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Atrophie Blanche - A Special Form Of Vasculopathy

C Y K Kwok, W K Cheong

ABSTRACT

Although atrophie blanche is classified as a vasculitis, its presentation and management differ from the classical vasculitis. We present a patient with idiopathic atrophie blanche and discuss its management.

Keywords: atrophie blanche, vasculitis, anti-thrombotics, pentoxifylline

INTRODUCTION

The clinical hallmark of atrophie blanche is the presence of ivory white atrophic scars on the extremities resulting from an underlying vasculopathy. Early lesions take the form of characteristic purplish papules and plaques.

We report a patient with idiopathic atrophie blanche and discuss the current literature reviews on the pathogenesis and treatment of this chronic disorder.

CASE REPORT

A 42-year-old Chinese housewife presented with a 10-year history of recurrent painful ulcers on both legs. These were precipitated by minor trauma and took up to 6 months to heal. Early lesions took the form of small purplish papules, which later turned haemorrhagic before ulcerating. Subsequent healing resulted in depressed white scars.

She was recently diagnosed to have non-insulin dependent diabetes mellitus with no systemic complication.

On examination, there was a 5 x 4 cm serpiginous ulcer with a necrotic purplish edge and sloughy base on the right foot and a 2 x 2 cm healing ulcer on the left foot.

The surrounding skin was also abnormal, with polymorphic lesions consisting of areas of erythematous and purpuric papules, telangiectasia, hyperpigmentation and white atrophic scars.

Investigations were carried out to elicit any underlying cause for the ulcers. The full blood count was normal (Hb 14.0 g/dL TWC 12,700/uL platelet 237,000/uL ESR 9 mm/hr). So was the connective tissue (ANA, ANCA, RF, anti-cardiolipin antibody, urinalysis - negative) and haematological screen (serum cryoglobulin - negative)

The wound culture however grew *Klebsiella* species and *Pseudomonas aeruginosa*. The random glucose was 17.3 mmol/L. This was later normalised with oral diabetic medication. Punch biopsy of an early papule was taken. The primary pathology appeared to be fibrin clogging of the dermal blood vessel lumina. There were also fibrinoid deposits in the vessel wall. In addition, some perivascular chronic inflammation and fibrosis were noted. The epidermis was atrophied. The histological findings fitted that of atrophie blanche.

The final diagnosis was:
1. Idiopathic atrophie blanche
2. Diabetes mellitus

The patient was prescribed a combination of dipyridamole and pentoxifylline. Low dose aspirin was initially prescribed but was later withdrawn after she developed an adverse drug reaction to it.

The secondary wound infection was vigorously treated with a course of antibiotics as well as meticulous wound dressing. The ulcers responded slowly to the therapy, accompanied by symptomatic improvement.

DISCUSSION

Atrophie blanche was first described by Milan in 1929[1]. It is derived from French, meaning white atrophy.

One can classify atrophie blanche into primary and secondary types[2]. In the latter, diseases that are associated include chronic venous insufficiency, atherosclerosis, connective tissue disorders (eg SLE, rheumatoid arthritis), and dysproteinemias (eg cryoglobulinemia, macroglobulinemia).

Idiopathic atrophie blanche is a chronic cutaneous disorder of young to middle-aged women characterised by persistent painful leg ulcers. It begins as purpuric infiltrated papules or plaques that undergo superficial ulceration which heals with white atrophic scars. There is usually associated telangiectasia and hyperpigmentation[1,3].

There are two schools of thought regarding the pathogenesis of atrophie blanche[4]. The first school considers atrophie blanche to be a primary vasculitic disorder. It is hypothesized that circulating immune complexes are deposited into vessel wall resulting in activation of complements, chemo-attraction of neutrophils and deposition of fibrin. Release of

National Skin Centre
1 Mandalay Road
Singapore 308205

C Y K Kwok, MBBS, MRCP (UK)
Registrar

W K Cheong, MBBS, MRCP (UK), Dip Ven (Lond)
Consultant Dermatologist

Correspondence to:
Dr C Y K Kwok

lysosomal enzymes subsequently lead to secondary vascular damage and inflammatory tissue destruction[4].

The second school of thought is the current favoured hypothesis. It considers idiopathic atrophie blanche to result from a primary coagulopathy. This is supported by the fact that fibrins deposit within vessels is the earliest pathogenic change. The paucity of neutrophils in the wall of blood vessels argues against it being a primary vasculitic disorder[2,4].

Although cutaneous vasculitis and idiopathic atrophie blanche may present with similar clinical picture of recurrent lower leg ulcerations, it is important to distinguish one from the other as treatment for each differs. Systemic corticosteroids, which may be used for recalcitrant cutaneous vasculitis, are ineffective in atrophie blanche. Besides, the mistaken prolonged use of corticosteroids in atrophie blanche may result in many adverse effects such as osteoporosis, Cushing's syndrome, hypertension and glucose intolerance.

Systemic therapy for idiopathic atrophie blanche is geared towards correcting the coagulopathic state. To this aim, the anti-thrombotics and pentoxifylline have met with moderate success and are the currently favoured drugs. Improvement is usually seen within one month of therapy.

Low dose aspirin and dipyridamole are the most common anti-thrombotics used. Aspirin acts by inhibiting the formation of thromboxane A_2, a platelet aggregator. Dipyridamole inhibits phosphodiesterase which leads to increased intracellular cAMP, thereby inhibiting platelet aggregation. They are generally well tolerated and have minimal side-effects[5,6].

Pentoxifylline (Trental) acts by increasing deformity of erythrocytes, thereby improving blood flow to ischaemic tissues to promote healing[7]. It however, is a comparatively costly drug.

Minidose heparin (SC heparin 5,000U 12 hourly) has also been reported to be effective. Heparin binds to anti-thrombin III which accelerates inhibition of activated clotting factors. With this therapeutic mini-dose, the partial thromboplastin time is not altered, and the risk of bleeding is minimised[8].

Phenformin and ethylestranol combination is of historical interest as the former has been withdrawn from the US market. The combination therapy act by increasing plasminogen activator in venous endothelial cells[9].

Nifedipine has been used on the premise that vasospasm may contribute to formation of fibrin by decreasing blood flow. Side-effects include erythromelalgia-like erythema and pain in the legs[10].

Sulphasalazine has also been used for treatment of atrophie blanche. It is thought to affect inflammatory cell function as well as platelet activity[11,12].

Although much has been said about chemotherapy, the local wound care and treatment of secondary infection is equally important. The use of hydrocolloid dressing may speed up the healing of ulcers. Systemic antibiotics based on wound culture sensitivity results may be required to eradicate any secondary wound infection.

CONCLUSION

Recognition of this distinct entity of atrophie blanche as a cause of chronic recurrent leg ulceration allows us to better manage this difficult condition by a two-pronged approach of meticulous wound care and attempt at correcting the underlying coagulopathy. The pitfall of using corticosteroids or even immunosuppressives to treat this condition must be avoided.

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