ORIGINAL ARTICLE

A cluster of chilblains in Hong Kong

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Objective To report a recent clustering of chilblain cases in Hong Kong.

Design Case series.

Setting A regional hospital and a social hygiene clinic in the New Territories West, Hong Kong.

Patients Patients with a clinical diagnosis of chilblains in February 2008.

Results Eleven patients with chilblains were identified; seven (64%) gave an antecedent history of prolonged exposure to cold. They all presented with erythematous or dusky erythematous skin lesions affecting the distal extremities, especially fingers and toes. Laboratory tests revealed elevated antinuclear antibodies titres in two, positive rheumatoid factor in two, presence of cold agglutinins in one, and a raised anti-DNA titre (>300 IU/mL) in one. Skin biopsies were performed in six patients, four of them showed typical histopathological features of chilblains. In the patient with systemic lupus erythematosus, features of vasculitis were suspected, and in the one with pre-existing juvenile rheumatoid arthritis, there were features of livedo vasculitis. In 10 (91%) of the patients, the skin lesions had resolved when they were last assessed (at the end of March 2008), but had persisted in the patient who had pre-existing systemic lupus erythematosus.

Conclusion The recent clustering of chilblains was possibly related temporally to the prolonged cold weather at the end of January to mid-February. In our series, most of the patients developed chilblains as an isolated condition and resolved spontaneously within a few weeks. Laboratory tests and skin biopsies for chilblains are not necessary, unless the condition persists, the diagnosis in doubt or an underlying systemic disease is suspected.

Key words
Chilblains; Cold/adverse effects; Fingers; Hypothermia; Toes

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Introduction

Chilblains, also known as pernio, is an inflammatory skin condition caused by prolonged exposure to cold temperatures above freezing, especially when the weather is damp.[1] Clinically, the affected area is characterised by erythematous, oedematous macules, papules, and plaques. Vesicles and ulceration may occur in severe cases. Chilblains often affect distal extremities but ears, nose, face, and thighs may also be involved. The condition is well documented in cold and temperate regions,[2,3] often occurs during winter, but is uncommonly encountered in local hospitals or dermatology clinics. Locally, the Chinese lay term "生蘿蔔" is commonly used to describe the condition. We report the clinicopathological features of a cluster of chilblains diagnosed in February 2008.

Methods

All patients diagnosed as having chilblains by dermatologists in Tuen Mun Social Hygiene Clinic or Tuen Mun Hospital in February 2008 were recruited. Their demographic and clinical features were recorded. History regarding occupation, precipitating factors, current and past medical history including drug intake were also documented. For all patients, blood tests for autoimmune markers were performed. Skin biopsies were performed if appropriate.

Results

A total of 11 patients with chilblains were identified, six were male; seven (64%) were in-

香港的凍瘡群體個案

目的 報告近期香港的凍瘡群體個案。

設計 臨床個案研究。

安排 香港新界西的區域醫院與社會衛生科診所。

病人 於2008年2月期間臨床診斷為凍瘡的病人。

結果 該期間共有11名臨床診斷為凍瘡的病人，其中7名(64%)事前有受凍的病史，而全部病人均於四肢末端出現紅或暗紅的皮疹，尤其是手指和腳趾較為明顯。化驗報告中，有2名病人的抗核抗體呈陽性，2名的類風濕性關節炎因子呈陽性，1名的冷凝集素呈陽性，1名的抗雙旋DNA抗體(>300 IU/mL)呈陽性。共有6名病人接受皮膚活體組織檢查，當中4名有凍瘡的典型要素。1名已知患有系統性紅斑狼瘡的組織檢查有疑似血管炎，而另1名患有幼年型類風濕關節炎的組織檢查則有網狀青斑血管炎的特徵。於3月底的評估中，10名(91%)病人的紅疹已消退，只有該名患有系統性紅斑狼瘡的病人紅疹仍未消退。

結論 近期發現的凍瘡個案與一至二月中的長期寒冷天氣有關。在我們的凍瘡個案當中，多為孤單性的，而皮疹並會於數星期內消退。除了過長的病史，診斷不確實或懷疑有潛在的系統性疾病，大多數情況下不需實驗室或皮膚活體組織檢查。

patients and four (36%) were seen in the out-patient clinic. Their mean age was 36 (range, 11-71) years and all of them were local Hong Kong Chinese. Five (45%) were aged 15 years or younger; the ages of the other six ranged from 43 to 71 years. The demographic features of these patients are shown in Table 1 and their clinicopathological features in Table 2.

