25
Filler injection	Dermatologists	Case by case basis	Clinic	25
Sclerotherapy	Dermatologists	Case by case basis	OT/ Clinic	20
Laser for treating skin pigmentation	Dermatologists	Case by case basis	OT/ Clinic	20
Laser for treating skin tumours	Dermatologists	Case by case basis	OT/ Clinic	20
Laser for skin rejuvenation (incl fractional)	Dermatologists	Case by case basis	Clinic	20
Laser for hair removal (e.g long-pulsed Nd-YAG, Diode)	Dermatologists	Case by case basis	Clinic	20
C. INVASIVE				
Laser for treating vascular lesions	Dermatologists	NA**	OT/ Clinic	
Chemical peels (Deep)	Dermatologists	NA	OT/Clinic	
Ablative skin resurfacing lasers	Dermatologists	NA	OT/ Clinic	
Hair transplant	Dermatologists	NA	OT	
Mechanical dermabrasion	Dermatologists	NA	OT/ Clinic	

PROCEDURES	CORE SPECIALISTS	NON-CORE SPECIALISTS	APPROPRIATE PREMISES NEEDED	PROCEDURES PERFORMED FOR NON-CORE SPECIALISTS
Phlebectomy	Dermatologists	NA	OT/Clinic	
Photodynamic therapy	Dermatologists	NA	OT/ Clinic	
Radiofrequency	Dermatologists	NA	OT/ Clinic	
Ultrasound device	Dermatologists	NA	OT/Clinic	
Tumescent liposuction	Dermatologists	NA	OT/ Clinic	

Note:
This list is subjected to be reviewed whenever there is new evidence-based treatment available.
**OT = Operation theatre, **NA = Not applicable*

3. PROCESS OF REGISTRATION

3.1 Medical specialists who intend to practise aesthetic medical practice are required to apply to the Cosmetic Dermatology and Laser Medicine (CDLM) Board of the Dermatological Society Malaysia.

3.2 The CDLM Board shall assess and/or examine medical specialists who intend to practice aesthetic/cosmetic procedures.

3.3 The CDLM Board will issue a Letter of Credentialing and Privileging (LCP) to the successful candidate, specifying the core specialty, aesthetic/cosmetic procedure(s) approved, and duration of validity. The LCP is valid for 5 year and renewable upon endorsement by the CDLM Board.

3.4 With the LCP, the medical specialists' names are eligible to be included in the National Registry of Aesthetic Medical Practice.

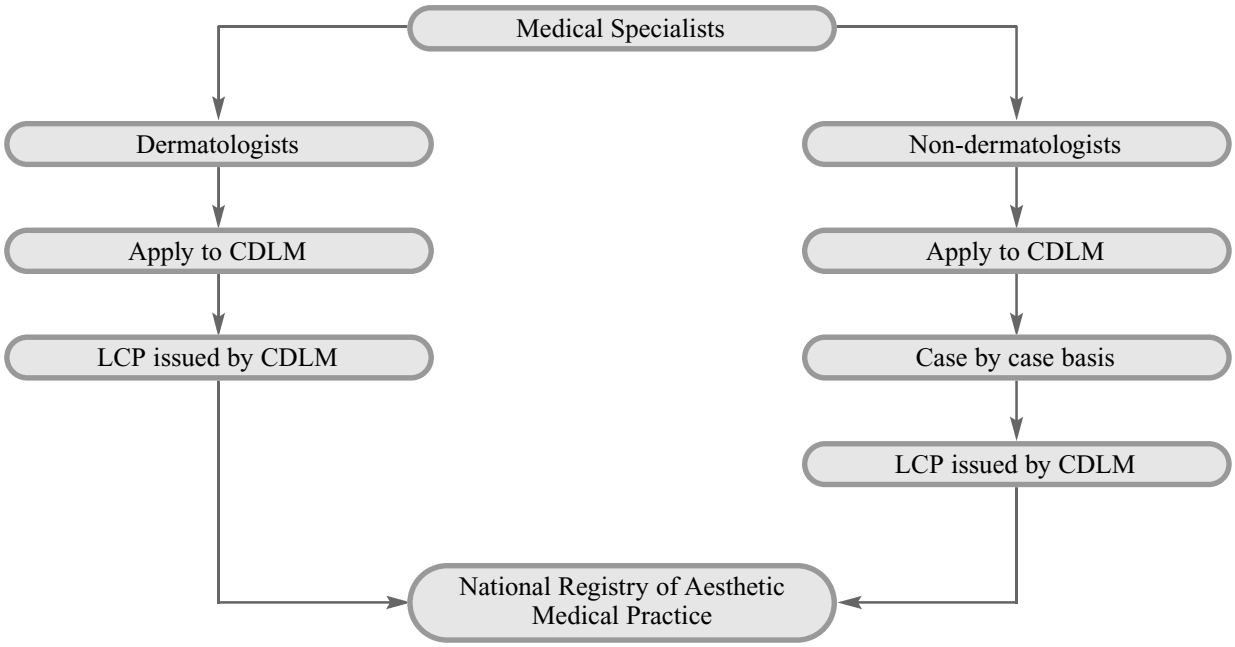

Figure 1 Process of registration for medical specialists.

