Chilblains in Iraqi patients in Hilla City (Clinical study)

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Abstract:

Chilblain is a major medical problem in Iraqi population in winter time and the purpose of this study was to determine the clinical patterns of involvement in patients at Hilla city.

A total of 60 patients with chilblains were assessed in the department of dermatology in Merjan teaching hospital during the cold months between January 2007 to February 2008. Their ages ranged from 1.5-70 years (mean ± SD, 22.6±13.6 years).The duration of the disease ranged from 2-90 days (mean ±SD, 24.5±20.7 days). There were 40 females (66.6%) and 20 males (33.4٪) with a sex ratio of 2:1. The majority of patients were between 11-30 years of age (71.6%) and females forming about 79.5%, female: male ratio was about 3.8:1.

All patients were presented in January and February and the attacks in majority of patients started in December (20 patients) and January (30 patients) forming about 83.3%. Family history in close relatives were positive in 14 patients (23.3 %).Thirty one patients developed similar attacks at previous winters (51.6%). Numbers of previous attack of perniosis / winter were from 1-215 attacks.

Associated systemic and dermatological diseases were: hypertension (3.3%), diabetes mellitus (3.3%), systemic lupus erythematosus (3.3%), pregnancy (3.3%), atopic dermatitis (3.3%), allergic rhinitis (3.3%), anemia (1.6%) and hand eczema (1.6%).

Regarding the sites of the lesions, chilblains commonly affected either hands alone (48.3%), feet alone (33.3%) and to a lesser extent both hands and feet (18.3 %). The feet alone were commonly affected in females (88.2%) while the hands alone in males (60 %). Hands and feet were affected together most commonly in females (91%).Ears, nose and buttock were also affected (1.6%) for each.

Clinically, the types of lesions in patients with chilblains were: erythematous cyanotic swelling (81.6%), some with diffuse welling sausage - like (13.3%),erythema multiforme –like picture with target lesions (20 %), purpuric rash (15 %),ulcerated lesions (11.6%), eczematization with fissuring (11.6%),blisters or vesicles (6.6%),dusky red papules (6.6%), lupus erythematosus – like rash on dorsum of the hands (5%) and nodules (3.3%).

In Iraq, chilblains clinically could be presented in a form of erythema multiforme-like, erythema nodosum –like, lupus erythematosus-like or sausage-like swelling with cold fingers and toes, pain, tenderness and itching.

مرض تشرين (الشرث) عند المرضى العراقيين في مدينة الحلة, دراسة سريريه:

تمت دراسة 60 حالة من المرضى المصابين بمرض تشرين تتراوح أعمارهم بين 1.5-70 سنة أي بمعدل 22.6 سنة. وجد أن اغلب المصابين هم من الإناث ونسبتهم 66.6 % وعدد الإناث هو 40 وبلغ عدد الذكور 20 و نسبتهم 33.4 %. اغلب المرضى تم رؤيتهم خلال شهري كانون الثاني وشباط من سنتي 2007 و 2008. القابلية الوراثية عامل مهم في تكوين المرض والذي تم تأكيده بوجود تاريخ المرض لدى الأقارب من الدرجة الأولى للمصابين بنسبة 23 % . يصيب مرض تشرين عادة إما اليدين فقط (48.3 %) أو القدمين فقط (33.3 %) وكحد اقل اليدين والقدمين (18.3 %) . لقد وجد أيضا ارتباط هذا المرض مع عدد من الأمراض الجلدية والباطنية: ضغط الدم (3.3 %) ، مرض السكر (3.3 %)، داء الذئب ألاحمراري (3.3 %)، الحمل (3.3 %)، الاكزما الاستشرائية (3.3 %)، حساسية الأنف (3.3 %)، فقر الدم (1.6 %) و اكزما اليدين (1.6 %)

Introduction

Chilblain or perniosis is a moderately severe form of localized cold injury which occurs after exposure to non freezing temperatures and damp conditions [1, 2].The lesions are erythematous, mauve or purplish swelling which give rise to throbbing pain and itching, usually occurring at the onset of cold spells in the winter and lasting 2-3 weeks [3, 4] .In severe cases, the lesions may progress to blistering and ulceration [5]. They are common over the dorsum of fingers and toes and other exposed areas such as lower legs, thighs, nose and ears [3].

