

The diagnosis and management of pernio (chilblains)

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Abstract

Perniosis (chilblains) is an abnormal reaction to cold that occurs most frequently in women, children, and the elderly. Chilblains may be idiopathic and self-limited, or associated with systemic diseases. This article discusses the approach to the diagnosis and treatment pernio and explains the concepts of acrocyanosis and erythrocyanosis.

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Introduction

Perniosis (chilblains) is a common, sometimes familial, condition. It is characterised by cutaneous lesions that often occur after exposure to cold and high humidity (dampness) during cold months of the year. It is an abnormal reaction to cold that occurs most frequently in women, children, and the elderly. Lesions tend to become worse in the elderly, and improve spontaneously in younger patients. The disease seems to be more common in environments where heating is inadequate for a few months of the year and is less common in localities characterised by harsh frigid winters where adequate home heating is the norm. Exposure to cold water sometimes seems to play a role.

The incidence of pernio in South Africa is unknown. The incidence varies with climate, approaching 10% annually in England. Chilblains may be idiopathic and self-limiting, or associated with systemic diseases, including chronic myelomonocytic leukaemia, viral hepatitis, HIV infection, rheumatoid arthritis, the use of weight reduction medications, anorexia nervosa, dysproteinaemia and connective tissue disorders, especially lupus erythematosus. Perniosis occurring in lupus erythematosus is known as chilblain lupus erythematosus or Hutchinson's lupus.

Pathogenesis / aetiology

The pathogenesis of pernio is not well-understood. Cold is a requirement for the development of symptoms. Chilblains are caused by a combination

of arteriolar and venular constriction, the latter predominating on rewarming with exudation of fluid into the tissues. Perniosis can be induced in susceptible individuals by prolonged exposure to temperatures above freezing point in humid climates. Tight clothing and in some cases, particularly during childhood, dysproteinaemia may play a part.

Clinical features

Patients present with recurrent painful, tender, erythematous-violaceous plaques on the distal extremities, especially the fingers and the toes (Figure 1). Pernio usually involves the hands, feet, ears and face in children, the legs and toes in women, and the hands and fingers in men. Lesions are more common on the dorsal aspect of the digits. Other exposed areas, such as the nose, face and ears, may also be affected. Lesions may be accompanied by pruritus and a burning sensation and may be complicated by blister formation or ulceration. Purpuric lesions are not uncommon. In most patients, the condition remits during summer but often recurs during the winter months. Lesions usually resolve spontaneously within three to five weeks leaving minor residual postinflammatory hyperpigmentation.

Horse-riding enthusiasts who wear tight clothing in cold weather may develop similar lesions on the thighs. This disease is associated with panniculitis, i.e. inflammation of subcutaneous tissue, and has been termed 'equestrian cold panniculitis'. Patients present with

Figure 1: Erythematous-violaceous patches:

Figure 1 (a): On the dorsal aspects of the fingers



Figure 1 (b): On the palms and fingers



doughy subcutaneous swellings on the thighs.

Perniosis may be accompanied by other clinical manifestations of cold sensitivity, such as acrocyanosis and/or erythrocyanosis.

Acrocyanosis

This type of 'poor circulation', often familial, is more common in females than males. The hands, feet, nose, ears and cheeks become blue-red and cold. The

palms are often cold and clammy. The condition is caused by arteriolar constriction, dilatation of the subpapillary venous plexus, and increases in blood viscosity induced by the cold temperatures.

Erythrocyanosis

Erythrocyanosis occurs in obese, often young, women. Purple-red mottled discoloration is seen on the buttocks, thighs and lower legs. Cold provokes the condition and causes an unpleasant burning sensation.

Diagnosis

The diagnosis of perniosis is usually based on clinical grounds. A skin biopsy may be helpful in cases where the diagnosis is in doubt. The four characteristic findings are scattered necrosis of individual keratinocytes, marked subepidermal oedema, perivascular lymphocytes and lymphocytic vasculitis. Perniosis may be difficult to distinguish from lupus erythematosus. Spongiosis and periecrine lymphocytes are more common in perniosis. In lupus erythematosus vacuolation of the basal layer is more common, mucin is usually increased in the dermis and the lupus band test (that is IgG at the dermo-epidermal junction) is positive. The presence of antinuclear antibodies favours a diagnosis of lupus erythematosus.

Treatment

First Steps

The therapeutic strategy comprises avoiding chronic exposure to cold temperatures and employing therapeutic agents that increase peripheral circulation.

The calcium channel blocker, nifedipine (10 mg three times daily or 20 mg twice daily) is very effective both in increasing the rate of resolution of pernio lesions and in preventing their appearance. This efficacy is due to the vasodilatory effect of this medication. While mild symptoms of peripheral oedema, flushing, headaches and hypotension may occur, these symptoms rarely require discontinuation of the medication when used in low doses. Blood pressure should be monitored at the start of treatment and at return visits.

Ancillary Steps

Keeping both the affected extremities and the core body dry and warm are essential in preventing pernio. Patients should wear thick socks and shoes. For

perniosis of the hands, gloves are recommended. Clothing should be loose-fitting. The ambient temperature should be warm. These environmental changes are most critical in preventing recurrences. Feet should always be kept dry as moisture enhances cold injury.

Alternative Steps

1. Nicotinamide (500 mg three times daily) may be useful alone or in addition to calcium channel blockers. Flushing and palpitation are problematic.
2. Hexylnicotinate (2% cream applied three times daily) may be useful for patients who are, intolerant of, or unwilling to take oral medication.
3. Amlodipine, (2.5-5 mg once daily) may be used as an alternative to calcium channel blockers. The long half-life and consequent once daily dosing of this drug are beneficial when chronic therapy is required.
4. Erythema doses of ultraviolet light (UVB) to affected areas 2 to 3 times a week at the start of winter may prevent the development of lesions.
5. Sympathectomy may be advised in severe cases.

Pitfalls

1. Pernio-like lesions occur in both discoid and systemic lupus erythematosus, as well as in sarcoidosis. The possibility of lupus should be excluded by appropriate laboratory tests and biopsy. Lupus pernio is a confusing term; it is related to neither pernio nor to lupus erythematosus, but is a particular manifestation of sarcoidosis.
2. Cryoglobulinaemia, cryofibrinogenemia and other hypercoagulable states should be considered, especially in atypical or refractory cases. Most cases of classic perniosis are not associated with such conditions but are environmentally triggered. 🙋

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