Except for patient 3 who reported similar but less severe attacks during past winters, this was their first attack. All of them were seen within 5 days to 4 weeks from symptom onset, nine (82%) of whom presented to a dermatologist within 2 weeks. Patients 2 and 7 had been previously diagnosed to have juvenile rheumatoid arthritis and systemic lupus erythematosus (SLE), respectively. Only the latter patient was in receipt of systemic medications (prednisolone, mycophenolate mofetil, and lisinopril). The other nine (82%) patients with no systemic disease, enjoyed good past health and were not on regular medications. The diagnoses prior to skin consultations were vasculitis in four (36%), cellulitis in three (27%), and dermatitis in one (9%). There was no initial diagnosis in three (27%) patients, until seen for a dermatologist's opinion. Prolonged exposure to cold was a relevant part of the history in seven (64%) of the patients. Such exposure included: cold weather, contacting with cold water, and playing 'TV' games with ungloved/unprotected hands exposed to cold. For the other four patients, cold exposure was not a definite precipitating factor. In our patients, protection from cold (woven gloves and socks, and heaters) had not been adopted. None of the patients gave a history suggestive of Raynaud phenomenon.

The main cutaneous features were painful, itchy, and dusky erythematous macules or purpura located on the distal extremities (Fig 1). In patients 2, 4, 5, and 11, the fingers and the toes were affected. Five (45%) patients had digital swelling, and painful erosion or ulceration were noted in two (18%). In our series, other chilblain susceptible areas (ears, nose, and thighs) were not affected.

Skin biopsies were performed in six (55%) of the

TABLE 1. Patient demographics and other relevant history

Patient No.	Sex/age (years)	Occupation	Relevant medical history	Implicated precipitating factor
1*	F/63	Housewife	No	Contacting detergent during housework
2	M/14	Student	Juvenile rheumatoid arthritis	Not known
3	M/45	Fishmonger	Recurrent episodes in winter time for few years	Contacting cold water
4	M/15	Student	Nil	Cold weather
5	M/14	Student	Nil	Not known
6	F/52	Housewife	Parkinson's disease	Cold weather
7	F/43	Housewife	Systemic lupus erythematosus on mycophenolate mofetil, prednisolone, and lisinopril	Cold weather
8	F/11	Student	Nil	Not known
9*	F/53	Gardener in football court	Recurrent episodes in winter	Contacting cold water
10*	M/71	Retired	Nil	Prolonged cold exposure in China
11*	M/13	Student	Nil	Prolonged cold exposure while playing TV games

* Seen as out-patient

† Not applicable or diagnosis not made to patient

patients, but were not undertaken in the remainder (whose lesions were resolving). In patients 3, 4, 5 and 6, the histopathological features entailed dermal perivascular lymphocytic infiltrates compatible with chilblains (Fig 2). Additional features included fibrin thrombi, red cell extravasation, epidermal necrosis, and re-epithelialisation. Patient 2 had livedo vasculitis, and vasculitis was suspected in patient 7 (with SLE). Blood tests for autoimmune markers were performed in all 11 patients, and no abnormality was detected in seven (64%). Patients 8 and 9 had raised antinuclear antibodies (ANA) titres; patient 7 had a positive anti-DNA antibody, and patients 2 and 9 were rheumatoid factor–positive. In patient 4, cold agglutinins were demonstrated but other immunological test results were not abnormal.

