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Atypical lichen myxedematosus: a rare case with complete resolution of skin

Karthiga S[1]*, Nusair MSM[1], Mayurathan P[1,2]

Abstract

Lichen myxedematosus (LM) is a rare disorder classified into three categories based on the type and distribution of skin lesions: generalised (scleromyxoedema), localised, and atypical variants. Scleromyxoedema, often associated with monoclonal gammopathy, exhibits multi-organ involvement and necessitates specific treatments such as intravenous immunoglobulin. Localised LM has benign characteristics and typically does not progress to scleromyxodema. The atypical variant does not meet the criteria of the previous two categories and may resolve spontaneously. We present a case of a 45-year-old woman who manifested an erythematous papular lesion on both shins over three months, which spontaneously resolved over one month. A skin biopsy confirmed the diagnosis of Lichen myxedematosus. However, clinical features were not suggestive of either Scleromyxodema or localised LM. The Patient was diagnosed to have an atypical variant of LM because the lesion, which was confined to bilateral shins, completely resolved within a month and was not associated with paraproteinemia. This report highlights the challenges in diagnosing and managing such atypical cases.

Key words: localised lichen myxedematosus, atypical lichen myxedematosus, scleromyxoedema, subclinical hypothyroidism

Introduction

Lichen myxedematosus (LM) is a rare papular mucinosis characterised by mucin deposition and fibroblast proliferation, with an unknown aetiology. It typically affects individuals in middle age, without a clear gender predominance. The condition is categorised into three forms based on the distribution of lichenoid eruptions, as described by Rongioletti(1): generalised papular and sclerodermoid eruption (scleromyxoedema), localised papular forms, and atypical or intermediate forms.

Scleromyxoedema, also known as generalised LM, is a rare cutaneous disorder with multi-organ involvement and is consistently linked to monoclonal gammopathy.(2) The criteria for diagnosing scleromyxoedema include: generalised papular and sclerodermiform eruptions, mucin deposition, fibroblast proliferation, and fibrosis in the skin biopsy, monoclonal gammopathy, and the absence of thyroid disease.

Localised forms of LM are further classified into five types: acral persistent papular mucinosis, discrete papular LM, self healing papular mucinosis (juvenile or adult variant), cutaneous mucinosis of infancy and nodular LM. Criteria for diagnosing localised LM include papular, plaque-like, or nodular eruptions, mucin deposits, varying fibroblast proliferation, and the absence of monoclonal gammopathy or thyroid disease.

The third category, atypical or intermediate forms, includes cases that do not meet the criteria for either scleromyxoedema or localised forms. This group comprises various subtypes, such as scleromyxoedema without monoclonal gammopathy,

*Correspondence:

Karthiga Selvaratnam
University Medical Unit
Teaching Hospital Batticaloa, Sri Lanka
E-mail: karthiyaa001@gmail.com

Full list of author information is available at the end of the article

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localised forms with monoclonal gammopathy and/or systemic symptoms, localised forms with mixed features of the five subtypes, and not well-specified cases.

Generalised LM (scleromyxoedema) requires specific treatment, such as intravenous immunoglobulin, due to its association with monoclonal gammopathy and multiple organ involvement. Other treatments for scleromyxoedema may include systemic corticosteroids. In contrast, different treatment options are available including topical therapies, destructive modalities and intralesional steroids for localised LM. Some types of localised LM and atypical variants may resolve spontaneously.(2) Other treatment options include melphalan, topical steroids, and various immunosuppressive drugs.(2,3)

Case presentation

A 45-year-old woman presented with bilateral lower limb swelling and an erythematous papular rash on her shins, which had persisted for three months. The rash initially appeared on her left lower limb and subsequently spread to her right lower limb, predominantly affecting the shin area. The rash was neither painful nor pruritic, and it was accompanied by progressive lower limb swelling that worsened over time, persisting throughout the day.

The patient also reported chest pain of a pricking nature, not associated with any autonomic symptoms. She experienced orthopnoea, paroxysmal nocturnal dyspnoea, and exertional shortness of breath but denied any history of fever, cough, or sputum production. She complained of a loss of appetite but denied any weight loss. The patient denied frothy urine, haematuria, nausea, or vomiting, and her urine output remained adequate.