4. THE CREDENTIALING AND PRIVILEGING COMMITTEE

The Cosmetic Dermatology and Laser Medicine (CDLM) Board of the Dermatological Society Malaysia will be the credentialing and privileging committee for the medical specialists.

5. THE SECRETARIAT FOR MEDICAL SPECIALISTS

Persatuan Dermatologi Malaysia (PDM)
Rumah Dermatologi
2-16-2, Block 2 (Remis) Pantai Panorama Condominium
Jalan 112 H, off Jalan Kerinchi
59200 Kuala Lumpur

BOOK REVIEW

Textbook of Laser and Light Dermatology in the Asian Skin

Edited by Yong-Kwang Tay and Yuin Chew Chan

Published by World Scientific Publishing Co Pte 2011. 140 pp.
ISBN-13:978-9814338868

It was a pleasure to review this well illustrated and informative textbook of laser and light dermatology. This is the latest textbook of laser and light dermatology for Asian skin just released in 2011. It is written and edited by Tay Yong Kwang and Chan Yuin Chew along with a further 10 other contributors. The book has 12 chapters and its contributors are all experienced dermatologists including Prof Goh Chee Leok, Joyce Lim, Chua SH and Melvin Ee from Singapore who gave practical pearls of wisdom and tips treating darker skin phenotypes.

The book is divided into 12 chapters covering a comprehensive range of laser and light based devices. It's only 140 pages, full of color images and is very readable. One of the outstanding features of the text is that it has numerous actual case presentations and the author would include the laser settings to be used in the treatment. Each chapter begins with an introduction giving the reader a general overview of the topic followed by mechanism of action, patient selection and treatment procedure including the laser parameters, precautions and post operative care. In some chapters the chapter would end with clinical case histories. The book include chapters on laser tissue interactions, CO2 laser, vascular, pigment, hair removal, ablative resurfacing, non ablative resurfacing, fractional laser, IPL and photodynamic therapy.

Of special interests are chapters on treatment of pigmented lesions, fractionated lasers, intense pulsed light devices and photodynamic therapy in the Asian skin. Conditions unique to the Asian skin such as Nevus of Ota, Hori's nevus, melasma, and post inflammatory hyperpigmentation were well covered in the text.

Currently there are very few reference textbook of laser and light devices on Asian skin. The *Textbook of Laser and Light Dermatology in the Asian Skin* will serve as a valuable reference textbook and an essential companion for all dermatologists and medical practitioners managing patients of Asian descent with laser and light based devices. The clinical information and the high-quality photographs will make this color textbook an enduring addition to any personal or institutional medical library.

Henry Foong Boon Bee FRCP (Edin)
Ipoh, Malaysia

TRUE GRIT

DR. SORYA A. AZIZ (1958-2011)

Dr. Sorya A. Aziz hailed from Johor Bahru where she was the eldest in her family. She completed her early school years in Johor and completed her MCE at Sekolah Seri Puteri, Kuala Lumpur. She found her calling in Medicine and went on to do her MD at the Universiti Kebangsaan Malaysia (UKM) and subsequently her Masters in Internal Medicine at the Universiti Sains Malaysia (USM). She served as a physician at Hospital Melaka for a few years before she pursued her dream to train in Dermatology. She joined the Department of Dermatology, Hospital Kuala Lumpur in 1998 and completed her fellowship training in 2001.

She developed an interest in Infectious Dermatology and went on to subspecialise in this field at the University of Amsterdam in 2003. She was involved in research projects involving Multiplex PCR in non-tuberculous mycobacterium. She returned to Malaysia in 2004 and headed the Infectious Disease Specialised Service in the Department of Dermatology, Hospital Kuala Lumpur.

