Devastating calcinosis in a patient with adult onset myositis

A 62-year-old Indian woman presented initially in 1993 with swallowing difficulty, limb weakness, bilateral ptosis and Raynaud's phenomenon. Investigation revealed an elevated creatine kinase and muscle biopsy suggested polymyositis. She responded well to treatment with prednisolone and azathioprine, which was gradually reduced and finally discontinued in 2004. She re-presented acutely unwell in 2006 with weight loss, dysphagia and gross weakness. Examination revealed cutaneous calcinotic plaques in both thighs and buttocks (fig 1), bilateral ptosis and global limb weakness. She was unable to stand erect, walking with a bent spine (fig 2A).

Serum creatine kinase was within normal limits (61 U/l), erythrocyte sedimentation rate was 67 mm/h, haemoglobin was 8.9 g/dl (normochromic normocytic) and albumin was 12 g/l. Autoantibodies revealed positive antinuclear antibodies (titre 1:160–640), double stranded DNA 108 IU/ml (normal range 0–30 IU/ml) and negative scl-70. EMG demonstrated low amplitude polyphasic units. MRI of the thighs showed high signal within all muscle groups compatible with active myositis. An area of calcification was found on plain x ray and video swallow anterior to the C4 and C5 vertebrae, causing significant oesophageal compression (fig 3). A moderate pericardial effusion was detected on transthoracic echocardiography. A repeat biopsy from the vastus lateralis of the quadriceps muscle revealed multifocal mononuclear inflammatory cell infiltrates and perifascicular atrophy compatible with a diagnosis of dermatomyositis (fig 4A). Immunohistochemistry showed overexpression of HLA class I antigens on all fibres, confirming the diagnosis (fig 4B).

Figure 1 Soft tissue calcification in the right thigh. Informed consent was obtained for publication of this figure.

Feeding was initiated through a percutaneous gastrostomy tube. However, the patient continued to deteriorate and 4 weeks later treatment was initiated with prednisolone 60 mg/day and ciclosporin 4 g/kg, which resulted in a rapid clinical improvement. She became self-caring and was able to walk upright (fig 2B).

Soft tissue calcification is well recognised in scleroderma,¹ systemic lupus erythematosus² and childhood dermatomyositis,³ where it is present in 40% of patients. It is extremely uncommon in adult onset dermatomyositis and does not seem to have been reported as a con-

Figure 2 (A) Patient's walking posture on admission. (B) Patient's walking posture 8 weeks later. Informed consent was obtained for publication of this figure.

tributory cause of dysphagia. The mechanism of tissue calcification is not well understood but is associated with delayed or inadequate treatment, and is typically found in periarticular tissues around the elbows, knees and in the buttocks. Complications include skin ulceration, joint dysfunction, secondary

Figure 3 Lateral x ray of the cervical spine, demonstrating a prevertebral soft tissue calcified deposit with oesophageal displacement.

Figure 4 Right quadriceps muscle biopsy showing (A) multifocal mononuclear inflammation and perifascicular atrophy (haematoxylin-eosin). (B) Strong HLA-ABC overexpression by immunohistochemistry.

infection and a local cutaneous inflammatory reaction.

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