Tumoral Calcinosis: Early Detection and Effective Treatment Can Reduce Morbidity

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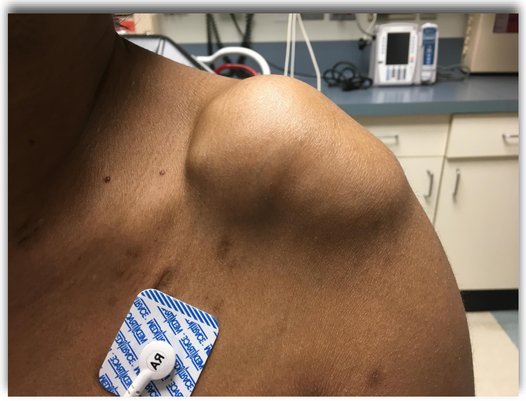
Introduction

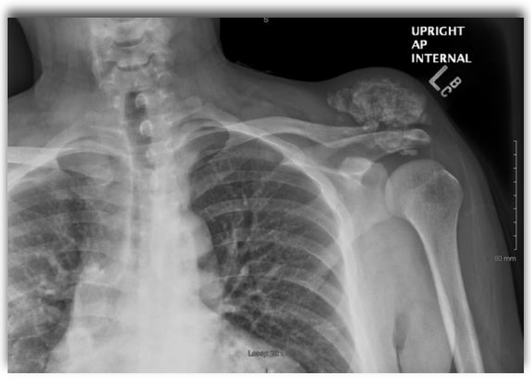
Tumoral calcinosis (TC) is characterized by calcium phosphate deposition in periarticular soft tissue, developing gradually over several years. It can be easily missed until large masses are noticed around weight bearing joints, restricting the range of motion. Hips and shoulders are most often involved. TC is a rare condition with controversial pathogenesis. Patients are predominantly of sub-Saharan African descent, and present with hyperphosphatemia and normocalcemia. TC can be subdivided into primary and secondary varieties; primary TC is relatively poorly understood and may be due to familial defects of metabolism or transport of phosphorus. We present a case of the secondary variety- TC associated with chronic renal failure resulting in hyperparathyroidism. The prevalence of TC in patients undergoing hemodialysis or peritoneal dialysis is 1.6%. In the past, these cases have been regarded as irreversible due to ineffective medical management and frequent recurrence after excision.



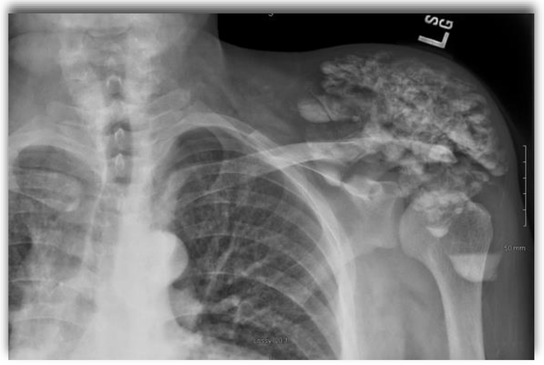
Case

A 40yo male with extensive past medical history significant for ESRD on hemodialysis presents with bilateral shoulder pain, progressively worsening over the past 3 months. Shoulders appear to have multilocular, cystic masses that are hard but mobile. They are mildly tender to palpation, without erythema, warmth, or ulceration. Symptoms include decreased range of motion secondary to the swelling around the shoulder joints, and intermittent pain that wakes the patient at night. CMP is significant for phosphorus 8.3, BUN 55, creatinine 8.97, GFR 8, while calcium was within normal range of 8.7. Radiographic studies of the left shoulder showed amorphous cloud-like calcification overlying the acromioclavicular joint and distal clavicle which measures 12.5 x 7.7 cm, while studies of the right shoulder reveal similar results measuring 10 x 5.3cm. The patient has been worked up for similar shoulder pain several times in the past, most recently one year before this presentation, at which time smaller calcifications were present, but no treatment was provided.

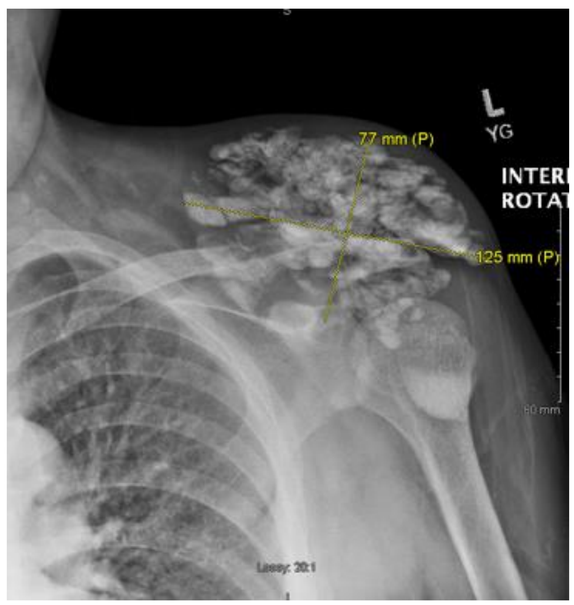
Imaging



May 21, 2016



February 19, 2017



Left shoulder radiograph on March 15, 2017 showing significant increase in amorphous cloud-like calcifications in soft tissues overlying the distal clavicle measuring approximately 77 x 125 mm.

Conclusions

Although TC is rare, patients with ESRD are at an elevated risk. Previous literature suggests that most patients with secondary TC do not respond to medical management. Due to the metabolic nature of the disease, surgical excision of the calcifications often results in recurrence. However recent studies have shown that treatment with a phosphate binder, sevelamer, along with dietary phosphate restriction can be sufficient to cause regression of the masses over several years. Calcium phosphate is readily exchanged with calcium and phosphate in the depleted serum. Further, refractory cases can be treated with subtotal parathyroidectomy, resulting in rapid regression of the masses over the course of a few months. Since the ESRD patients are at an increased risk of developing TC, they would benefit from early diagnosis and treatment to prevent disfigurement and suffering.

References

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