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Big Gain, No Pain: Tumoral Calcinosis

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PRESENTATION

A patient's odd physical presentation was the result of what could be termed a mineral deposit. The 40-year-old man reported that his right shoulder had been swelling progressively for more than 6 months. His medical history was significant for chronic kidney failure, and he had been undergoing hemodialysis for the previous 10 years. He had no other systemic diseases, such as diabetes mellitus, hypertension, rheumatoid arthritis, or gout. Further, he had no history of trauma.

ASSESSMENT

During the physical examination a firm mass was noted at the patient's anterolateral shoulder (**Figure 1**). Yet he had no erythema or tenderness (**Figure 1**). His shoulder motions were free, and he had no motor deficits or sensory impairments.

Ultrasonography revealed multiple cysts with homogeneous echoes and scattered irregular hyperechoic plaques (**Figure 2A**). Upon compression and decompression, free particles within the cysts were easily displaced and sent whirling. Ultrasound-guided aspiration was performed, and 38 mL of milky fluid was aspirated (**Figure 2**). Fluid analysis revealed no leukocytosis or crystals.

The patient's medical records indicated that bilateral extensive calcifications, including small calcifications around the right shoulder, had been seen on a chest radiograph 1 year before this episode (**Figure 3**). The chest radiograph in **Figure 3** was taken 2 days before the ultrasonography and revealed calcifications around bilateral shoulders, which were larger than the calcifications noted 1 year ago. Three years earlier, a chest radiograph showed almost no calcification.

Computed tomography (CT), ordered after the ultrasonography, demonstrated bilateral multiple amorphous calcifications, some with fluid–fluid levels, around the glenohumeral joint (**Figure 4**). Laboratory tests yielded the following results: serum calcium level, 9.08 mg/dL (normal, 8.6-10.3 mg/dL); serum phosphate level, 6.6 mg/dL (normal, 2.5-5 mg/dL); alkaline phosphatase level, 360 U/L (normal, 34-104 U/L); and parathyroid hormone level, 1339 pg/mL (normal, 12-65 pg/mL). An ultrasound image of the parathyroid gland disclosed a growth that was suspected to be an adenoma.

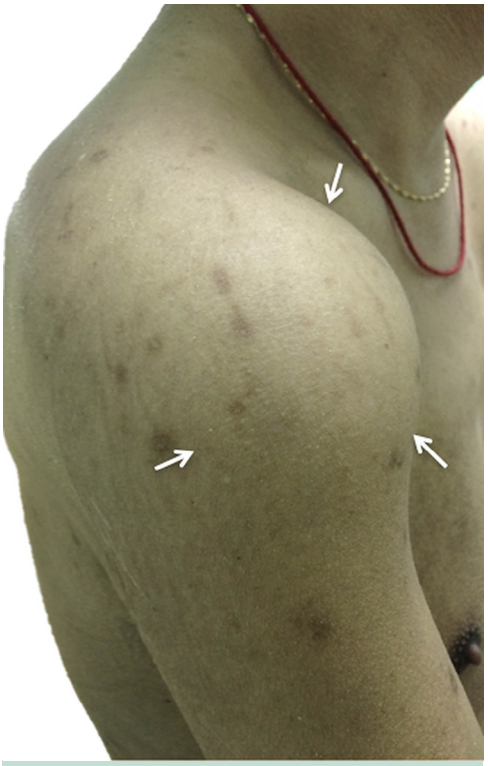

Figure 1 Arrows delineate a swollen, firm mass over the patient's anterolateral right shoulder. This was not associated with redness or tenderness.