Unfortunately, her health declined gradually and she was diagnosed with end stage renal failure in 2006 and commenced on renal replacement therapy. Despite all this, Sorya remained in high spirits and continued to work full-time as a Dermatologist which was commendable. She was transferred as the Head, Department of Dermatology, Hospital Sultan Ismail, Johor Bahru in 2008 where she served until her untimely demise in 2011.

Sorya will be remembered by all who knew her as exemplary and dedicated Dermatologist with immense patience and courage. More importantly, she was a true friend and great listener who was with us through thick and thin. In her unassuming ways, she was an astute diagnostician in our department. She was a mentor not only to her juniors but also to her colleagues.

Sorya will also be remembered as a fun-loving person who was in high spirits until the end. She remained optimistic despite all the hurdles that came her way. We are sure that Sorya would like us to celebrate her life as she lived it.

To a great Dermatologist, a superb teacher, a true friend and a wonderful person.

We miss you….
Al- fatihah
Semoga Allah mencucuri rahmat ke atas rohnya
"May her soul rest in eternal peace"

Asmah Johar, Suganthi Thevarajah, Dawn Ambrose, Noor Zalmy Azizan

ANSWER TO CLINICOPATHOLOGIC CHALLENGE

49 year-old lady referred by nephrologist for increasing number of skin nodules on both legs for past 2 years. Systemic lupus erythematosus with lupus nephritis was diagnosed 10 years ago when she presented with Raynauld's phenomenon, hair loss and nephrotic syndrome. Patient was treated initially with monthly pulses of IV cyclophosphamide and oral prednisolone ranging between 30-10mg. Her renal function is preserved with instituted treatment. On presentation, she was still on prednisolone 7.5mg, mycophenolate mofetil 500mg bd and telmisartan for hypertension which she developed 5 years ago. On further questioning, patient did notice progressive hardening of both legs for 2 years but attributed it to past cellulitis in 2008. Hence, she only consulted her nephrologist when prickly lesions on thighs caused discomfort. She has no associated ulceration or discharge. She still has Raynauld's phenomenon but did not have difficulty in swallowing, thickening of facial skin or skin on her upper limbs. Fig 3 Biopsy of a lesion on her right calf showed atrophy of skin with loss of rete pegs, thickened collagen in haphazard arrangement and subcutaneous calcinosis

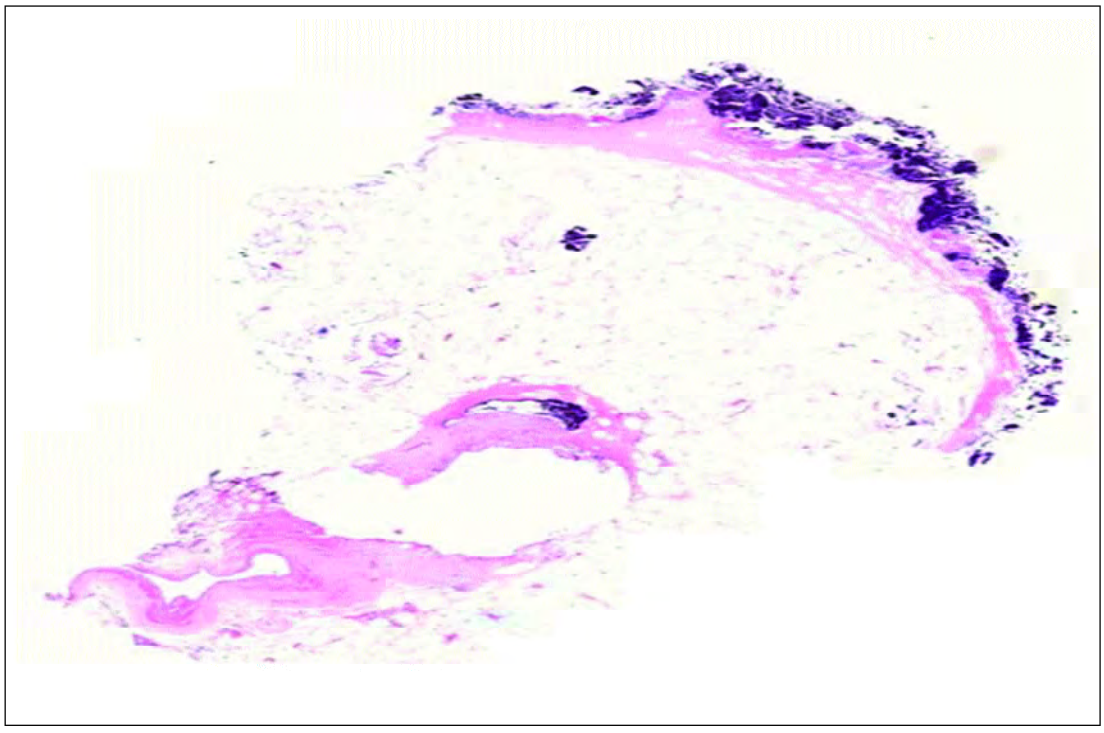

Figure 3 shows calcinosis in the subcutaneous tissue.

