Perniosis (Chilblain): Report of three cases

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ABSTRACT

Perniosis (chilblain) is inflammatory cutaneous lesions, located on acral surfaces (fingers, toes, nose, auricula), which present in association with cold exposure. They can appear as an idiopathic (primary) dermatosis or with an underlying autoimmune disease (secondary). The primary or idiopathic form is not associated with an underlying disease and is clinically indistinguishable from the secondary form. The use of cutaneous biopsy to distinguish between both types is controversial. The secondary form is associated with an underlying condition such as connective tissue disease, monoclonal gammopathy, cryoglobulinemia, or chronic myelomonocytic leukemia. Histopathology cannot accurately help distinguish the primary from secondary forms of chilblains.

The aim of these 3 case reports was to raise awareness of this condition and to present simple recommendations and treatment for perniosis to avoid unnecessary investigation and anxiety.

KEYWORDS: Perniosis, chilblain, cold injury

INTRODUCTION

Perniosis (chilblains) is acral cutaneous eruptions which occur with exposure to cold. The disease can be classified as primary or secondary forms. The primary or idiopathic form is not related to any disease and cannot be clinically differentiated from the secondary form. The secondary form is related to an underlying clinical disorder such as connective tissue diseases, primarily systemic lupus erythematosus (SLE), monoclonal gammopathy, cryoglobulinemia or myelomonocytic leukemia. Histopathological differentiation of the two forms is difficult. [1]

Clinicians are not sufficiently knowledgeable about perniosis, which causes unnecessary hospital, laboratory and radiological costs and even the use of harmful medications. Generally these patients present with itchy and sometimes painful erythematous or blue-purple patches or plaque on acral surfaces (fingers, nose, ears) after exposure to cold. In histopathological examination, there is generally seen to be endothelial swelling and perivascular lymphocytic infiltration in the lower and upper dermal plexus together with subcutaneous fat tissue. [2] Three cases are presented here with the differential diagnosis, clinical diagnosis and treatment approach for perniosis disease, which is rarely seen in temperate climates and can be brought under control with simple treatment approaches.

CASE REPORT

Case 1:

A 43-year old construction site worker presented with complaints of swelling, bruising and pain in the hands which had been ongoing for 2 months. These complaints improved in the spring and summer months. There was no history of any known systemic disease and there was no abnormal finding in the laboratory parameters. In the physical examination, a 1x1.5cm erythematous lesion was observed on the nose and at the distal tips of both hands there was a blue-purple colour change and swelling. [Figure 1, 2]. The patient was diagnosed with primary perniosis and treatment with Nifedipin (AdalatCrono) 2 x 30mg was started.
Case 2:
A 34-year old female presented at the Physical Medicine and Rehabilitation outpatient clinic in the month of January with complaints of cold, burning, swelling and blue-purple colour changes in the ends of the fingers and toes bilaterally after going outside. In addition, there was a history of ‘white attack’ not in the winter months which had been evaluated as Raynaud’s phenomenon in the hands.

The patient history revealed that when going out in cold weather, several times redness had developed in the neck and facial area, which had recovered with antihistamines. The patient had a history of Hashimoto thyroid disease for many years. In the physical examination, blue-white colour changes and swelling were observed in both hands. [Figure 3].

In the laboratory evaluation, values were determined as sedimentation rate: 37 mm/hr, C-reactive protein: 16 mg/dl (0-3), rheumatoid factor (<20.0 IU/ml) negative, sT3:2.97pg/ml (1.8-4.5), sT4:1.35 ng/dl (0.93-1.7), TSH: 4.3 uIU/ml (0.27-4.2), ANA: 1/100 (weak positive, mottled pattern). Full blood count and routine biochemical test results were normal. The patient was evaluated as having Raynaud’s phenomenon, cold urticaria, autoimmune thyroiditis and secondary perniosis.

In accordance with the 2014 American Rheumatology Association SLE criteria (2/11 document), the patient was not evaluated with SLE. The patient was recommended to take precautions against the cold and treatment with pentoxifylline 2x400 mg po (Trental), and Diltiazem 2x60 mg po (Diltiazem) was started. After 3 weeks the symptoms had recovered.