Perniosis occurs chiefly in women, children and elderly and there is a familial tendency [3]. Exact incidence of the disease is not known and the frequency of the chilblain varies with weather conditions [6]. In its early stages, there is a distinct seasonal incidence, the symptoms developing with cold weather and disappearing with warm weather [7]. In its later stages when chronic ulceration of the skin is present, the lesions tend to persist all year round. Thus, perniosis may be divided into 2 stages, an acute stage which is completely reversible and chronic stage in which permanent tissue changes have developed and in which ulceration may or may not be present, a stage which is never completely reversible[7].

The usual sites of the lesions are the dorsal aspects of the proximal phalanges of the hands, on the plantar aspect of the toes, along the inner border and dorsum of great toe and the region of the heel and Achilles tendon[7, 8]. With the onset of cold weather, the extremities begin to burn and itch and if examined at this stage, the skin will be red or cyanotic, cold and slightly swollen. Also there were erythematous papules and nodules, on blanching, commonly these lesions had a purpuric element. This rash was tender, cold to touch when compared to adjacent skin. In few cases, the rash was very similar to erythema multiforme[8]. Blebs of varying size, sometimes hemorrhagic with well defined margins develop if the exposure is prolonged. Upon coming into a warm environment, the itching and burning become intensified and too rapid warming by placing the extremities near an open fire or in hot water may produce vesicle not previously apparent[7,8].

The acute stage is fully developed within 12-24 hours of the initial exposure and lasts only a few days if the exposure to cold not repeated. If hemorrhagic blisters have developed, these usually rupture with the development of a superficial, weeping, raw-ham tinted patch. This fades to a brownish hue which persists covered by a thin layer of scaly skin [7, 9].

Pathophysiology of chilblain is largely unknown; however it represents an abnormal vascular response to cold [6]. Lewis (1941) elaborated his theory of the release of H-substance from skin cells as a result of damage by cold, such release would of course account for the itch, the redness and the swelling [10].

Chilblain may be idiopathic or secondary to an underlying disease like chronic myelocytic leukemia [11], cryoglobulinemia, dysprotenemia[12], anorexia nervosa[13] and systemic lupus erythematosus[14].

Chilblain lupus erythematosus (Hutchinson) is a chronic unremitting form of lupus erythematosus seen predominantly in women [15]. It occurs commonly on the digits, calves and heels. They are precipitated by cold damp climate[15]. Also can be precipitated by pregnancy[16]. Usually the chilblain lesions occur some years after the development of the discoid lesions on the face. When the discoid lesions remit with treatment, the chilblain persists[15]. The chilblain lesions are the result of microvascular injury secondary to exposure to cold and possibly hyper viscosity from immunological abnormalities[15].

The direct cause of chilblain in all cases is cold exposure. Although perniosis is generally benign in nature, an investigation of an underlying cause should be undertaken in selected cases which are recurrent chronic extending into warm seasons or poorly responsive to treatment [5, 17].

Most cases of chilblains resolve within few weeks without any residual effects. However, there is a tendency of recurrences of chilblains every year with the approach of winters[18]. The most important point in the management is prophylaxis with adequate clothing and warm living conditions to avoid exposure to cold [18].Once the lesions appear, treatment is usually symptomatic with avoidance of further exposure to cold. Many treatment modalities have been tried with variable results [3]. These include rewarming of the affected parts of the body, vasodilator calcium channel blockers like nifedipine, topical minoxidil, prazosin, topical antipruritic and iontophoresis [3].

Aim of Study: Chilblain is a major medical problem in Iraqi population in winter time. The purpose of this study was to determine the clinical patterns of involvement in patients at Hilla city.

