

Idiopathic Chilblains

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ABSTRACT

BACKGROUND: Chilblains is a benign condition infrequently encountered in clinical practice; its resemblance to vasculitis or peripheral thromboemboli can often result in an extensive and unnecessary diagnostic work-up.

METHOD: Three cases of chilblains seen by our Rheumatology service, along with 113 documented cases, were reviewed.

RESULTS: Chilblains is characterized by painful red-to-purple papular lesions involving the acral surface of fingers or toes that resolves with symptomatic treatment. Female sex and low body mass index are risk factors.

CONCLUSION: Distinct clinical features of chilblains can be used for early recognition and management, thus avoiding unnecessary diagnostic testing and delays in patient care. © 2009 Elsevier Inc. All rights reserved.

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KEYWORDS: Idiopathic chilblains; Papular rash; Pernio

Idiopathic chilblains, often referred to as pernio, manifests as inflammatory cutaneous lesions in patients exposed to nonfreezing weather during late winter or early spring.¹ These lesions typically present as painful erythrocyanotic discoloration (often with cutaneous necrosis) of the fingers or toes (or both) and is frequently misdiagnosed as vasculitis or an embolic event, leading to an elaborate and expensive workup. We recently encountered 3 patients with idiopathic chilblains, all of whom possessed the characteristic skin lesions along with other key findings. We propose certain clinical features that can be utilized to make the diagnosis of chilblains at the time of first encounter. Immediate diagnosis of chilblains can eliminate a time-consuming diagnostic process that adds expense as well as an unnecessary prolonged period of patient discomfort without definitive management.

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ILLUSTRATIVE CASE

A 33-year-old woman was referred to the rheumatology service for painful red discolorations with ulcers on her toes. Her symptoms first occurred in February, with painful red blisters on the distal plantar aspect of her right first toe. She subsequently noted similar small painful lesions on both of her feet (Figure). She did endorse cold extremities, both hands and feet, which were especially worse during the



Figure Typical chilblains lesion on the toes of patient 2.

winter months. The patient denied trauma, skin tricolor changes, fever, or joint pain. She was a nonsmoker with no known past medical history. Concurrent medications included oral contraceptives and ibuprofen. The family history was noncontributory. On physical examination, the patient was a thin female, body mass index of 18.6 kg/m, with extremely tender erythematous macules on the plantar surface of her bilateral great toes. All of her toes were cool to touch, without any other significant skin changes. The laboratory values include normal complete blood count, erythrocyte sedimentation rate (ESR), hypercoagulability panel, and a negative antinuclear antibody. A lower extremity duplex scan to evaluate arterial flow also was unremarkable. The patient was advised to discontinue oral contraceptives and keep her feet warm. During the follow-up visit in about 1 month, the patient had complete resolution of her toe lesions.

REVIEW OF OUR CASES

Three cases were referred to our rheumatology service for evaluation from December through February. All were female, with bilateral lesions involving fingers and toes. The clinical features of these patients are displayed in Table 1. The primary cutaneous findings were painful erythematous or purplish papules on the acral surfaces of fingers and toes. One patient had digital edema and one had erosion and bleeding from these lesions. Extensive laboratory workup for hypercoagulability and vasculitis was unremarkable, except for an elevated ESR in patients 1 and 3. The elevated ESR likely represents a nonspecific marker of an inflammatory process at the lesion site as well as these patients' older age. All patients had complete resolution of their lesions once the diagnosis was considered.

LITERATURE REVIEW

Chilblains was originally observed in damp, nonfreezing weather of Western European countries such as Great Britain and France;² however, more recent studies are reporting increasing numbers of cases from North America, possibly due to climate changes. Our review of the literature uncovered 113 reported cases of idiopathic chilblains available in the English language. The clinical features, treatment, and outcomes of these cases are shown in Table 2. The data clearly reveal female predominance, with 77 females, compared with 36 male cases. All cases were seen during the winter to early spring months of November to April.

