

Erythrocyanotic Discoloration of the Toes

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A 35-year-old woman from Virginia presented with purplish, tender toes. She was diagnosed with pernio and successfully treated, initially with nifedipine and subsequently with management of the affected area's environment. Clinical presentations and treatment strategies are discussed below.

Pernio—also called perniosis or chilblains—is a common, inflammatory, dermatologic condition associated with a cold, humid climate. The inflammatory lesions of pernio may be pruritic or painful, erythematous to violaceous, plaques, papules, or nodules, which may have overlying blisters or ulcerations. The condition is frequently misdiagnosed. Proper diagnosis relies primarily on the patient's history and clinical picture. Histologic examination is typically not needed or definitive. Our patient had moderately disabling pernio, which responded promptly to therapy with a calcium channel blocker.

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Case Report

A 35-year-old white woman from northern Virginia, in excellent general health, presented in January with purplish toes that were markedly tender to pressure. This clinical picture was now recurrent for the third successive winter with a trend toward worsening. Initially, she had experienced only an itchy, burning sensation in her toes. Diflorasone diacetate cream and a systemic steroid dose pack had partially moderated symptoms during her first episode 2 years ago. Discomfort from the pressure of shoes and even socks induced her to go bare-footed around the house. The discoloration and other symptoms were constant, not episodic. The patient's condition disappeared with the occurrence of warm weather (late spring) each year and also during a mid-winter visit to Florida during the second winter.

Physical examination revealed splotchy to mostly confluent erythrocyanotic discoloration of the plantar (Figure 1) and distal toe pads (Figure 2). The involved areas were very tender to pressure. Her distal pulses were normal, as was the rest of her cutaneous examination.

Based on the patient's suggestive medical history and physical findings, a laboratory evaluation and a therapeutic trial of nifedipine (10 mg twice daily)



FIGURE 1. Erythrocyanotic discoloration of the plantar toe pads.



FIGURE 2. Erythrocyanotic discoloration of the distal toe pads.

were initiated prior to considering a biopsy. Laboratory studies included: complete blood count; hepatic, renal, and electrolyte studies; serum protein electrophoresis; cryofibrinogen titer; cryoglobulin titer; cold agglutinin titer; cold hemolysis screen; and an antinuclear antibody test using human substrate. All showed normal findings. She had noticeable improvement within 2 days of initiating therapy, and marked improvement in symptoms and physical findings by 7 days. Additionally, the patient was advised that both nylon socks and sneakers would increase foot perspiration, which combined with cool temperatures could aggravate her problem. She remained free of pernio in 2 subsequent years with the prophylactic use of nifedipine from mid-October to April. Mild headache and dizziness associated with her nifedipine dose would dissipate within 1 hour of onset. By the third year post diagnosis, she was able to control her pernio with only warmth, low local moisture, and exercise.

Comments

Pernio (perniosis or chilblains) is a common inflammatory condition characterized by symptomatic skin lesions, which are precipitated by exposure to a moderate degree of cold in a humid climate.^{1,2} Lack of central heating is a predisposing factor.³ Humidity is thought to augment the chilling effect by enhancing thermal conductivity.¹ Perniosis is frequently misdiagnosed by both primary care physicians and specialists because of its relatively low incidence in low-humidity, centrally heated North American homes. It may also be confused with secondary pernio of chilblain lupus erythematosus or other illness. A detailed history and physical examination best make the diagnosis of pernio. Perniosis occurs in either sex at

any age, but it is most common in women, children, and the elderly.^{2,4} The lesions typically worsen in elderly patients and resolve spontaneously in younger patients.⁴ Patients describe their symptoms as a burning sensation, itching, or pain.² On examination, the pernio lesions are inflammatory, erythematous to violaceous macules, papules, plaques, or nodules, sometimes with overlying blisters, erosions, or ulcers.¹ Typically, the lesions are bilaterally symmetrical and acral.⁵ Superficial perniosis typically involves the terminal digits, heels, nose, and ears.⁵ Deep perniosis is seen on the thighs and buttocks as dusky erythematous plaques that histologically show change extending into the subcutis.¹ The superficial pernio shows upper dermal change only.

