Tumoral calcinosis in a rheumatoid arthritis patient – a rare case report

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INTRODUCTION

Tumoral calcinosis (TC) is a rare clinical entity characterized by calcium deposition in the soft tissues around joints. The deposits are grouped into lobulated masses constituted by calcium hydroxyapatite, calcium carbonate and calcium phosphate, and are usually painless with an insidious growth over the years[1].

TC typically affects the shoulders, elbows and hips, but may also be present in other locations such as the metacarpals/metatarsals bones. It can be classified as primary (rare familial forms of hyperphosphatemic or normophosphatemic TC) or secondary (mainly associated to chronic renal failure with secondary or tertiary hyperparathyroidism). Few cases of association with rheumatoid arthritis (RA) have been described, and their pathophysiology remains controversial[2-4]. High values of calcium, phosphorus and parathyroid hormone may reflect the underlying pathophysiological processes. The diagnosis is based mainly on imaging where a typical appearance of amorphous, multilobulated and cystic calcifications with a periarticular location is observed.

CASE REPORT

A 78-year-old female was referred to our rheumatology clinic with a one-month history of inflammatory arthralgia affecting hands, wrists, shoulders and knees. She had a background history of osteoarthritis, hypothyroidism, obesity and left nephrectomy due to infectious complications in 1970. In addition, in the past two to three years she had been complaining of mechanical left shoulder pain, treated with paracetamol. She denied previous history of Raynaud's phenomenon or cutaneous lesions. Physical examination revealed symmetric polyarthritis of the metacarpophalangeal and proximal interphalangeal joints of the hands, wrists and knees, and soft tissue swelling of the left shoulder. Blood tests showed erythrocyte sedimentation rate of 96 mm/1st h, C-reactive protein of 6.28 mg/dL, rheumatoid factor present in the serum (119.3 UI/mL), negative anti-cyclic citrullinated peptide, and antinuclear antibody (ANA) of 1/160 (fine speckled) but with negative extractable nuclear antigens (ENA) and dsDNA double stranded DNA (dsDNA). Radiographic assessment of hands showed soft tissue swelling of the joints with space narrowing and erosion on the right ulnar styloid process, as well as the presence of periarticular amorphous calcifications in both hands (Figure 1). In addition, chest radiography revealed massive periarticular calcifications on the left shoulder (Figure 2). Ultrasound identified synovitis of the metacarpophalangeal joints, with presence of power-Doppler, and synovitis of the wrists (Figure 3). A left shoulder Computed Tomography showed a cluster of multiple calcifications, mostly with nodular morphology, extending approximately 8 cm longitudinally and 6 cm transversely, mainly located in the anterior region (Figure 4). Further investigation to the phosphocalcic metabolism and renal function did not find relevant changes (serum creatinine 0.86 mg/dL [0.5-0.9], calcium 9.9 mg/dL [8.6-10.2], phosphate 3.8 mg/dL [2.5--4.5], 25-OH vitamin D 37.0 ng/mL [≥30] and parathyroid hormone 37.4 pg/mL [14-72]). The diagnosis of TC in a patient with new-onset RA was established.

DISCUSSION / CONCLUSION

TC is an uncommon disease and to our knowledge only

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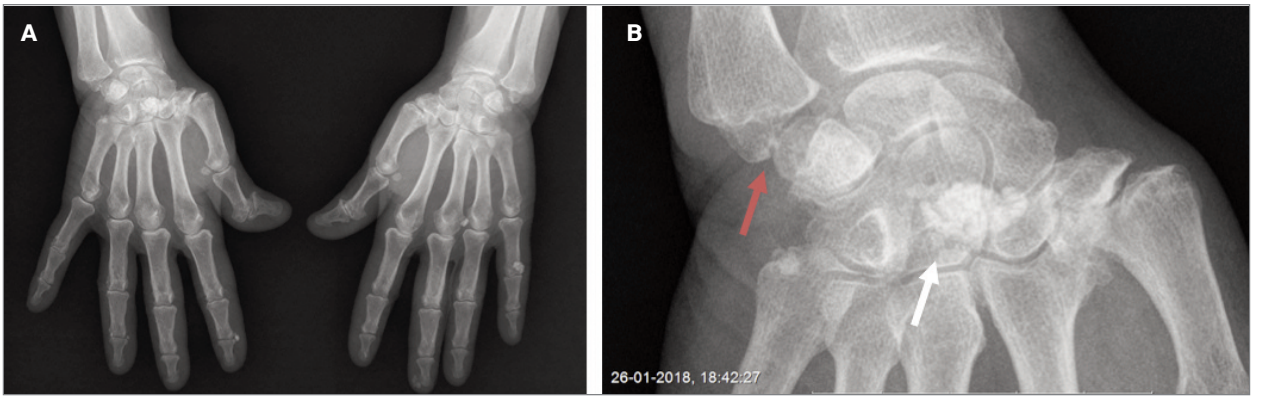