Subjects' weight may be a predisposing factor; Simon et al³ and St Clair et al⁴ have reported 6 cases of chilblains with low body mass index (BMI). White et al⁵ specifically documented that all 3 of their patients, 18%-22% below

the ideal body weight, had complete resolution of their symptoms with weight gain and keeping the affected area warm. These findings highlight the possibility that there may be an association between low BMI and thermal irregularity predisposing patients to chilblains.

The most common sites of chilblains lesions are the hands (41 of 113 cases) and feet (90 of 113 cases). The apparent large difference in prevalence between upper and lower extremities is due to inclusion of the Spittell et al⁶ study, which excluded patients with only upper extremity chilblains lesions. Other less frequent locations were ear, nose, and thigh. All patients were treated with conservative measures by keeping the affected area warm, along with occasional addition of vasodilatory agents such as nifedipine or prazosin. However, 23 cases treated with nonpharmacological therapy alone had similar outcomes. All except 3 of the patients had complete resolution of their lesions. One

CLINICAL SIGNIFICANCE

- Chilblains present as bilateral painful lesions on distal surfaces of fingers and toes, resembling vasculitis.
- Chilblains occur in nonfreezing and damp conditions, typically in late winter, early spring.
- It is a clinical diagnosis and can be treated conservatively with excellent prognosis.
- Low body mass index and female sex could potentially be predisposing factors.

Table 1 Clinical Features of Three Cases Referred to Cedars-Sinai Medical Center

Patient	Age (Years)/Sex	BMI (kg/m ²)	Time of Year	Location of Lesion	Bilateral	Laboratory Testing	Treatment	Resolution
1	84/F	23	Jan	Fingers	Yes	ESR 64; CRP 1.01	Keep warm Revatio	Yes
2	33/F	18.6	Dec-Jan	Toes	Yes	WNL*	Keep warm	Yes
3	66/F	N/A	Feb	Fingers and toes	Yes	ESR 74; CRP low	Keep warm, tramadol, topical antiseptics, prednisolone, clindamycin	Yes

BMI = body mass index; CRP = C-reactive protein; WNL = within normal limits; N/A = not available; ESR = erythrocyte sedimentation rate.

Table 2 Reported Cases of Idiopathic Chilblains¹⁻¹⁶

Author, Ref	# of Cases	Sex or M:F	BMI (kg/m ²)	Time of Year	Hands	Feet	Other	Bilateral Lesion	Treatment	Resolution
Chan et al ⁷	11	6/5	NA	February	8	7	0	NA	1. Keep affected area warm 2. Nifedipine (1 of 11)	Yes (1-no response)
McCleskey et al ¹⁵	1	M	NA	February	1	0	0	Yes	1. Keep affected area warm 2. Oral prednisone (to reduce edema)	Yes
Simon et al ³	5	0/5	15-18	Dec-April	3	5	0	Yes (4)	1. Keep affected area warm 2. Nifedipine (2 of 5)	Yes
Wessagowit et al ¹⁴	1	M	NA	NA	0	1	0	Yes	1. Keep affected area warm 2. Topical steroid	Yes
Parlette and Parlette ¹³	1	F	NA	January	0	1	0	Yes	1. Keep affected area warm 2. Nifedipine	Yes
Goette ¹	7	4/3	NA	Nov-Jan	4	3	0	Yes (3)	1. Keep affected area warm 2. Nifedipine (4 of 7)	Yes (1-unknown)
Jacob et al ⁸	1	F	NA	March	0	1	0	Yes	1. Keep affected area warm 2. Prazosin	Yes
Weston and Morelli ¹⁰	8	4/4	NA	November March-April	5	7	4 (ear)	NA	Keep affected area warm	Yes
White et al ⁵	3	0/3	NA*	Nov-April	0	3	0	Yes	1. Keep affected area warm 2. Weight gain	Yes (1-improvement)
Price and Murdoch ¹²	5	4/1	NA	NA	0	0	5 (thigh)	Yes (5)	Keep affected area warm	Yes
St Clair et al ⁴	1	F	16	NA	1	1	0	Yes	Weight gain – with gluten-free diet	Yes
Herman et al ¹⁶	9	F	NA	NA	2	9	0	Yes (2)	Antibiotics – with no effect No other specific therapy noted	Yes
Scurry and Cowen ⁹	1	M	NA	NA	0	0	1 (ear)	NA	NA	NA
Spittell and Spittell ⁶	39	11/28	NA	NA	1**	39	0	Yes (34)	1. Keep affected area warm 2. Prazosin	Yes
Viguier et al ¹¹	21	5/16	NA	NA	16	13	4 (3 ear; 2 nose)	NA	NA	NA

*Did not provide BMI (body mass index), but ideal body weight was significantly below normal (18%-28% below ideal body weight).