The exact pathogenesis of pernio is unknown, but many authors suggest it is of vascular origin.⁶ The histologic description of perniosis is controversial.¹ This may be due, in part, to the tendency of physicians not to distinguish between idiopathic pernio and secondary pernio lesions occurring in the setting of chilblain lupus erythematosus or other illness.¹ As part of the diagnostic process, appropriate laboratory studies should be obtained to rule out systemic disease. Systemic diseases, such as systemic lupus erythematosus, Behçet's disease, and chronic myelomonocytic leukemia, may mimic idiopathic perniosis.^{1,3}

Proper management of pernio can speed its resolution. Sufficiently warm clothing, appropriate home heating or warm climate, and avoidance of abrupt temperature change can lessen or resolve pernio symptoms.^{4,5} Also, keeping feet dry is important since moisture enhances cold injury. Weight-reduction and exercise are described as helpful in preventing recurrent perniosis⁴; our patient is convinced that aerobic

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exercise is an effective part of her treatment. Avoidance of smoking should also be recommended to patients.² Less frequently recommended therapies include nicotinamide, phenoxybenzamine, erythrocytic doses of ultraviolet B light, and sympathectomy.^{4,5}

Nifedipine therapy has proven to be the most effective for pernio. Rustin *et al.*⁶ found that no patient on nifedipine developed new pernio, but all patients on placebo relapsed or developed new pernio lesions. Nifedipine's mode of action is unknown, but it possesses vasodilatory, anti-inflammatory, and anti-platelet properties. Using Laser Doppler velocimetry, increased digital cutaneous blood flow was noted in patients treated with nifedipine.⁶ In the same study, nifedipine in daily doses of 20 to 60 mg resolved existing lesions and prevented new ones. Nifedipine is usually well tolerated, but it may cause headaches, facial flushing, dizziness, ankle swelling, and nausea at therapeutic doses.^{6,7} These side effects may be reduced by initiating therapy with small doses and gradually increasing to therapeutic doses.⁷ Management with nifedipine is effective in 70% of patients with severe unremitting chronic idiopathic pernio of both superficial (acral) and deep (thigh) types.⁷

Pernio is a very bothersome problem that is often misdiagnosed. Once the diagnosis is made, a thorough history, physical, and laboratory evaluation are required to rule out systemic disease. Lesions of pernio have been associated with macroglobulinemia, dysproteinemia, myelodysplastic disease, and lupus erythematosus.⁸ Therapy of idiopathic pernio is usually simple and very effective.

REFERENCES

1. Crowson AN, Magro CM: Idiopathic perniosis and its mimics: A clinical and histological study of 38 cases. *Human pathol* 28: 478-484, 1997.
2. Duffill MB: Milkers' chilblains. *N Z Med J* 106: 101-103, 1993.
3. Fisher DA, Everett MA: Violaceous rash of dorsal fingers in a woman. *Arch Dermatol* 132: 459-464, 1966.
4. Berger TG, Elias PM, Wintroub BU: Manual of Therapy for Skin Diseases, pp 51-52. New York, Churchill Livingstone, 1990.
5. Page EH, Shear NH: Temperature-dependent skin disorders. *J Am Acad Dermatol* 18: 1003-1017, 1988.
6. Rustin MHA, Newton JA, Smith NP, *et al.*: The treatment of chilblains with nifedipine: the results of a pilot study, a double-blind placebo-controlled randomized study and a long-term open trial. *Br J Dermatol* 120: 267-275, 1989.
7. Greaves MW, Dowd PM, Hawk JLM: Dermatopharmacology: drugs around the corner. *J Am Acad Dermatol* 19: 323-329, 1988.
8. Goette DK: Chilblains (perniosis). *J Am Acad Dermatol* 18: 257-262, 1990.