Patients and methods

This is an observational study conducted in Merjan teaching hospital at Hilla city in the department of dermatology during the cold months between Jan 2007 and Feb 2008 in which 60 patients with chilblains were seen.

All patients with persistent painful or pruritic erythematous or dusky erythematous papules and plaques or painful swelling involving digits or other acral parts of the body during the period of the study were registered. A detailed history was taken from each patient regarding age, sex, occupation, duration of present episode of chilblains and family history of the disease. Each patient was also asked for any associated dermatological or systemic illness. History of atopy (hay fever, bronchial asthma, atopic dermatitis and allergic conjunctivitis) was evaluated in every patient and his close relatives.

Lesions of chilblains were examined to note the site and extent of involvement. In addition, dermatological as well as systemic examinations were carried out to find any associated diseases. Patients with a long duration of disease more than 3 months were investigated to rule out connective tissue diseases like systemic lupus erythematosus by doing complete blood count and antinuclear factors (ANFs) and patients who seem to be anemic were investigated by doing PCV count. Diagnosis of chilblain was based on history and clinical examination.

Results:

A total of 60 patients with chilblains were assessed .Their ages ranged from 1.5-70 years with a mean ± standard deviation (SD) of 22.6±13.6 years. The duration of disease ranged from 2 – 90 days with a mean ± SD of 24.5 ± 20.7 days. There were 40 females (66.6%) and 20 males (33.4%) with a female to male ratio of 2:1. The majority of patients were between 11-30 years of age (43 patients) forming about 71.6% and 30 patients were females forming about 79.5%, female to male ratio was about 3.8:1 (tables1&2).

All patients gave a history of exposure to cold weather or washing their hands and feet with cold water. All the patients were presented in January and February and the attacks in majority of patients were started in December (20 patients) and January (30 patients) forming about 83.3 %.Three patients had a longer duration of disease about 3 months. Family history in close relatives was positive in 14 patients (23.3%).

Thirty one patients developed similar attacks at previous winters (51.6%).Number of previous attacks of perniosis per winter were from 1-5 attacks table (3). Twenty - nine patients (48.4%) had the disease for the first time. Associated systemic and dermatological diseases are shown in table 4.

Hypertension was seen in 2 middle aged females with perniosis 50 and 58 years. Diabetes was also seen in 2 middle aged patients 47 years old female and 63 years old male. Chilblain lupus in association with SLE was seen in 2 female patients 25 and 32 years old with positive antinuclear factors. There were 2 pregnant females 18 years with 9 missed periods and 24 years with 5 missed periods. The first pregnant patient also had anemia and hypotension

Personal history of atopy was seen in 4 patients, 2 patients with atopic dermatitis, 8 years old male and 13 years old female and 2 patients with allergic rhinitis 12 years old male and 23 years old female .

Regarding the sites of lesions, chilblains commonly affected either hands alone in 29 patients (48.3 %)or feet alone in 20 patients (33.3%) and to a lesser extent both hands and feet in 11 patients (18.3%). The feet alone were commonly affected in females (88.2%) while the hands alone in males (60%). Hands and feet were affected together more common in females (91%) The parts commonly affected with perniosis were shown in table (5).

Clinical presentations in patients with chilblains were shown in table (6) Figures [1 – 8].

The rash in the majority of patients was bilateral symmetrical cold to touch with cold hands and feet associated with pain and itching. Lupus erythematosus-like rash affecting the dorsum of hands sparing the knuckles was seen in 3 female patients. Two patients were with real systemic lupus erythematosus with malar rash, arthralgia and positive antnuclear factors, one of them with positive anti DS-DNA Abs. These two patients had a long duration of chilblain about 3 months and got similar attacks at previous winters. The third female patient had similar rash of one month duration with negative ANFs.

