Between a rock and a hard place

Dermatomyositis-Morphoea overlap with diffuse dystrophic calcinosis cutis

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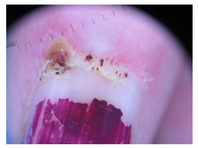
Introduction

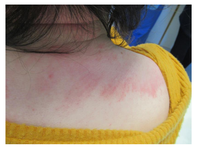
Correlation between systemic sclerosis (SSc) and dermatomyositis is well described in the literature. (1) Association between localised scleroderma (limited plaque morphoea) and dermatomyositis is however rare. We present a case report of localised scleroderma and Dermatomyositis presenting within a six-month time frame.

Report of a case

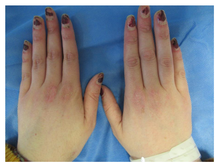
A 29-year-old female presented with a 3-week history of rash associated with itch to back and metacarpophalangeal (MCP) joints. She complained of progressive muscle weakness. History was significant for plaque morphoea, biopsy proven three months prior. Clinical examination was as follows:



Nailfold dermoscopy: Cuticle overgrowth, telangiectasia



Shawl sign affecting the neck, shoulders, upper back

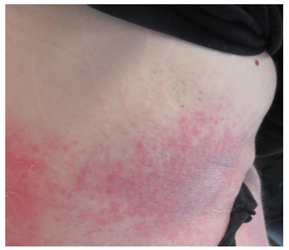


Gottron's papules at the MCP and PIP joints

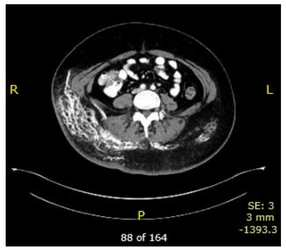
Investigation, treatment and progress

- Creatine Kinase: 8716 U/L.
- Anti-nuclear antibody: positive; 1:1280
- Myositis panel: Anti MI-2 antibody positivity.
- Anti-centromere, anti-topoisonerase-1 - negative.
- MRI: Significant muscle oedema.
- Deep muscle biopsy: perifascicular and perivascular inflammation, atrophy
- Skin biopsy: mild epidermal atrophy, scattered apoptotic keratinocytes.
- Echocardiogram, chest x ray, cardiac MRI: normal.

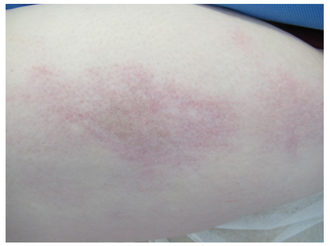
The patient was commenced on IV methylprednisolone and noted an initial improvement in rash and myositis. However, myositis and associated myopathy returned on weaning to oral steroids, and she was readmitted to hospital. IV Rituximab treatment was initiated, and oral azathioprine commenced.



New rash, flank and thighs, presenting after commencing treatment



CT revealing calcinosis corresponding to area affected by rash, subsequently biopsied



This patient had previously presented to the dermatology clinic with bilateral skin changes to the thighs. Anti-nuclear antibodies were noted at 1:1280 dilutions. Extractable nuclear antibodies (ENA), double-stranded DNA (dsDNA) and System Sclerosis profile (SSC) were negative at that time. Skin biopsy: focal collagen sclerosis in deep dermis with accompanying lymphoplasmacytic perivascular infiltrate in skin and subcutis, in keeping with localised Scleroderma. With potent topical steroids this had significantly improved prior to clinical presentation with dermatomyositis.

Despite prompt initiation of treatment, she subsequently represented to dermatology with new erythema over bilateral flanks. Incisional skin biopsy showed a deposit of calcium at the base, and subsequent computed tomography and ultrasound guided biopsy revealed widespread, calcinosis cutis, affecting the flanks and bilateral thighs. This has not progressed, ulcerated or caused symptoms. She is now improving on current treatment, and to date has not developed features of systemic sclerosis.

Discussion

Scleromyositis – an overlap between systemic sclerosis and dermatomyositis, or more commonly polymyositis, is well described. (1) Studies have shown 10-38% of patients with systemic sclerosis develop overlap syndromes i.e., features of systemic sclerosis with another connective tissue disease (2) However, on a review of published case reports, very few cases describe an overlap between localised scleroderma and dermatomyositis. (3,4) Rituximab has previously been used in treatment of scleroderma/myositis overlap to good effect, (4) and was also effective in our patient.

In cases of scleromyositis, muscle involvement has generally been described as mild, with minimal elevation in muscle enzymes, and patients generally respond well to low or moderate doses of steroid therapy. (1) In contrast our patient had significantly elevated CPK and severe myositis necessitating treatment with Rituximab, and subsequently developed widespread dystrophic calcinosis cutis despite treatment.

Widespread calcinosis cutis is common in juvenile dermatomyositis but seen more rarely in adults (6). There are no specific guidelines for treatment, and few randomised control trials exist, making treatment challenging (7). This patient has shown improvement with rituximab, which has previously been used in treatment of calcinosis cutis with success. (7)

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Literature cited

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