Advice on keeping extremities and chilblains prone areas warm was given to these patients. Topical steroids were prescribed in nine (82%), and systemic antibiotics to three (27%) who were initially diagnosed as having cellulitis. Patient 2 received nifedipine after skin consultation for his significantly painful lesions. Further enquiry about the progress of skin lesions (by the end of March) within the next 6 weeks, revealed that the skin lesions had resolved, except in patient 7.

Discussion

Hong Kong's climate is subtropical, tending towards temperate for nearly half the year.[4] Whilst days with cold spells are usually brief, early this year Hong Kong experienced its longest cold spell in the past 40 years. Cold weather warnings were announced and spanned from 24 January to 18 February 2008 (more than 24 days); about 595 hours were deemed cold.[5] At the Hong Kong Observatory, the mean minimum temperature during the cold spell (Fig 3) was 10.8°C, the lowest being 7.9°C. On nine (35%) of these days, the relative humidity was high (≥85%; Fig 4). All our patients lived in the west region of New Territories. Because of the lack of comparative data, it is not certain if this cluster of cases was the largest in Hong Kong, but we are not aware of other similar reports affecting the local population. However, chilblains can be very mild and escape medical attention. Detailed history revealed that our patients dated onset of their symptoms from the end of January to mid February, corresponding to the period of cold weather warnings by the Hong Kong Observatory. Young and elderly patients have a predilection to chilblains, which were also evident in our series, five of whom were aged 15 years or less and the others were middle-aged or elderly.

Diagnosis of chilblains is not difficult. Common features include a history of exposure to cold, usually for recreational or occupational purposes; presence of painful or itchy erythematous skin lesions affecting distal extremities, especially the digits; and a self-limiting course upon cessation of cold challenge. Differentiation of chilblains from other clinical mimickers (eg leukocytoclastic vasculitis, cellulitis, and contact dermatitis) is important. Leukocytoclastic vasculitis can be preceded by respiratory tract infections or intake of drugs (eg antibiotics or non-steroidal anti-inflammatory agents). It typically presents with painful, palpable, purpuric lesions on lower limbs, especially the legs. Venulitis with fibrinoid necrosis, perivascular neutrophilic infiltrates, and leukocytoclasia are the classical histopathological

Date for skin consultation (estimated date of onset)	Duration before presentation	Diagnosis before skin consultation
13 Feb (3 Feb)	10 days	Nil†
16 Feb (2 Feb)	2 weeks	Vasculitis
13 Feb (3 Feb)	10 days	Cellulitis
16 Feb (2 Feb)	2 weeks	Vasculitis
20 Feb (10 Feb)	10 days	Vasculitis
26 Feb (5 Feb)	3 weeks	Cellulitis
17 Feb (3 Feb)	2 weeks	Vasculitis, lupus erythematosus
11 Feb (6 Feb)	5 days	Cellulitis
13 Feb (31 Jan)	2 weeks	Nil
28 Feb (1 Feb)	4 weeks	Nil
28 Feb (30 Jan)	4 weeks	Hand and foot dermatitis

TABLE 2. Patient clinicopathological features

Patient No.	Symptom/sign	Location of lesion	Biopsy
1*	Dusky rash, swelling	Hands and fingers	ND†
2	Painful dusky rash	Fingers and toes	Livedoid vasculitis
3	Itchy, swelling dusky rash, erosion	Hands and fingers	Chilblains
4	Dusky rash	Fingers, feet, and toes	Chilblains
5	Painful dusky rash, swelling	Fingers, feet, and toes	Chilblains
6	Pain and itchy purpuric rash	Feet and toes	Chilblains
7	Dusky rash and ulcer	Feet and toes	Suspicious of vasculitis
8	Dusky rash, swelling	Feet and toes	ND
9*	Pain, itchy dusky rash, swelling	Fingers	ND
10*	Itchy, dusky rash	Fingers	ND
11*	Dusky rash	Fingers and toes	ND

