









Pernio is a non-infectious inflammatory skin condition presenting after exposure to cold as primary or an

History

Most patients with pernio present with a history of recurrent painful and/or pruritic, erythematous, violaceous papules or nodules on the fingers and/or toes. Most cases of pernio resolve within 2-3 weeks. Elicit a history of cold exposure or repeated episodes of cold exposure.

Physical

Pertinent findings in pernio are limited to the skin. Cutaneous pernio lesions present 12-24 hours after cold exposure as red or violaceous macules, papules, nodules, or plaques, which may form vesicles or ulcerate. Pernio lesions occur on acral areas, are associated with burning or pruritus, and last 1-3 weeks.

Causes

The direct cause of pernio is cold exposure; specifically, exposure to both mild nonfreezing cold and humidity seems to be required.^{2,3} Chronic pernio may be secondary to various systemic diseases as follows:

- Chronic myelomonocytic leukemia⁴
- Anorexia nervosa⁵ : Low body mass index may predispose to pernio.²

- Dysproteinemias
- Macroglobulinemia
- Cryoglobulinemia, cryofibrinogenemia, cold agglutinins
- Antiphospholipid antibody syndrome
- Raynaud disease⁶
- Celiac disease⁷
- Variants
- Kibes (equestrian cold panniculitis): Erythrocyanotic plaques occur on the upper lateral thighs of women who ride horses. Histology is characterized by an intense perivascular infiltrate extending into subcutaneous fat.
- Chilblain lupus erythematosus^{8,9} : Violaceous "pernio" plaques appear prominent over dorsal interphalangeal joints, often with positive antinuclear antibody (ANA) or rheumatoid factor (RF). Histologic and immunofluorescent evidence of lupus is present in the skin lesions. Half of the patients have associated facial discoid lupus lesions, and 15% develop systemic lupus.
- Drug-induced pernio: Sulindac induced cases have been reported

Laboratory Studies

- CBC count and sedimentation rate should be obtained to rule out associated leukemia.
- Antiphospholipid antibody panel: Review of patients presenting with pernio shows an increased incidence of antiphospholipid antibody syndrome.
- Cryoglobulins, cryofibrinogen, and cold agglutinins generally are absent but should be considered as part of the laboratory evaluation in a patient with chronic pernio. Because of occasional false-negative cryoprecipitate screening results, consider hepatitis C antibody screening or even rheumatoid factor (RF) as a marker for cryoglobulinemia in select cases.
- Antinuclear antibody (ANA): Pernio lesions can occur in the setting of lupus erythematosus.
- Serum protein electrophoresis (SPEP) and quantitative immunoglobulins: Dysproteinemias and macroglobulinemia, causing increased serum viscosity, may be associated with pernio.

Histologic Findings

Pernio can often be diagnosed on the basis of clinical findings. Biopsy may be indicated to rule out other inflammatory processes in difficult chronic cases. Punch biopsy is adequate. There is variable epidermal spongiosis or necrosis. Intense papillary dermal edema is present. A superficial and deep perivascular lymphocytic infiltrate is seen, with the described "fluffy edema" of vessel walls. Lymphocytic vasculitis may be present.

Medical Care

- Prophylactic warming of acral areas, achieved by heat and appropriate clothing, best prevents pernio.
- Ultraviolet light, given at the beginning of the cold, damp season, has been touted as preventing outbreaks of pernio in prone individuals. Pathogenesis was loosely based on damaging the minute vessels and minimizing their ability to vasoconstrict with subsequent cold exposure. However, in at least one double-blind study, ultraviolet therapy was of no value in prophylaxis of pernio.¹¹
- Avoidance of nicotine may help alleviate pernio

The use of topical and systemic steroids, vasodilators, intravenous calcium followed by intramuscular vitamin K, and ultraviolet B radiation has been anecdotally reported in the literature. In most cases of pernio, the value of these agents is questionable and often disputed.

Calcium channel blockers

Peripheral arterial vasodilators may be effective in the treatment and prevention of pernio.^{7,12,13}

Nifedipine (Procardia)

Relaxes coronary smooth muscle and produces coronary vasodilation, which in turn improves myocardial oxygen delivery. Small studies have shown this drug to be effective in reducing symptoms associated with severe recurrent pernio. Currently is considered DOC.

Adult

10-20 mg PO tid

Pediatric

Not established