FIGURE 1. Radiography of the hands: A) Posteroanterior view of both hands showing multiple calcifications: right second finger - adjacent to the distal interphalangeal joint; right fifth metacarpal - adjacent to the base; right carpus - on the capitate, trapezoid and scaphoid bones; left third finger - on the finger pad and near the metacarpal head; left fifth finger - near the proximal interphalangeal joint; and left carpus - near the triquetrum and ulnar styloid. B) Right carpus with the presence of amorphous calcifications projected on the capitate, trapezoid and scaphoid bones (white arrow) and erosion of the right ulnar styloid process (red arrow).

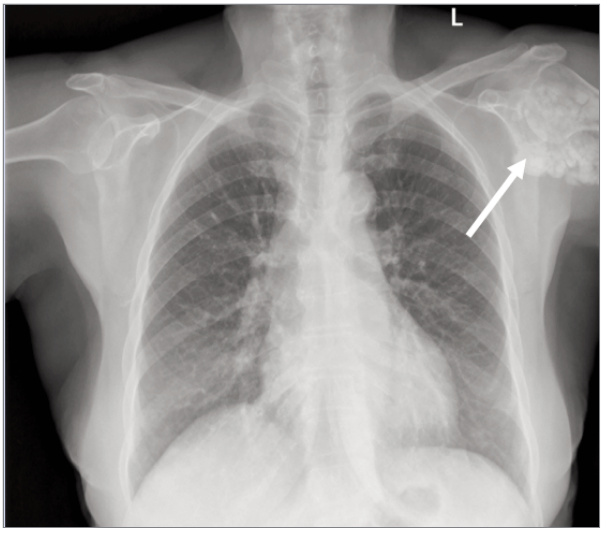

FIGURE 2. Chest radiography revealing massive periaricular calcifications on the left shoulder (arrow)

three cases of association with RA have been described[2-4]. Unlike other cases, our patient seemed to have developed RA two to three years after presenting shoulder complaints later recognized as being related to TC. Therefore, RA could not have acted as an inflammatory stimulus to promote TC, highlighting that the underlying pathophysiological mechanism remains to be clarified. In addition, no secondary cause related to abnormalities in the phosphocalcic metabolism was identified in this case. The authors draw attention to this rare clinical and imaging challenge that may occur in patients with RA.

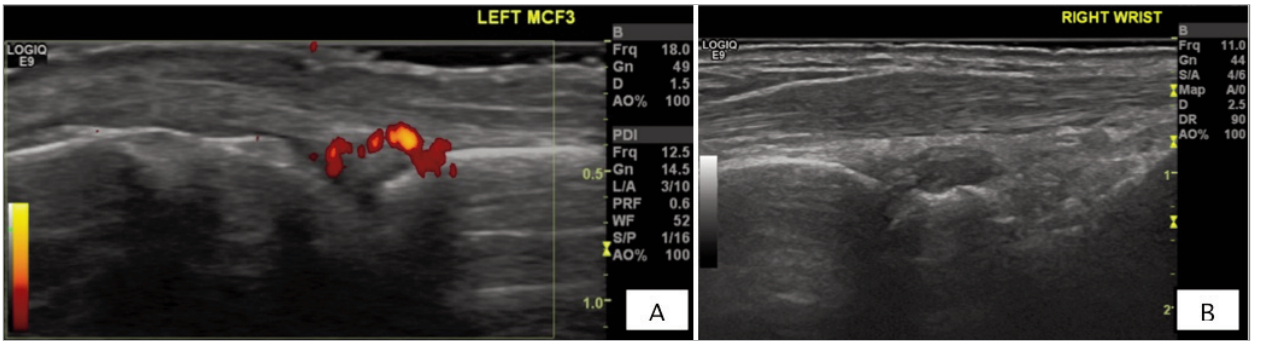

FIGURE 3. Ultrasound of the hands and writs: A) Moderate synovitis of the 3rd metacarpophalangeal joint with the presence of power Doppler. B) Mild synovitis of the right radiocarpal joint.