* Seen as out-patient
† Not done
‡ ANA denotes antinuclear antibody

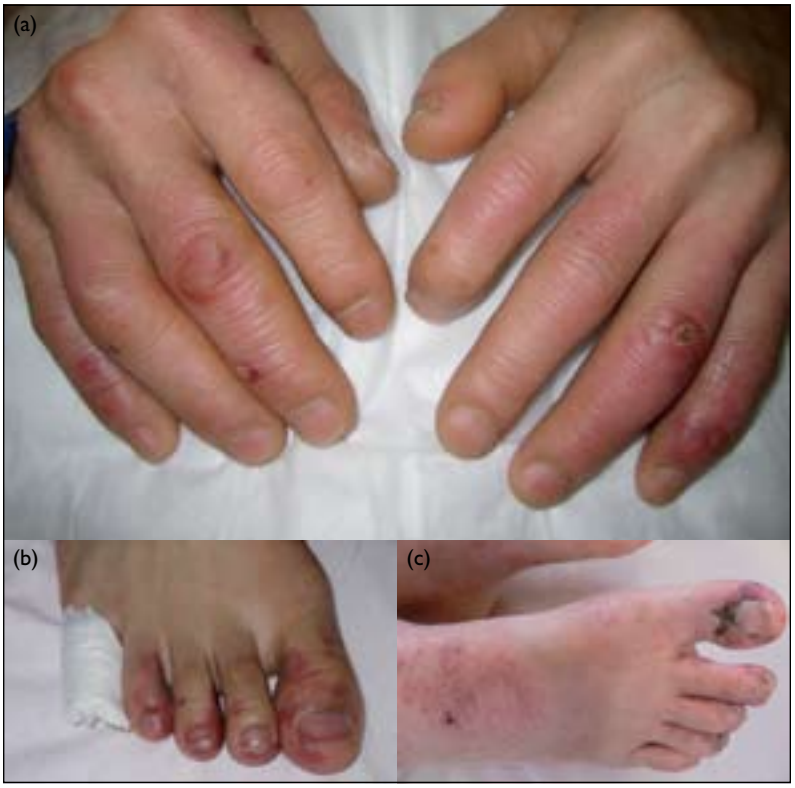

FIG 1. (a) Dusky erythematous patches with erosions and oedema on the dorsal aspect of fingers in patient 3. (b) Dusky red oedematous purpuric macules with central greyish discolouration, affecting the toes in patient 2. (c) Dusky erythematous lesions on the right foot with ulceration at the base of the big toe in patient 7

features, while lymphocytic infiltrate is scant in early lesions, and a strong relationship with cold exposure is not recognised. Cellulitis is an infection of skin involving dermis and subcutaneous tissue. Tissues or blood cultures may grow pathogenic cocci and prompt systemic antibiotic therapy is indicated. Contact dermatitis is not cold related. Housewife dermatitis is a form of irritant contact dermatitis with features that include: dry, rough, and chapped skin, with a background of erythema mostly affecting the hands. The toes are not usually affected. Dusky erythema, oedema, and ulceration are not typical of housewife dermatitis. While patient 1 attributed her onset of symptoms to contact with detergents during housework, it was more likely that her chilblains were precipitated by contact with cold water.

While the diagnosis of chilblains is mostly based on clinical grounds, a skin biopsy may assist differentiation from mimicks. The basic histological pattern in chilblains is a lymphocytic vasculopathy. The lymphocytic infiltrate is generally dense and involves the superficial and deep dermal vasculatures.[2] The vasculopathic changes take the form of fibrinoid lesions in vessel walls and/or intraluminal thrombi that occur to a variable extent. Leukocytoclasia (a breaking down of leukocytes forming nuclear dust) is not a feature. In some lesions, a lymphocytic eccrine hidradenitis or lymphocytic infiltrate of the eccrine acrosyrinx is present.[2] This pattern overlaps with lupus dermatitis, and other epidermal and stromal changes are necessary for differentiation. The often-stated prominent papillary dermal oedema was not seen in our biopsies. In all of our biopsies, there were central epidermal changes in form of epidermal necrosis or erosions. Interface vacuolar change was not a prominent feature in all.