The patient also denied experiencing joint pain, swelling, excessive hair loss, or oral ulcers. She exhibited no symptoms like constipation, hoarseness of voice, or cold intolerance. There was no history of sore throat, chronic cough, haemoptysis, night sweats, or a contact history of tuberculosis. She denied memory impairment, limb weakness, numbness, headache, dizziness, or visual disturbances. The patient reported no recurrent infections, bleeding manifestations, high-risk sexual activity, blood transfusions, drug abuse, or long-term drug use, apart from her routine medications. She had a history of hypertension, ischaemic heart disease, and heart failure but no family history of heart disease or malignancy.

On examination, the patient was afebrile and not pale. She exhibited an erythematous papular rash on both shins, with associated non-pitting oedema. The papules measured 2-3 mm in diameter and were confined to the bilateral shin area. Skin thickening was also noted in this region (see figures 1). There was no skin thickening on the face or arms.

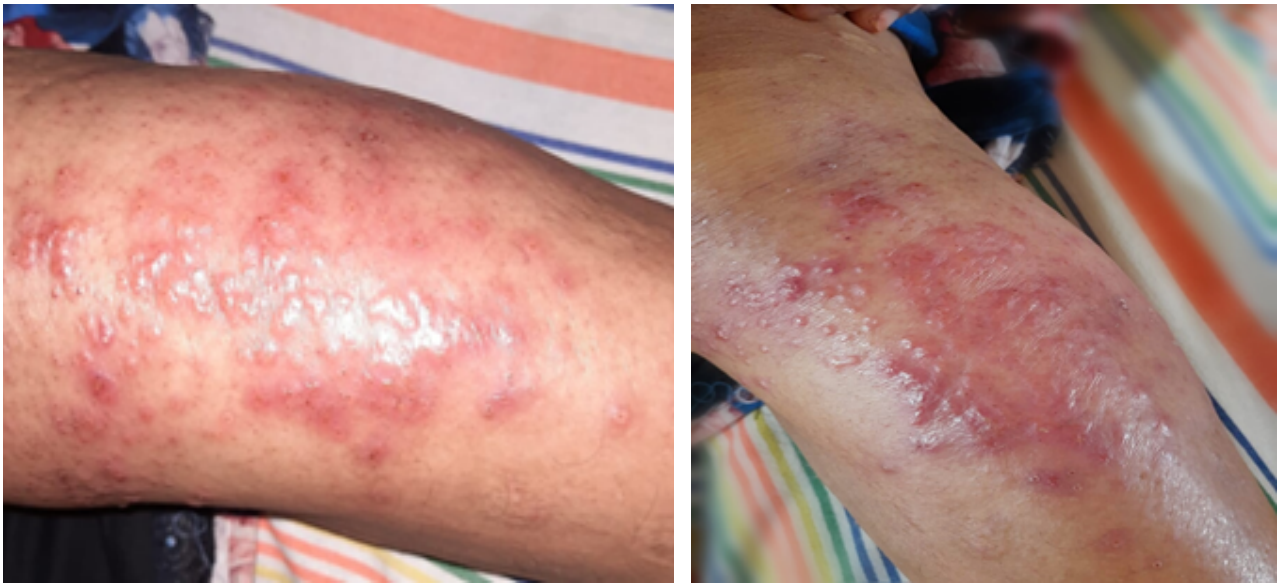

Figure 1 - Erythematous papular rash confined to bilateral shins

In terms of her cardiovascular assessment the patient had elevated blood pressure (180/110 mmHg) and an elevated jugular venous pressure. A pansystolic murmur radiating to the axilla at a mildly displaced apex, was observed. However, examinations of other systems were unremarkable.

Full blood count, liver function tests, renal function tests, and erythrocyte sedimentation rate were all within normal ranges. Chest X-ray and abdominal ultrasound scans revealed no abnormalities. Antinuclear antibody (ANA) testing returned negative results. A 2D echocardiogram revealed an ejection fraction (EF) of 35-40%, concentric left ventricular hypertrophy, and diastolic dysfunction.