DIAGNOSIS

The ultrasound images, combined with the evidence of hyperparathyroidism and the patient's history of chronic renal

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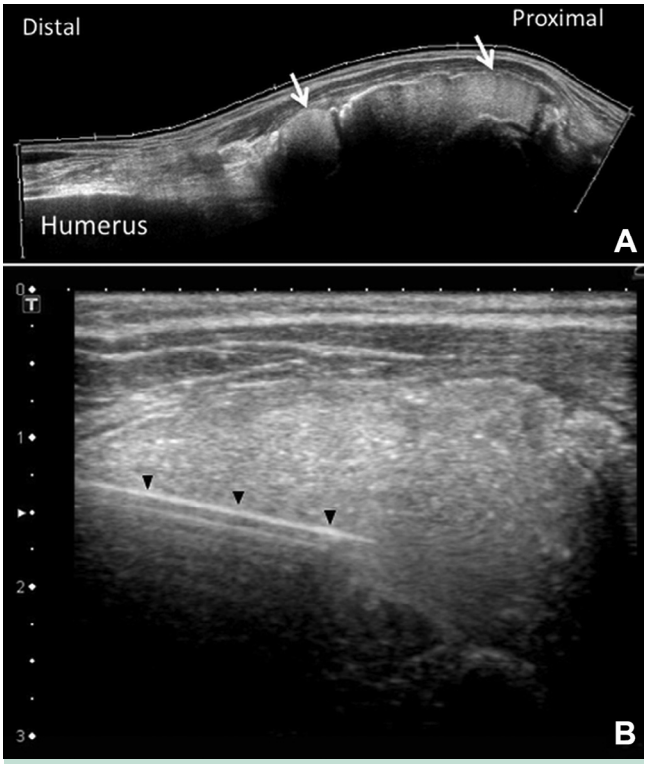

Figure 2 (**A**) Ultrasound imaging of the patient's right shoulder, shown here in this panoramic view, revealed multiple, round, mass-type lesions with homogenous echoes (arrows) and irregularly scattered hyperechoic plaques with acoustic shadows. (**B**) Ultrasound-guided aspiration was performed using the direct in-plane technique. The arrowheads indicate the needle.

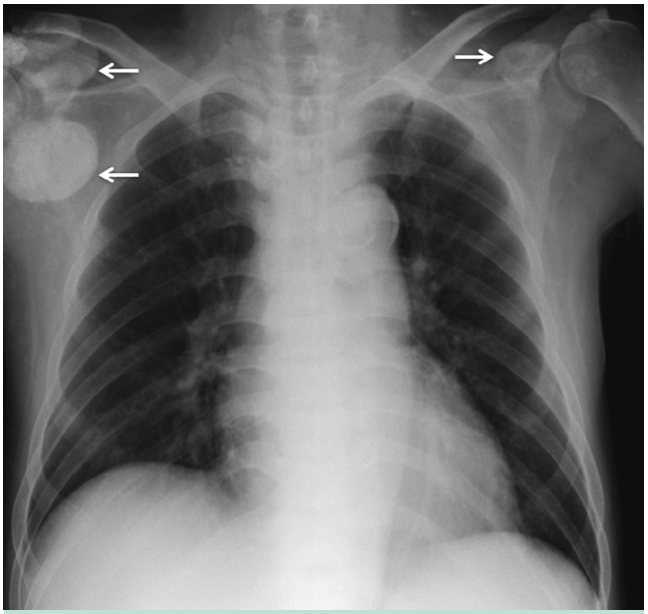

Figure 3 A chest x-ray demonstrated bilateral rounded calcifications and areas of increased density overlying bony structures (arrows). These corresponded with areas of tumoral calcifications and were larger on the right side than on the left.

failure, led to a diagnosis of tumoral calcinosis, a rare disorder characterized by multilobulated calcium salt deposits in periarticular soft tissues. Classically, tumoral calcinosis occurs at large joints, such as the hip, elbow, shoulder, ankle, and wrist, forming on the extensor surface of a joint or on the bursa.[1,2] Primary calcinosis, a familial condition with variable genetic subtypes, is classified into hyperphosphatemic and normophosphatemic types, according to the patient's serum phosphate levels. Secondary calcinosis is often associated with chronic renal failure and hyperparathyroidism.[1,3,4]

The diagnosis of tumoral calcinosis largely depends on findings from radiography, CT, and/or magnetic resonance imaging (MRI). Distinctive radiographic features include amorphous, cystic, and lobulated calcifications at the affected periarticular areas. Computed tomography may further reveal the sedimentation sign, illustrated by layered calcium deposits and associated fluid–fluid levels. Homogeneous patterns of calcifications on CT images might also indicate decreased activity of the disease. On our patient's images, each calcification looked the same all the way through; no one area was of higher density than others.