Laboratory finding‡	Treatment(s)	Outcome (assessed in March 2008 from disease onset)
Negative	Topical steroid	Resolved in 6 weeks
Rheumatoid factor, 17.3 IU/mL	Topical steroid, nifedipine	Resolved in 5 weeks
Negative	Antibiotics (commenced before skin consultation), topical steroid	Resolved in 3.5 weeks
Cold agglutinin titre, 1:128	Nil	Resolved in 4 weeks
Negative	Topical steroid	Resolved in 3 weeks
Negative	Topical steroid	Resolved in 6 weeks
Anti-DNA antibody, >300 IU/mL	Topical steroid for non-ulcerated lesions	Persistent lesions at 6th week
ANA, 1:160; rheumatoid factor, 35 IU/mL	Antibiotics (before skin consultation)	Resolved in 4 weeks
ANA, 1:640; anti-DNA antibody, <10 IU/mL; rheumatoid factor, 346 IU/mL	Topical steroid	Resolved in 4 weeks
Negative	Topical steroid	Resolved in 5 weeks
Negative	Topical steroid	Resolved in 5 weeks

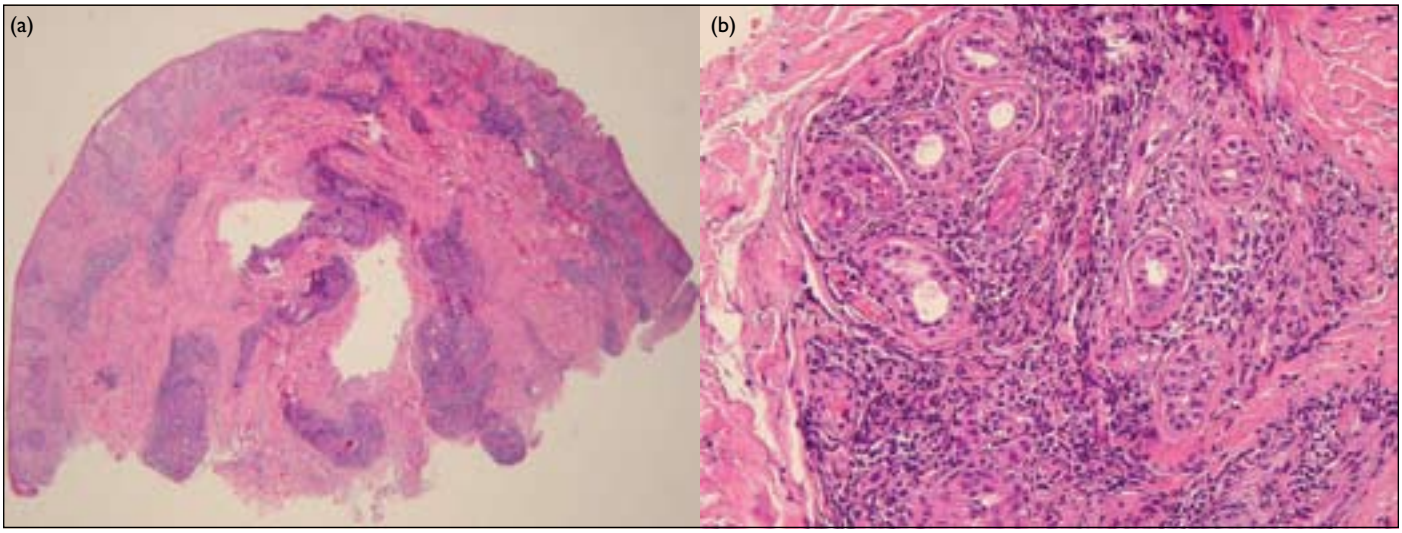

FIG 2. (a) Distinct superficial and deep perivascular and perisudoriferous lymphocytic infiltrate. The epidermis is partly necrotic and ulcerated (original magnification, x20). (b) High-power view showing dense sudoriferous lymphocytic infiltrate (original magnification x200)

