Unusual and memorable

Series editor: Gary D Wright

A 38 year old Arab woman presented recently to the hospital with cellulitis of the left hand and ischaemia of the right second toe. Multiple, coarse, and firm cutaneous nodules in various sites of the body were also noted in the examination. She had been diagnosed with systemic lupus erythematosus/myositis overlap syndrome for approximately 10 years and had not been aware of telangiectasis, sclerodactyly or dysphagia. She has had Raynaud's phenomenon for years, but it has recently improved. Her nodules began to develop some four to five years ago. Later, they progressed to involve the mandible, hands, arms, axilla, chest, and abdominal wall (fig 1) and the legs.

Radiologically, the abdominal wall was the site most heavily involved by these lesions where they appeared as tumoral-like calcinosis (fig 2). The feet were spared.

Blood test including creatine kinase and lactate dehydrogenase, and parathormone assay, were of normal values. Anti-nuclear factor was positive. Anti-U1 ribonucleoprotein and Jo-1 antibodies were negative. Anti-centromere antibodies repeatedly tested positive. These antibodies are known to be associated with calcinosis in systemic sclerosis and CREST syndrome. None the less, both of the latter conditions were not manifested by our patient at that time.

It is rather unusual to find such a widely spread lesion with extensive involvement of the abdominal wall in an adult with such a syndrome. Moreover, the upper limbs are the sites most frequently affected by soft tissue calcinosis in connective tissue diseases. From the therapeutic point of view, both cellulitis and microvasculopathy were managed appropriately and calcinosis was treated with diltiazem, 60 mg thrice daily. An anecdotal success has been reported with diltiazem in inducing remission of calcinosis in scleroderma¹ and CREST syndrome.² The response in this case needs, however, to be closely observed in the follow up.

Figure 1

Figure 2

1 Dolan AL, Kassimos D, Gibson T, Kingsley GH. Diltiazem induces remission of calcinosis in scleroderma. *Br J Rheumatol* 1995;34:576-78.

2 Farah MJ, Palmieri GMA, Sebes JI. The effect of diltiazem on calcinosis in a patient with the CREST syndrome. *Arthritis Rheum* 1990;33:1287-93.

Contributors: HAIDER M AL ATTIA, AL NADHIR M SHERIF, RIMA N EL ABASSI *Department of Internal Medicine and Orthopaedics, Mafraq Hospital, Abu Dhabi, United Arab Emirates*