**Patients with only upper extremity chilblains lesions were excluded.

patient was lost to follow-up,¹ one had no response and was later found to have lupus chilblains,⁷ and one was not followed long enough to see full recovery.⁵

DISCUSSION

The exact pathogenesis of idiopathic chilblains is unknown. Several studies have described the process as a vasculopathy. It is hypothesized that susceptible individuals may be at risk of developing these lesions secondary to a disruption of normal neurovascular responses to dermal temperature changes. The concept of a cold-induced vasodilatory reflex has been described as a protective physiologic mechanism that intermittently opens blood flow to allow reperfusion and prevent skin ischemia.¹⁷ Studies have shown abnormally high levels of vasoconstrictive agents such as endothelin-1 in subjects with idiopathic Raynaud phenomenon and acrocynosis.¹⁸ It has been suggested that patients with chilblains have persistent or prolonged cold-induced vasoconstriction leading to hypoxemia and a subsequent secondary inflammatory reaction.^{1,3,6,8} The association of chilblains with a low BMI noted in prior studies,³⁻⁵ also seen with 2 of our patients, suggests that this might be a predisposing factor for this unusual condition. Although there are no pathognomonic histological findings for chilblains, many

studies have shown prominent perivascular inflammatory processes involving mononuclear cell infiltration with intimal wall and papillary dermal edema.^{19,20}

Chilblains are almost always seen during the months of November to April under nonfreezing and damp conditions. It is thought that humidity adds to the chilling effect by enhancing thermal conductivity.¹³ The characteristic lesions usually appear within a few hours of cold exposure of the affected area.^{1,3,12} Although chilblains occur in all ages and sexes, its female predominance might reflect choices of footwear. Typically, it manifests as painful erythematous or purple lesions with associated swelling or itching. Occasionally there can be overlying erosions, ulceration, or blistering.^{1,2} The findings mostly affect bilateral acral areas of hands and feet, as seen in all 3 of our cases. Other sites such as nose¹¹ and ear,⁹⁻¹¹ as well as the lateral aspect of thighs^{12,21} and buttocks,²² also have been reported. In Table 3, we have outlined the key diagnostic features of chilblains that would be helpful in a clinical setting to make an early and specific diagnosis.

The key therapy for chilblains is protecting the affected area and avoiding further cold exposure. Although pharmacological modalities like vasodilators have been employed, there are no controlled studies to show if they are effective. Acute chilblains or pernio is a self-limiting process that

Table 3 Key Diagnostic Features of Chilblains

Hands or feet with characteristic lesions – painful, dusky erythematous/purplish papules with edema
 Bilateral lesions
 Temporally associated with non-freezing cold exposure
 Self limiting — improves with warming the affected area
 Predominantly seen in females
 Associated with low body mass index
 Presentation during late winter/early spring (Nov-April)

usually resolves within days to months; although there can be a chronic form of waxing and waning lesions with persistent cold exposure.

CONCLUSIONS

Idiopathic chilblains, or pernio, is an uncommon entity that is characterized by bilateral painful bluish-purple lesions, frequently affecting the fingers and toes associated with prior exposure to nonfreezing and humid conditions in the late winter and early spring months. Although the pathogenesis is unknown, factors such as low BMI and sex might predispose patients to such a condition. We propose that the key features of chilblains can be utilized to help distinguish this condition from other entities such as thromboembolic disease or vasculitis. Recognizing these clinical features will prevent further delays in diagnosis and therapy, as well as unnecessary utilization of costly diagnostic procedures.

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