Patient 7 was known to have SLE, and although the skin biopsy showed suspicion of vasculitis, they did not refute a clinical diagnosis of chilblains; in her chilblain lupus was likely. Chilblain lupus denotes chilblain lesions occurring in cutaneous and systemic forms of lupus erythematosus. Morphological differentiation of skin lesions between idiopathic chilblains and chilblain lupus is difficult. Viguier et al[6] reported two patients who initially presented with prolonged chilblains and lymphocytopenia and later developed SLE.[6] They suggested that persistence of cutaneous lesions beyond cold weather and female gender were significant factors associated with chilblain lupus. Patient 7 had persistent cutaneous lesions despite cessation of cold weather, which was in concordance with the study by Viguier et al.[6] Autoimmune markers were checked in all of our patients. Discounting patient 7 who had SLE, two had a positive ANA titre. Patient 4 had cold agglutinins in his serum. At the time of writing this article, none of the three other patients testing positive for such markers showed evidence of any underlying

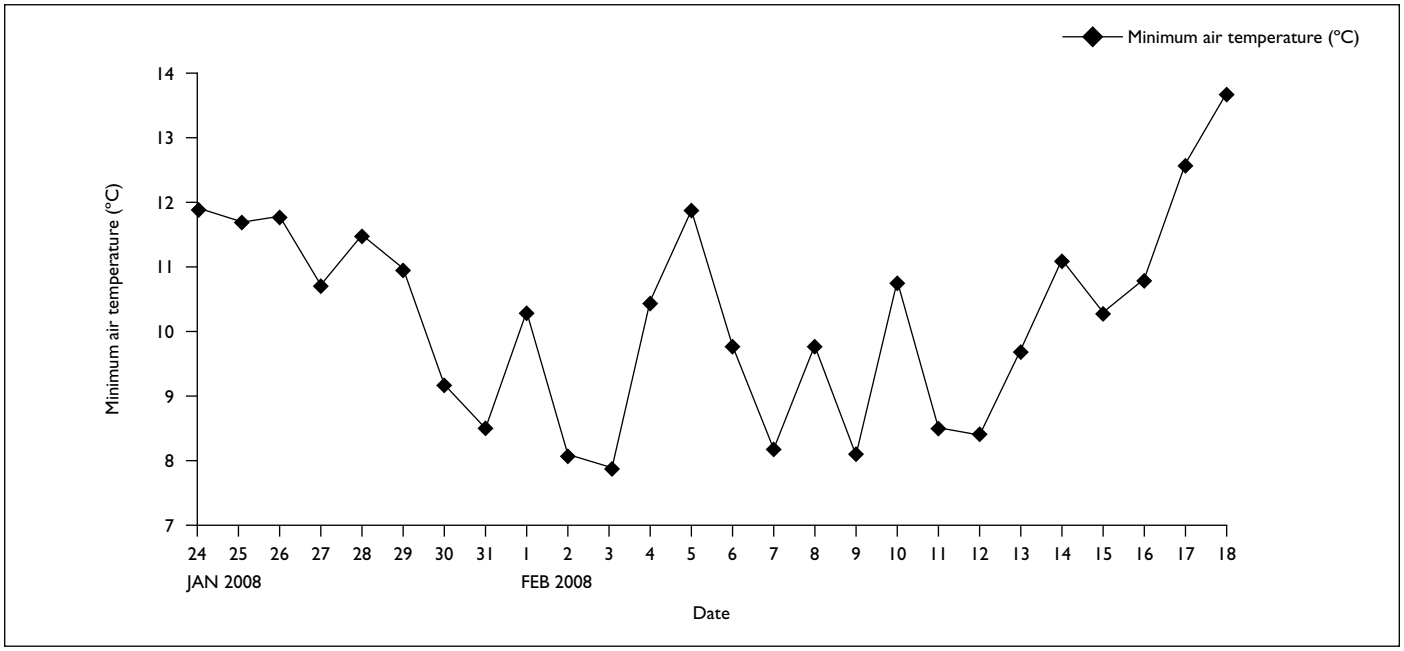

FIG 3. Minimum air temperatures during the prolonged cold weather from 24 January to 18 February 2008

