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On the Cover

What is the diagnosis?

Case Description

A 44-year-old male with kidney failure on peritoneal dialysis and multiple comorbidities, including severe peripheral vascular disease, heart failure with reduced ejection fraction, superior vena cava syndrome from bilateral subclavian thrombosis, and chronic sternal osteomyelitis secondary to a prior episode of staph epidermidis endocarditis, was admitted for chest pain and hyperkalemia. He had significant history of extremely poor adherence with peritoneal dialysis and oral phosphate binders. He was on suppressive antibiotics for his chronic sternal osteomyelitis. On presentation, he was afebrile and hemodynamically stable. Physical exam revealed a tender, firm, immobile nodule at the mid-sternum and multiple hard nodules on his hands and fingers. He had mild erythema and tenderness at the incision site of a prior below-the-knee amputation. His serum potassium was 6.3 mmol/L, phosphorus was 13.5 mg/dl, calcium was 7.2 mg/dl, and parathyroid hormone was 995 pg/ml. Electrocardiogram did not reveal peaked T-waves or signs of ischemia. After the patient's electrolyte disarray was corrected, imaging was performed for further evaluation.

Image Description

Left: CT scan of the chest revealed a lobulated, calcific mass (10.4 × 9.7 × 11.3 cm) arising from his previous sternotomy site. The cystic lesions contained multiple fluid-calcium levels, completely replacing the superior half of the sternum and encasing the previous sternotomy wires.

Center: Imaging of his hands revealed extensive soft tissue calcifications throughout the hand and fingers. There were also postsurgical changes from a prior amputation of the third metacarpal.

Right: Right knee plain film showed several lobulated, dense calcifications near the tibial and fibular stumps with extensive vascular calcifications. The findings were consistent with severe vascular calcification and tumoral calcinosis.

Teaching Points

Tumoral calcinosis is characterized by calcium deposition in periarticular soft tissue. It typically manifests as subcutaneous masses around major joints such as the hips, shoulders, and knees, with case reports of it occurring in the axial skeleton. The underlying pathophysiology can be divided into primary or secondary in etiology. Primary tumoral calcinosis occurs as a result of genetic mutations, which lead to pathologic calcification processes. There is a normophosphatemic entity and a hyperphosphatemic entity, each associated with its own mutation and a familial pattern. Secondary tumoral calcinosis is most commonly associated with kidney failure, as highlighted in this case. The pathophysiology is related to longstanding hyperphosphatemia from secondary or tertiary hyperparathyroidism. Tumoral calcinosis is difficult to treat, regardless of the etiology. The best success comes from aggressive control of serum phosphorus levels and kidney transplantation in those with kidney failure. Although surgical excision is an option, deposits can recur, and patients often have comorbidities that make them poor surgical candidates.

(Images provided by Kirsten Lee Koons, University of Virginia, Internal Medicine, Charlottesville, Virginia)