Method

Table 1: The age distribution in female patients with Chilblains

Age / year	No. of patients	Percentage
1-10	5	8.3 %
11-20	16	26.6 %
21-30	14	23.3 %
31-40	2	3.3 %
41-50	2	3.3 %
> 50	1	1.6 %

Table 2: The age distribution in male patients with Chilblain

Age / year	No. of patients	Percentage
1-10	2	3.3 %
11-20	8	13.3 %
21-30	5	8.3 %
31-40	3	5 %
41-50	0	0 %
50>	2	3.3 %

Table 3: Number of previous attacks of perniosis

No. of previous attacks	No. of patients	Percentage
1	19	31.6 %
2	7	11.6 %
3	1	1.6 %
4	1	1.6 %
5	3	5

Table 4: Associated Dermatological and systemic disease with perniosis

Associated diseases	No. of cases	Percentage
Hypertension	2	3.3 %
Diabetes mellitus	2	3.3 %
SLE	2	3.3 %
Pregnancy	2	3.3 %
Atopic dermatitis	2	3.3 %
Allergic rhinitis (atopy)	2	3.3 %
Anemia	1	1.6 %
Hand eczema	1	1.6 %

Table 5: The affected sites in patients with chilblains:

Site of lesion	No. of cases	Percentage
Sides of fingers and dorsum of hands	34	56.6 %
Sides and dorsum of toes and feet	26	43.3 %
Soles and sides of heels and plantar surfaces of forefeet	8	13.3 %
Palms and palmar surfaces of fingers	6	10 %
Ears	1	1.6 %
Nose	1	1.6 %
Buttock	1	1.6 %

Table 6: Types of lesions in patients with chilblain:

Type of lesion	No. of patients	Percentage
Diffuse erythematous cyanotic swelling	49	81.6%
some with :Sausage-like diffuse swelling	8	13.3%
Erythema –multiforme like picture with target lesions	12	20 %
Ecchymotic (purpuric) rash.	9	15%
Ulcerated lesions	7	11.6%
Eczematization, exfoliation with fissuring.	7	11.6%
Vesicular or blistering.	4	6.6%
Dusky red papules.	4	6.6%
Lupus erythematosus-like rash on dorsum of hands	3	5%
Nodules.	2	3.3%

* Some of patients presented with more than one type of lesions.

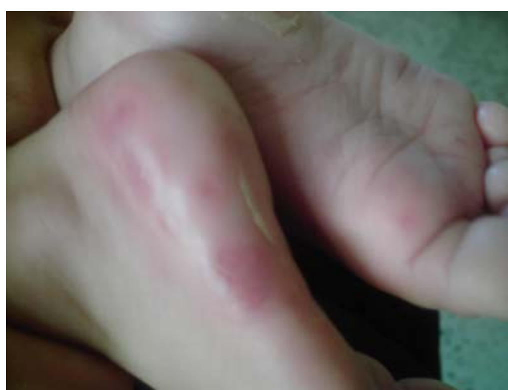

Figure 1: Chilblain affecting heels and sides of feet in a child (erythematosus cyanotic swelling)

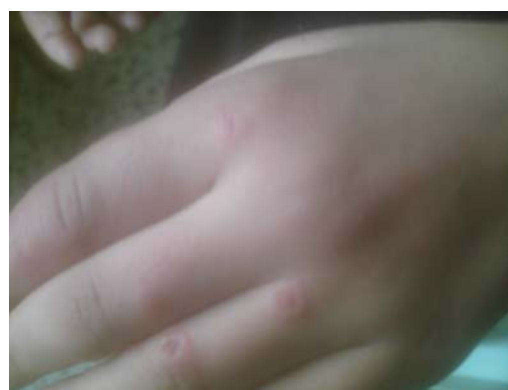

Figure 2: Chilblain affecting the dorsum of fingers, EM-like (target lesions)

