Tumoral calcinosis in systemic sclerosis

Sirs,

Systemic sclerosis (SSc) is a systemic inflammatory disorder affecting many organs, particularly the lungs, the gastrointestinal tract and the skin (1). Calcinosi is also a frequent manifestation in SSc, being encountered in 8 to 40% of patients (2-4). While calcinosi most often involve the hands (85% of cases), it may also affect other troublesome sites, including periarticular areas (3,4). Moreover, although calcinosi may vary in size, it rarely leads to tumoral masses (5). We recently observed a case of particular interest, as the patient with limited SSc (lcSSc) developed a pseudo-tumor involving the anterior part of the left shoulder related to tumoral calcinosi.

A 61-year-old woman was diagnosed with lcSSc in 1995. Systemic manifestations of lcSSc included: 1) Raynaud’s phenomenon associated with telangiectasia, pitting scars and cutis calcinosi involving the hands; 2) esophageal impairment characterized by the absence of peristalsis in the lower two-thirds of the esophageal body and low pressure in the lower esophageal sphincter; and 3) interstitial lung disease. Autoantibody screening tests were positive for antinuclear (1:1000) and anticientromere antibodies. Naiifold capillaroscopy revealed enlarged capillary loops. The patient received combination treatment with diltiazem, cisapride and omeprazole. In July 2001, she presented with a painful tumoral mass on the anterior part of the left shoulder, which had developed over 5 months and was exquisitely tender to gentle palpation (Fig.1a). Laboratory data included: ESR 12 mm/ hour, hematocrit 8.5 mmol/L, white blood cell count 5.9 x 10^9/L, and platelet count 259 x 10^9/mm^3. Results of renal and liver tests, blood protein electrophoresis, serum calcium and phosphorous levels were within normal limits. Bone radiographs revealed marked conglomerate of soft tissue calcifications along the anterior part of the left shoulder (Fig.1b), and CT-scan demonstrated calcific deposits with a focal cortical clavicle erosion underlying the extensive calcifications (Fig.1c). Because the patient exhibited both discomfort and stiffness caused by the location of tumoral calcinosi, local excision surgery of the mass (composed of hydroxyapatite crystals) was made with a favorable outcome.

Calcinosi is a frequent complication, occurring mainly during the course of lcSSc with anticientromere antibodies (2,4). It usually remains asymptomatic, calcific deposits being detected by systematic bone radiographs showing linear and punctuate patterns of a few millimeters (2). In contrast, calcinosi mimicking a tumoral process is uncommon, occurring in less than 1% of SSc patients (5). In this instance, our patient presented with tumoral calcinosi involving the anterior part of her left shoulder, which was a marked cause of disability compared with the other components of lcSSc. CT-scan was further useful in accurately revealing the detailed anatomic extent of calcinosi and underlying bone clavicle erosion. Because of both severe discomfort and decreased dignity related to the location of the tumoral calcinosi, our patient underwent surgical excision of the lesion, as suggested previously (3).

Finally, although tumoral calcinosi is uncommon in SSc, our findings suggest that when this type of complication is noted, an evaluation for systemic sclerosis, including a clinical evaluation, a search for antinuclear antibodies and nailfold capillaroscopy should be systematically performed.

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References

Letters to the Editor

(a) Voluminous pseudo-tumor along the anterior part of the left shoulder. (b) Bone radiographs revealed marked soft tissue calcifications conglomering along the anterior part of the left shoulder. (c) CT-scan demonstrated tumoral calcific deposits in the left shoulder area with a focal cortical clavicle erosion underlying the extensive calcifications.

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