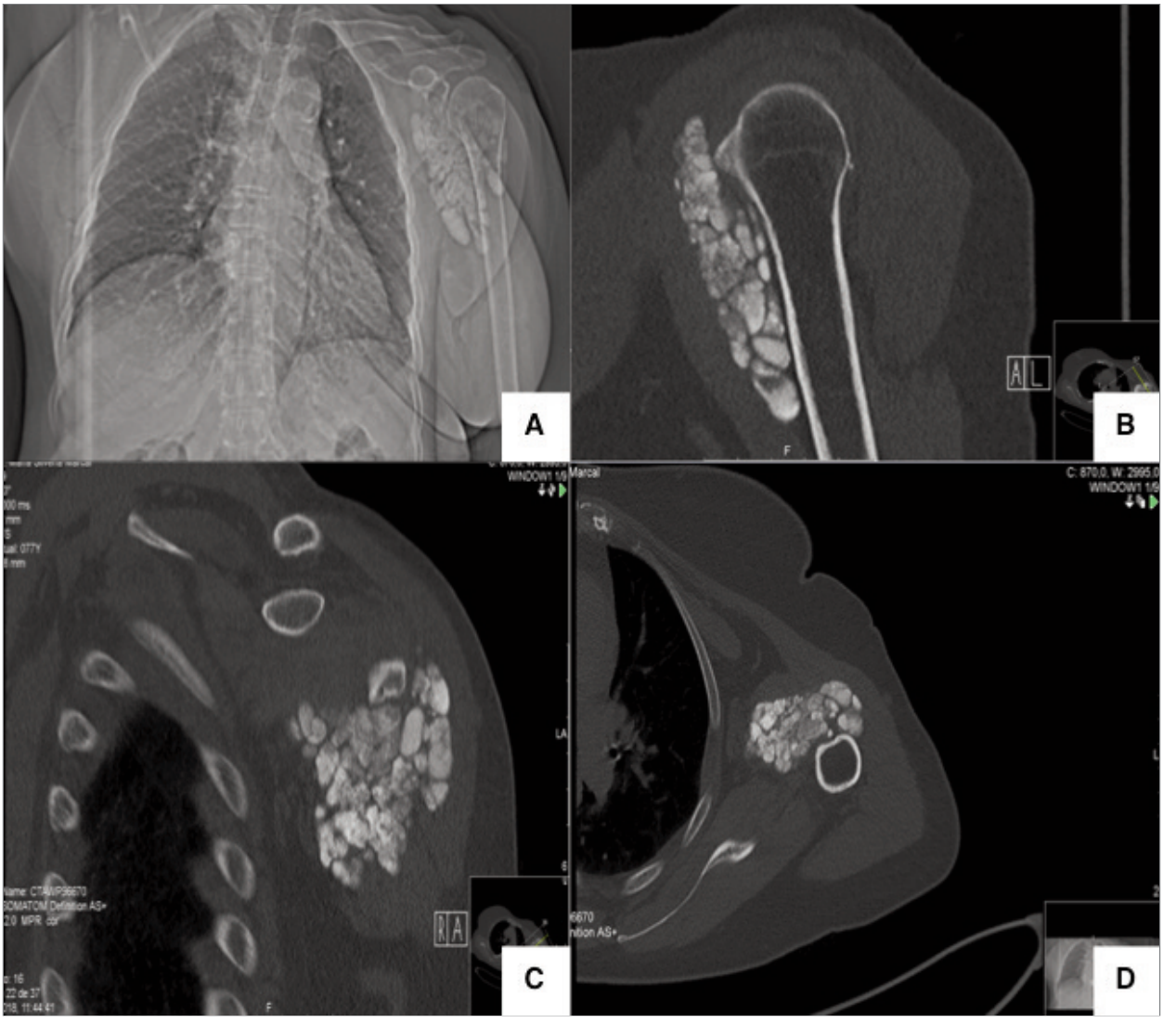

FIGURE 4. Computed Tomography of the left shoulder showing tumoral calcinosis in different planes: A) Reference topography; B) Image in the oblique sagittal plane of the left shoulder; C) An oblique coronal plane image of the left shoulder; D) Image in transverse plane.

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