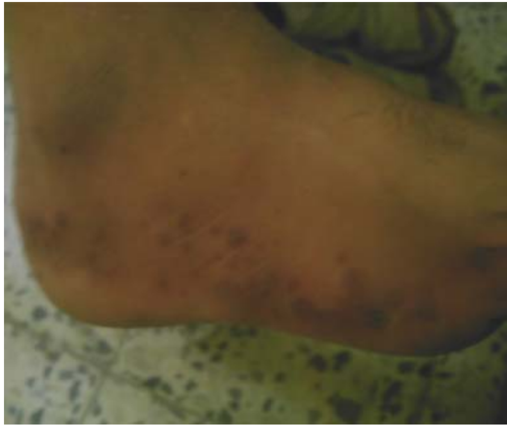

Figure 3: Chilblain affecting the dorsumand sideof foot (ecchymotic rash)

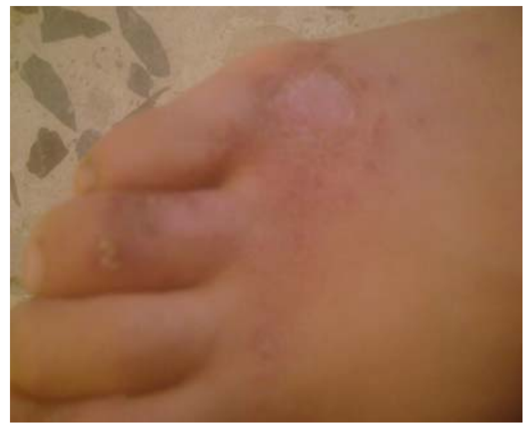

Figure 4: Chilblain affecting dorsum of toes

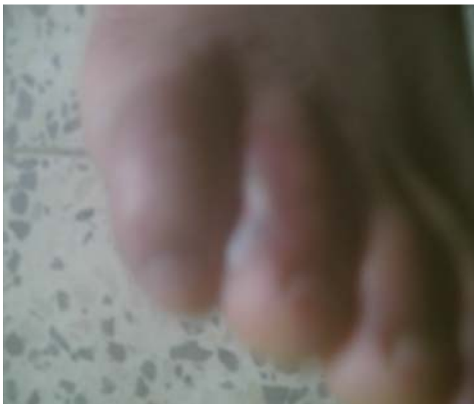

Figure 5: Chilblain affecting the toes of foot with erythematous swelling and blistering.

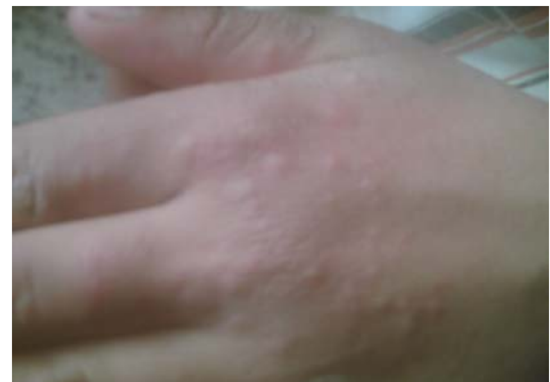

Figure 6: Chilblain affecting the dorsumof hand in a form of red papules.

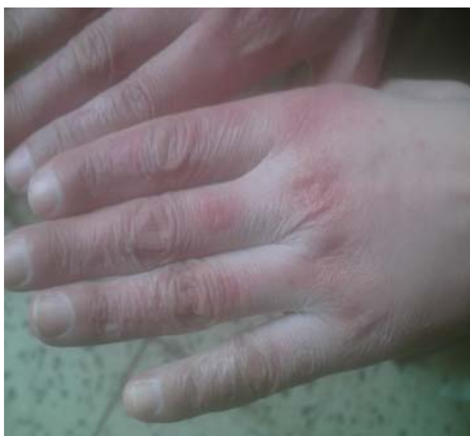

Figure 7: Chilblain affecting the dorsum of hands in LE-like rash involving the spaces between fingers joints sparing the knuckles.