T1-weighted MRI typically demonstrates inhomogeneous low-signal intensity. That is, the signals are generally of low intensity but by different degrees. On T2-weighted images, tumoral calcinosis can have various signal intensities, including mixed signals; however, it usually appears inhomogeneous with high signal intensity.[1,3] In addition to ascertaining the diagnosis, CT and MRI also help to clarify the extent to which nearby tissues are involved with the tumoral calcinosis.[5]

For our patient, ultrasound was a readily accessible and real-time imaging modality that helped provide a prompt diagnosis. Treatment quickly ensued. In acute cases, in which a mass is initially forming or beginning to enlarge, and vascular signals are more detectable, tumoral calcinosis may present as a well-circumscribed homogenous mass with an irregular inferior border of calcifications. In chronic cases, which tend to be defined by more cystic changes and fibrous septa, ultrasound may reveal a lobulated cystic mass with septa and a calcified rim. The sedimentation sign may also be observed.[6,7]

MANAGEMENT

In patients with massive tumoral calcinosis, treatment should focus on the management of underlying problems, such as serum calcium or phosphate imbalances and hyperparathyroidism. Reports indicate that parathyroidectomy or renal transplantation has led to rapid resolution in some patients.[8,9]

Surgical excision of the mass can often relieve symptoms due to focal compression. However, because the recurrence rate is high, surgery is usually combined with phosphate deprivation and administration of acetazolamide for long-term control. Cystic lesions containing white to pale yellow chalky materials have been identified during surgery.[1,2] Needle

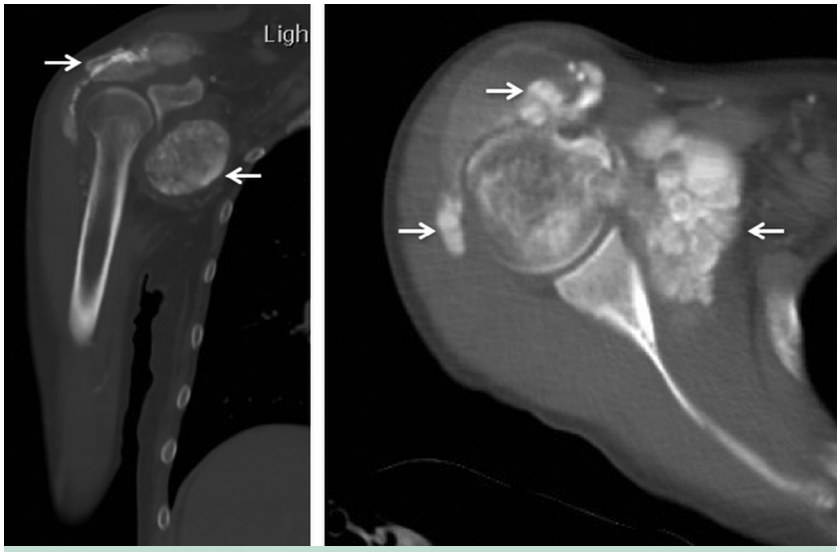

Figure 4 Computed tomography of the right shoulder disclosed multiple amorphous soft tissue calcifications of different sizes (arrows).

aspiration can also help relieve symptoms, while guiding the differential diagnosis of tumoral calcinosis.[10]

Our patient underwent ultrasound-guided aspiration of the tumoral calcinosis, and the procedure sufficiently resolved the mass. His hyperphosphatemia was treated with oral anhydrous sevelamer carbonate. At the 1-month follow-up visit, he had no recurrence of the disease.

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