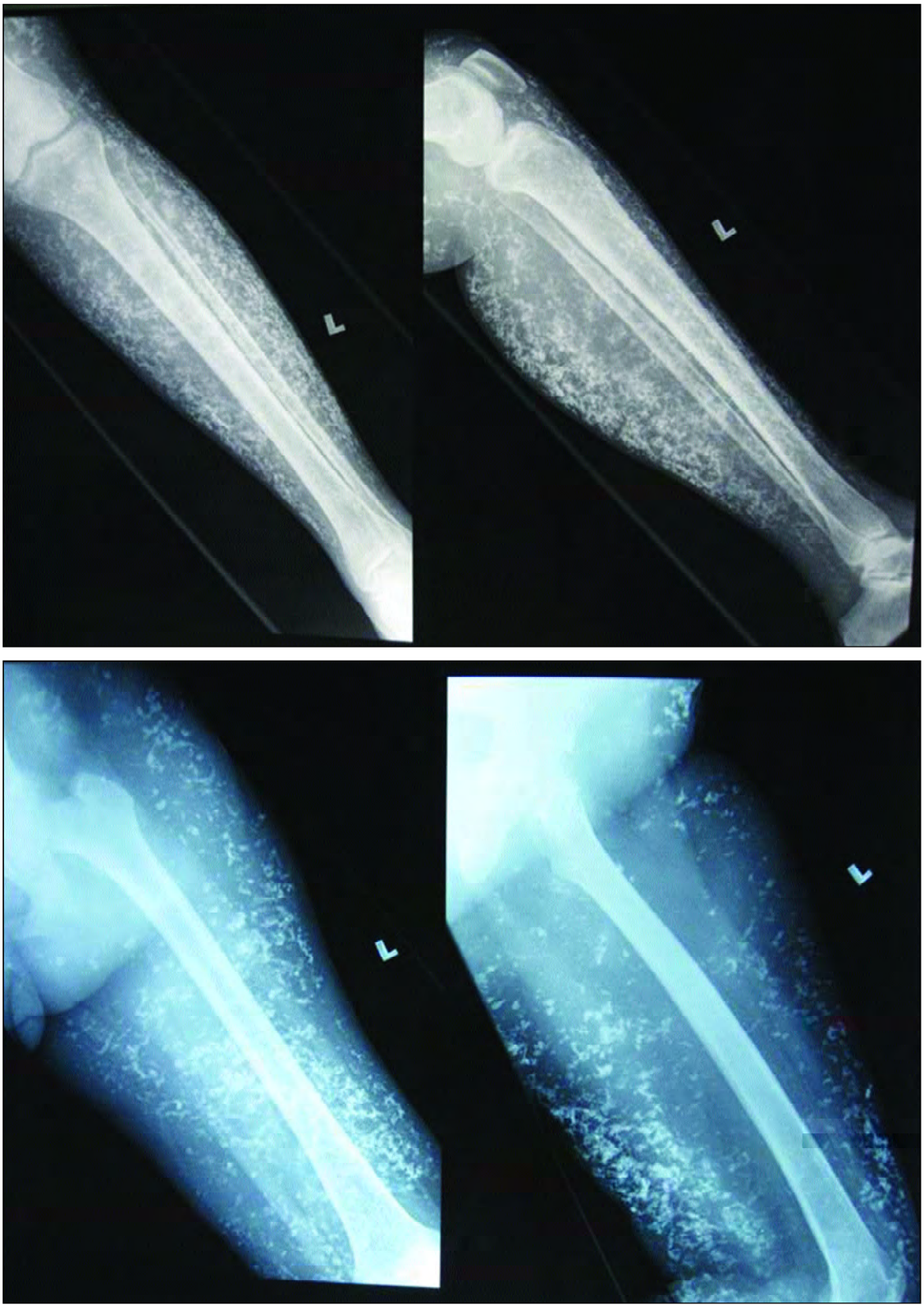

Figure 4 A & B show diffuse fluffy calcinosis affecting soft tissues of both lower limbs.