Case 3:
A 69-year old male patient presented with complaints of bruising in the right hand and discharge on the palmar surface at the level of the 3rd finger metacarpophalangeal joint. There was no history of trauma or exposure to cold. In the physical examination, soft tissue infection at the right hand 3rd finger metacarpophalangeal joint was determined, cyanosis in the skin of the right hand, venodilatation in the whole arm and hardness in the vascular walls with palpation. [Figure 4]

There was no history of any known systemic disease. In the laboratory evaluation, sedimentation rate was 33mm/hr. In the culture taken from the lesion area, staphylococcal proliferation was determined.

On venous Doppler ultrasonography of the left upper extremity, venous insufficiency was determined. There was venous dilatation rather than peripheral vasoconstriction (acrocyanosis) which is seen in perniosis. The patient was considered to have a peripheral circulatory disorder and soft tissue infection. Treatment was started of amoxicillin clavulonate 2 x 1gr po, ciprofloxacin 2 x 500 mg po and enoxaparin 2 x 0.6ccsc daily.
DISCUSSION

Non-adaptation of the human body to cold causes a wide range of injuries. Just as this damage may be localised in one part of the body, it may also be in the form of systemic hypothermia. Perniosis generally occurs with exposure to cold in the form of localised, inflammatory, cutaneous lesions on the acral surfaces (fingers and toes, nose, ear). Patients present with itchy, painful, burning blue-purple colored acral plaque or patches on the fingers and toes. Over time the lesions become ulcerated or pustular and secondary infections may develop. This makes differential diagnosis more difficult. Lesions may occur as idiopathic (primary) or with an underlying autoimmune disease (secondary) and in cases with a similar clinical appearance with no association with cold, the term ‘perniotic-like’ is used. [2-4]

Primary perniosis is an idiopathic vasculopathy associated with the cold which affects the soft tissues of the hands and feet. The secondary form is seen together with chronic autoimmune connective tissue diseases and various forms of vasculitis. [5] Although the idiopathic form may be seen at any age, it is rarely reported in children and most patients are young and female. Relapse is often seen in the winter months. Single or multiple lesions may be seen as macular, papular or plaque, erythematous or blue-purple in colour and often in acral areas such as the hands, feet, nose and ears. Contrary to assumptions, perniosis is a frequently seen complaint. It is generally benign and not a cause of systemic symptoms. Good knowledge of the disease would prevent unnecessary tests and investigation. [6,7] The first case presented here is a typical example of primary perniosis both in the history and the examination findings.

Perniosis differential diagnosis includes SLE, cryopathy or connective tissue disease, polymyositis, septic emboli, Raynaud’s phenomenon, atheromatous emboli, sarcoid pernio, erythromalagia, vasculitis, peripheral arterial disease, cold urticaria, cutaneous leukaemia, acrocyanosis and blue finger syndrome associated with anticoagulant treatment. [8] With a detailed history, physical examination and selected laboratory tests, diseases which may be underlying the secondary form can be discounted in the differential diagnosis. As in second case, we evaluated as secondary form of perniosis. She was 34 years old and may develop any kind of connective tissue disorder elderly.

Perniosis is a complaint seen more often in the winter months. Various studies have been published in literature related to perniosis cases in different climates. In a case-control study of perniosis cases in Diyarbakır/Turkey, Akkurt et al [9] evaluated the relationship with body mass index (BMI) and concluded that a low BMI could be a risk factor increasing the development of perniosis and personal predisposition could be important in the development of lesions independently of external factors. The first two cases have normal body mass indexes.

The epidemiology and basic mechanisms of the disease are still at the hypothesis stage and there is ongoing debate concerning treatment. The disease is bilateral and is related to exposure to cold. Spontaneous recovery often occurs after the winter months then a relapse is seen the following winter. It may be seen as an occupational disease in individuals who work in a cold environment. It does not demonstrate systemic symptoms and laboratory test results are normal. Physical examination is sufficient for diagnosis. [4].
In the histopathological examination, there is lymphocytic vasculitis characterised by lymphocytic infiltration in the vascular walls. This inflammation pattern is different from other causes of perivascular lymphocytic infiltration such as SLE, erythema multiform, lymphocytic infiltration of the skin (Jessner) and medication-related eruptions. Despite this, differentiation cannot be completely made. Histopathological examination provides differentiation from cellulitis, herpes simplex infection and disseminated gonococcaemia. [10]

The application of cutaneous biopsy for differentiation of primary and secondary subtypes from each other is controversial. In a study by Boada et al [11], histopathological analyses were applied to idiopathic and autoimmune disease-related (secondary) perniosis cases and while perivascular lymphocytic infiltration, pericorneal distribution, dermal oedema and necrotic keratinocytes were found in cases of idiopathic perniosis, perivascular localized lymphocytic infiltration, vacuolisation in the basal membrane and necrotic keratinocytes without pericorneal distribution were observed in secondary cases. The conclusion was reached that the single histopathological differentiation that could be made was lymphocytic infiltration with peri-ecrine distribution.

