dosage to 200 mg/day and finally even to 250 mg/day. But we were not able to reduce the glucocorticoid dosages until immunosuppression was further intensified using i.v. immunoglobulins 20 mg/day for 3 days [6] (Supplementary Data Fig. 5).

Intensive physiotherapy, local necrosectomy (scrotal skin and epiorchium) and intensive immunosuppression finally led to the resolution of the symptoms (Supplementary Data Movie 2).

Subsequent to neurological rehabilitation the patient is currently successfully being treated with cyclophosphamide following the Austin-scheme.

Here we describe a rare case of PAN where the need for close interdisciplinary collaboration is obvious. The well-known testicular pain as a criterion for diagnosis of PAN is suggested to be explained by vasculitis and consecutive stenosis of the testicular artery and or involvement of small testicular and or epididymitic vessels causing a combination of inflammatory and ischaemic pain.

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Breast calcinosis, panniculitis and fat hypertrophy in a 35-year-old woman with dermatomyositis

Sir, We report the case of a 35-yr-old woman with a 12-yr history of dermatomyositis. She originally presented with proximal muscle weakness, fatigue, a heliotrope rash and Gottron’s papules. Investigations confirmed a raised creatine kinase and typical histological changes on needle muscle biopsy. After a reducing dose of steroids, she was maintained on azathioprine 150 mg per day, switched due to lack of efficacy to weekly oral methotrexate 3 yrs later. Four years later she developed conspicuous weakness and wasting of the left upper arm, with clinical evidence of muscle atrophy affecting the left triceps. Hydroxychloroquine was later introduced due to active skin disease (typical facial rash, Gottron’s papules and florid nail fold capillary changes) and subsequently cyclosporin, with good benefit, at a present dose of 75 mg bd.

For the last 2 yrs she has had an insidious and eventually marked asymmetry of her two arms, thought to be due to further wasting of her left triceps (Fig. 1). However, subsequent MRI imaging (Fig. 2) showed marked fat hypertrophy of her dominant right arm with no significant muscle atrophy. Biopsy of this was not performed.

One year ago she developed a right-sided breast lump and mammography was organised (Fig. 3). This showed diffuse calcification in all quadrants bilaterally. This breast calcification was not detectable clinically and there were no other areas on her body of palpable subcutaneous calcification.

Over the last year, marked loss of subcutaneous fat over both cheeks, consistent with a panniculitis (for cosmetic reasons no biopsy has been performed) (Fig 4). There has been no progression of lipo-atrophy or hypertrophy elsewhere.

Discussion

In summary, we present a patient with dermatomyositis who has developed the fascinating combination of likely panniculitis...
and subsequent fat atrophy of her face, together with marked fat hypertrophy affecting her right arm. To our knowledge this combination has not previously been described. In addition, the patient shows the interesting development of breast calcification. Chronic relapsing ‘Polydermatomyositis’ with predominant involvement of the subcutaneous fat (panniculitis) was described as early as 1924 [1]. Panniculitis has subsequently been described in a number of connective tissue diseases including dermatomyositis [2] and in systemic lupus [3]. The development of ‘lipodystrophy’ associated with panniculitis causing facial disfigurement is well-described [4]. Biopsy typically shows panniculitis with a lymphocytic infiltrate. The mechanism through which our patient developed fat hypertrophy is unclear but it could be hypothesized that this is a paradoxical response to subcutaneous fat inflammation. In this case, biopsy of both areas would have been interesting but would have been unlikely to alter management.

Soft tissue calcification normally occurs in the juvenile form of dermatomyositis [5], but breast calcification has previously been reported in the context of adult dermatomyositis. The findings are not common but when reported may manifest as extensive bizarre, dystrophic subcutaneous calcific deposits [6]. Additionally subcutaneous calcification is a key feature of the CREST variant of scleroderma.

There is an established link between dermatomyositis and malignancy and, with the added risk of immunosuppressive medication, the development of a breast lump requires appropriate investigation.

In summary, this case demonstrates an overlap of connective tissue disease with features of adult dermatomyositis, breast calcification and both fat atrophy and hypertrophy.

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