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.

The authors have declared no conflicts of interest.

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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. 1). 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...
Fig. 4. Anterior and lateral view of face showing fat atrophy.

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 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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Adalimumab-associated pulmonary fibrosis – reply

Sir, We thought that the response to our case report (by Collins et al.) about a patient with adalimumab-associated pulmonary fibrosis was interesting and raised some important points. The presumption that there was pre-existing interstitial pulmonary fibrosis (IPF) may be correct, however, in the absence of a pre-treatment high resolution CT or lung biopsy, this remains a presumption. Acute pneumonitis associated with methotrexate use tends to occur earlier in treatment than in this patient who had been taking it for 2 yrs. The patient concerned certainly was asymptomatic prior to treatment and deteriorated rapidly within 10 weeks of starting adalimumab. Whether fibrosis was pre-existent or not, there is a temporal relationship in this case between initiation of anti-TNF therapy and rapid deterioration in lung function. This suggests a cause and effect relationship, and certainly does not change the level of concern.

Reduction in gas transfer is a more sensitive test than a chest radiograph in screening for IPF but this test is not specific and generates false negatives. However, in the light of this case and others, there may be a case for gas transfer measurements as part of the screening process.

We believe that above all there is accumulating evidence suggesting increased caution when commencing these drugs. Most importantly, however, there should also be increased awareness and dissemination of information to general medical physicians of the potential side effects that these drugs can cause.

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Reply: Adalimumab-associated pulmonary fibrosis

Sir, We enjoyed reading the letter from Huggett and Armstrong in the October issue of the journal [1], and agree that their case report adds to the mounting body of evidence that suggests that most, if not all, anti-tumour necrosis factor (TNF) drugs may precipitate sudden, severe and often fatal respiratory failure in patients with rheumatoid arthritis (RA). However, we believe that it also highlights the importance of understanding which patients are at greatest risk of this development. The case described [1] had a normal chest radiograph prior to commencing adalimumab but a CT scan <3 months into treatment showed clear evidence of established interstitial pulmonary fibrosis with honeycombing in both lung bases. This pattern is typical for interstitial pulmonary fibrosis (IPF) complicating RA itself [2] and is most unlikely to have developed exclusively over the 2 weeks of symptomatic dyspnoea prior to presentation. This strongly suggests that pulmonary disease was established prior to the commencement of anti-TNF therapy.

The authors have declared no conflicts of interest.

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