In a study by Viguer M et al [12], in 22 of 33 patients with perniosis lesions, one or more abnormalities were determined which could suggest connective tissue disease and 8 were diagnosed with SLE. In addition, the rates of female gender and exposure to cold were found to be much higher in SLE-related perniosis cases.

Histopathologically, SLE-related perniosis cases differ from idiopathic cases in that a high rate of skin perisudoral infiltration is seen. In those showing findings of connective tissue disease, a relationship has been found between anti-nuclear antibody (ANA) positivity and positive cutaneous immunofluorescence especially in the circulation. In conclusion, it can be said that chronic perniosis cases may be related to connective tissue diseases and may develop severe SLE including autoimmune abnormalities. Therefore, these patients must have long-term follow-up. [13] The second case presented here which is evaluated as secondary perniosis, because of accompanying autoimmune diseases, primarily SLE, she will be followed up clinically in respect of other connective tissue diseases.

In a study by Crowson et al [4], skin biopsies were evaluated from 39 patients diagnosed clinically and pathologically with perniosis. In 17 of the patients, systemic or extracutaneous disease was determined such as SLE, antiphospholipid antibody, HIV disease, viral hepatitis, rheumatoid arthritis, cryoglobulinemia, hypergamaglobulinemia, iritis and Crohn’s disease. In the other 21 patients, while other serological markers were negative and there were no symptoms or findings related to specific systemic disease, primary (idiopathic) perniosis was diagnosed by determining Raynaud’s phenomenon in the majority of those with positive ANA, and arthralgia atopy or family history of other connective tissue disease in the small joints. [4]

An inexperienced clinician may confuse the pustular and ulcerated erythematous lesions of perniosis disease on the hands and feet with MRSA infection and incorrect treatment may be given. There is a case report in literature of a young adult female who developed redness in the feet the day after a pedicure, which was thought to be MRSA infection, but despite a long period of antibiotic therapy, the lesions did not recover. It was concluded that the toes remaining for long period in a damp environment during the pedicure was a risk factor for the development of perniosis. [14-17] In the 3rd case presented in this paper, the cyanotic, white-blue appearance of the right hand could on first impression be thought to be perniosis, but as it was asymmetric, there was no history of cold exposure and venous dilatation, this diagnosis was not considered. In addition, it must not be forgotten that after itching and trauma in perniosis lesions, secondary infections may develop. So, we should take into consideration the infectious diseases in differential diagnosis of perniosis.

Treatment for perniosis includes warm clothing as protection in cold weather, not staying immobile, appropriate nutrition, exercise and not staying in cold and damp environments. Nicotine and alcohol use should also be reduced. Nifedipin or amlodipin are used as medical treatment. These medications increase digital blood flow, shorten the recovery time and reduce pain and irritation. In the past, ultraviolet light, topical corticosteroids, calcium preparates, intramuscular vitamin injections and vasodilators have been used but could not achieve sufficient treatment. [18-20] There are also reports which have claimed that thyrocacitin and haemodilution could be useful in very serious cases. [4]

CONCLUSION

Perniosis is the term given to painful or itchy, erythematous or violet-coloured acral papular or nodular lesions which develop associated with cold. Although perniosis is a rarely seen clinical event in temperate climates, it may be encountered in the cold months. Therefore, clinicians in these climatic regions may not be familiar with this disease. The disease may be in the primary form or the secondary form may develop related to underlying disease. Therefore, perniosis cases showing a chronic course may develop connective tissue diseases (SLE) and autoimmune abnormalities. These patients should be followed up long-term. Treatment of the disease is simple and when clinicians have good knowledge of the clinical table, unnecessary testing is avoided.

REFERENCES


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