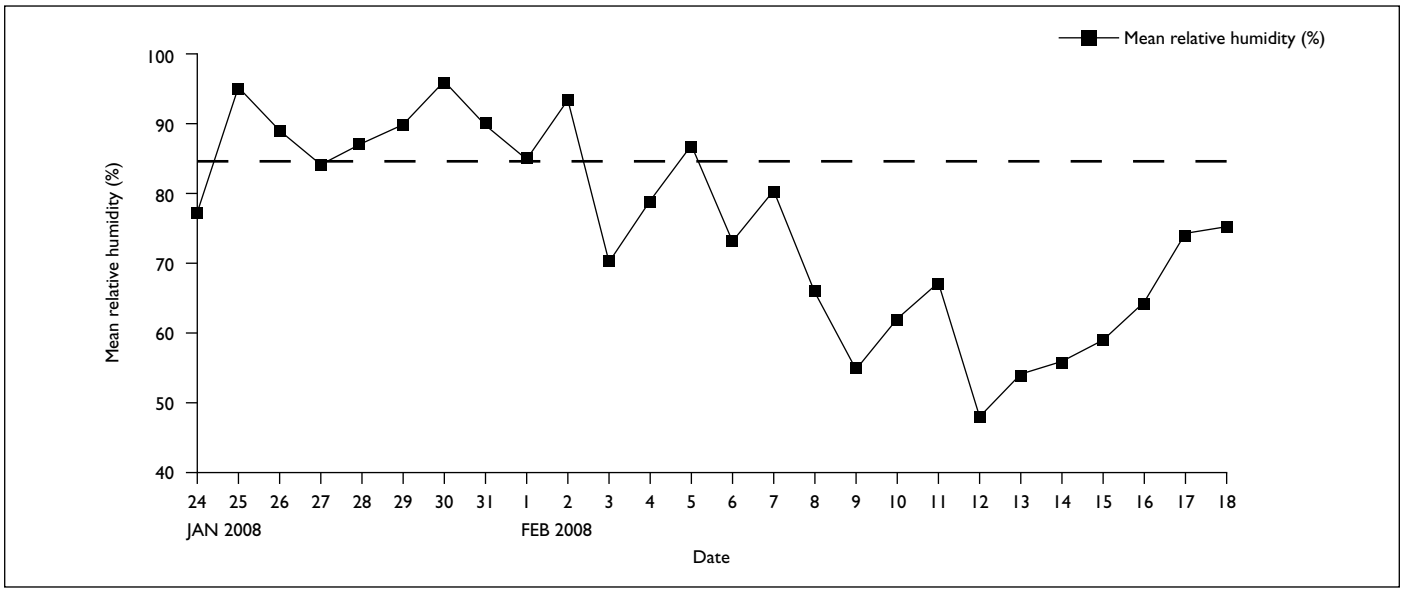

FIG 4. Mean relative humidity during the prolonged cold weather from 24 January to 18 February 2008

autoimmune disease.

General management of chilblains includes: cessation of or no smoking, avoiding cold exposure, and keeping extremities warm. These measures were used for all of our patients. Calcium channel blockers (nifedipine in particular) have been reported to be of benefit[7,8]; nifedipine was used to treat the significant symptoms of patient 2. Boehm and Bieber[9] reported successful treatment of one patient with refractory chilblain lupus, using mycophenolate mofetil. Incidentally, this was also used in our patient with SLE. However, her chilblains skin lesions had persisted up to her latest dermatological evaluation. In concordance to other reports,[6] the chilblains in most of our cases resolved spontaneously within a few weeks. Persistence beyond 6 weeks only occurred in patient 7. Recurrence of chilblains upon cold exposure is possible, as described by patient 3 who had attacks every winter for the past few years, though this winter the attack was the most severe.

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