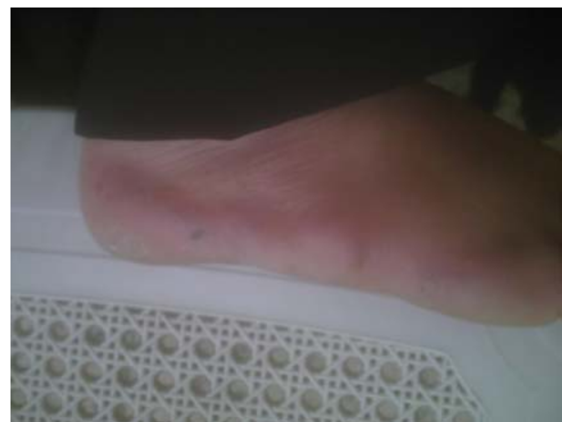

Figure 8: Chilblain affecting the side of heel with erythematous nodules.

Discussion:

Chilblain or perniosis is a localized cold injury which represents an abnormal response to non-freezing temperatures in humid conditions[19]. Incidence now become less frequent in the well developed countries due to more efficient central heating and their people protecting themselves from cold with suitable warm clothing[20]. Although Iraq is one of the subtropical countries, still perniosis is common diseases because there is always diurnal and seasonal fluctuation of temperature between day and night, summer and winter. These changes of temperature are often more important in precipitating the disease than cold alone. In addition, there is some lack of central heating and many unaccomodated susceptible people not protecting themselves from cold by suitable warm clothing [8, 21].

Chilblains can occur at any age are said to be more common in females [17, 22]. In the present study, the majority of patients were between the age of 11-30 years (71.6%) with a female-to male ratio of about 3.8:1. These results were nearly comparable to the results of a previous Iraqi study(1993) in which the patients were between the ages of 11-30 years forming about 81.6 % and female to male ratio was about 2.8:1[23]. In a study that conducted in Pakistan at Abbott Abad, a moderately cold weather station in 2006,the main age groups affected were between 11-30 years (60%) and males were affected more often than females with a male to female ratio was about 1.5:1. This is because the study was achieved at a military hospital and the majority of patients were serving soldiers[24].

In this study and in a previous Iraqi study (1993), the increase in female-to male ratio might be influenced by the following factors:
1- Genetic factors: female have genetic susceptibility of perniosis more than males.
2- Hormonal influence: female sex hormones exacerbated the disease.
3- Thick subcutaneous fatty layer in females with its poor blood supply is more liable for perniosis attack.

The present work had shown that sudden change in temperature and cold exposure as occur in December, January and February was the most important precipitating factor. In addition, repeated immersion of hands and feet in cold water as a part of housewife care might increase the incidence of the disease. Long standing might also increase the incidence of perniosis in the feet.

Genetic susceptibility was an important factor in the development of the disease as confirmed by positive family history in close relatives of patients in 23.3 % of cases.

In this study, the fingers and toes were commonly affected bilaterally and nearly symmetrical. Unilateral and asymmetrical lesions like involvement of one hand and one foot was also observed and seen in 9 patients (15%).Rare involvement sites like buttock was seen in 1.5 years old child female. This was not mentioned in literatures.

In the present attack, it was noted that, the affected parts were the same areas which were affected during the previous attacks, so there was area of susceptibility for perniosis in relation to each patient.

Types of lesions in chilblains were nearly similar to what had been reported but in the present work as in previous Iraqi study (1993)[23], lesions of multiple papules on the hands with target lesions similar to erythema multiforme (20%) and nodules on the heels and sides of soles similar to erythema nodosum (3.3 %) were demonstrated. In addition, the swelling of whole finger can be diffused, sausage-like in 13.3 % of cases.

Three patients had lupus erythematosus – like rash involving the dorsum of the hands and sparing the knuckles, two of them were with positive ANFs and other features of systemic lupus erythematosus like malar rash, renal involvement and arthralgia with a longer duration of chilblain, but in one patient, the onset is acute with negative ANFs and no other features of SLE, so in our study, chilblain could present in a form of LE-like rash even with negative investigations and without other manifestations of SLE.

In conclusion, in Iraqi patients, chilblains clinically could be present in a form of erythema-multiforme-like, erythema nodosum –like, lupus erythematosus - like or sausage-like swelling with cold fingers and toes, pain, itching and tenderness.

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