The patient's thyroid-stimulating hormone was 6.60 mIU/L (normal range: 0.46-4.58), and free T4 was 14.9 pmol/L (normal range: 10.0-28.2). TSH and free T4 were repeated in 6 weeks which showed high TSH (5.83) with normal free T4. Corrected calcium was 2.6 mmol/L (normal range: 2.1-2.5), and serum magnesium was 0.8 mmol/L (normal range: 0.7-1.0). Serum protein electrophoresis indicated a chronic inflammatory process. A skeletal survey revealed no abnormalities. Repeated calcium was normal.

The patient was subsequently referred to the Dermatology clinic, where a skin biopsy was performed on the bilateral shins. The biopsy revealed mucin deposition in the superficial dermis and focal fibrosis of the dermis, supporting the diagnosis of Lichen myxedematosus (see figure 2).

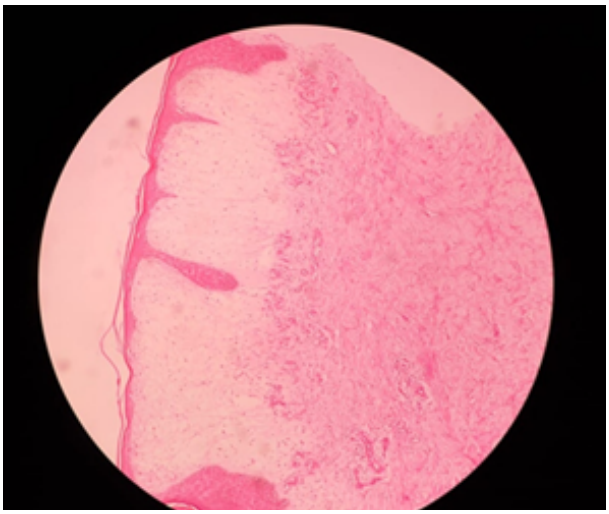

Figure 2 - Skin biopsy H & E stain ; showing mucin deposition of superficial dermis and focal fibrosis of dermis

In this case, the patient presented with an erythematous lichenoid rash localised to her shins, which resolved spontaneously. Notably, there was no evidence of monoclonal gammopathy, but she had subclinical hypothyroidism. Despite the skin biopsy favouring a diagnosis of Lichen myxedematosus, the type and distribution of the lesions in our patient did not align with either the generalised or localised forms of LM. According to Rongioletti's classification, our case was classified as an 'Atypical LM.'

Discussion

Lichen myxedematosus is a rare dermatological condition with three distinct categories based on the type and distribution of skin lesions. Localised LM is typically milder than scleromyxoedema, which is often associated with multi-organ involvement.(4) Atypical localised LM cases are reported rarely in medical literature, making their diagnosis and management challenging.

The diagnosis of LM can be particularly difficult due to its rarity and variable clinical presentations. Approximately 200 LM cases have been reported in the literature(5), with no documented cases in Sri Lanka until now.

In our case, the patient presented with subclinical hypothyroidism and a localised distribution of an erythematous rash confined to lower limbs which resolved completely. There was no association with paraproteinemia, and the skin biopsy showed mucin deposition in the superficial dermis and focal fibrosis of the dermis. Remarkably, the rash spontaneously resolved within a month without any treatment. Despite the skin biopsy suggesting lichen myxedematosus, the type and distribution of the rash, as well as the thyroid involvement in the absence of monoclonal gammopathy and complete disappearance of the lesion with time, did not align with a diagnosis of scleromyxoedema or localised LM form. Some localised forms such as self healing papular mucinosis and cutaneous mucinosis of infancy may resolve spontaneously. However, localised forms are not associated with thyroid disease. Therefore, according to Rongioletti's clinicopathologic criteria, the patient was categorised as an atypical variant of LM.

Atypical localised LM cases are extremely rare, which presents challenges in diagnosis and management. There were no specific guidelines available for the management of such cases, and our patient's lesions resolved without any treatment.

Conclusion

This case highlights the need for further research and documentation of atypical variants of LM to better understand their clinical characteristics and aid in their diagnosis and management. Although our patient's lesions resolved completely with time, future cases may not follow the same course, emphasising the importance of continued study and reporting in the field of dermatology.

Declarations

Conflicts of interest
The authors declare that they have no conflicts of interest to be addressed regarding this case report.

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Author details
[1]University Medical Unit, Teaching Hospital Batticaloa, Sri Lanka
[2]Faculty of Health care science, Eastern University of Sri Lanka

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