The detached subcutaneous tissue (Fig. 4) also showed obvious calcification. Hence, patient's chief complaint was due to calcinosis cutis. Calcification of the skin and subcutaneous tissue is known to occur in a variety of disorders and may be classified as dystrophic, metastatic, idiopathic or iatrogenic calcification, and calciphylaxis[1]. Dystrophic calcification appears as a result of local tissue damage in patients with normal serum calcium and phosphate levels. Metastatic calcification is characterized by an abnormal calcium and/or phosphate metabolism, leading to the precipitation of calcium in cutaneous and subcutaneous tissues. Idiopathic calcification occurs without any underlying tissue damage or metabolic disorder. Iatrogenic calcinosis cutis is a side effect of therapy reported after IV calcium gluconate infusion. Calciphylaxis is defined as small-vessel calcification mainly affecting blood vessels of the dermis or subcutaneous fat. There may be disturbed calcium and phosphate metabolism and hyperparathyroidism. This potentially fatal syndrome predominantly occurs in patients with end-stage renal disease.

Blood investigations including full blood count, renal function tests, muscle enzymes, serum calcium and phosphate were unremarkable. Hence, patient has dystrophic calcinosis cutis. Widespread fluffy opacities affecting both legs and thighs were seen on radiography (Fig.5). Dystrophic calcinosis cutis is the most common type of cutaneous calcification. The ectopic calcified mass typically consists of hydroxyapatite and amorphous calciumphosphate[1]. The pathophysiology of the disorder is still unclear although it has been suggested that phosphate-bound denatured proteins of necrotic cells serve as a nidus for ectopic calcification, and that alterations in collagen, elastin and subcutaneous fat promote the calcification process. Histopathologically, calcium deposits stain dark blue with hematoxyline and eosin stain and black with von Kossa stain. Fine granules of calcium are usually seen in the dermis while large, irregular calcium masses occur in the subcutaneous tissue. A foreign body reaction with inflammation and fibrosis may be seen around larger calcified deposits.

Dystrophic calcification is associated with a variety of disorders, including connective tissue diseases, inherited disorders, cutaneous neoplasms and infections. Among the connective tissue dieases, calcinosis is most commonly seen in juvenile dermatomyosistis. Calcinosis cutis in systemic lupus erythematosus (SLE) is rare and usually asymptomatic. Calcinosis cutis is a common finding in systemic sclerosis (SS), especially in the limited cutaneous form of systemic sclerosis (CREST syndrome)[2,3]. Subcutaneous or intracutaneous calcification occurs in 25% to 40% of patients with limited systemic sclerosis (LcSS), typically 10 years or more after disease onset. Clinically, nodules and plaques of calcium deposits occur at sites of recurrent microtrauma such as the forearms, elbows, or fingers. Ulceration of the overlying skin and discharge of chalky material may occur. Diffuse and tumoral calcinosis is rare but reported in localised systemic sclerosis.

So does patient has SS, SLE or ovelap syndrome with calcinosis cutis? Closer examination of patient's biopsy showed atrophy of epidermal layer with loss of rete pegs and atrophic eccrine glands (Fig.2), haphazardly arranged thickened collagen and scanty inflammatory infiltrates, supporting a diagnosis of systemic sclerosis. SLE has much more inflammatory infiltrates with immunoglobulin deposition at the dermoepidermal junction and within small blood vessels whereas direct immunoflourescence studies are usually negative in SS. Indirect immunofloresecence test showed positive ANA with titre of 1:640 and a homogenously distributed speckled pattern in the nucleus which is characteristic of anti-centromere antibody[4,5]. Enzyme-linked immunoassay was negative for double-stranded DNA, anti-Smith, anti-RNP, anti SSA, anti-SSB, antiJo1 and anti-topoisomerase (anti-Scl 70). Anti-topoisomerase and anti-centromere antibodies are the classic ANA found in systemic sclerosis[4,5]. They are intrinsically specific for SSC and rarely found in healthy individual or patients with other connective tissue disease. Anti-topoisomerase antibody is associated with diffuse SS and predicts early incidence of interstitial lung disease, renal and heart involvement[4,5]. Anti-centromere antibody in SS patient is associated with limited cutaneous

disease, peripheral vascular damage, calcinosis and later onset of pulmonary hypertension. When anticentromere antibody is found in patients with Raynauld's phenomenon, it predicts future development of localised cutaneous systemic sclerosis. Final diagnosis is limited cutaneous systemic sclerosis with dystrophic calcification.

There is no recommended standard treatment for calcinosis cutis[6,7]. Success had been reported with use of warfarin, diltiazem, aluminium hydroxide, minocycline, cefuroxime, laser therapy and excision[6,7].

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