A rare case report of tumoral calcinosis syndrome in an adult with rheumatoid arthritis

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Summary

Tumoral calcinosis is a rare benign disease entity that mimics a neoplasm and is manifested by calcium deposition in the soft tissues around the large joints. It can be primary or secondary to renal failure and hyperparathyroidism. This is, to our knowledge, the first reported case of tumoral calcinosis in a Hispanic male with rheumatoid arthritis.

KEY WORDS: tumor calcinosis; rheumatoid arthritis.

Introduction

Tumoral calcinosis is a distinct disease of unknown etiology. The disease was first recognized in 1899 by Duret who reported cases of a 7-year-old girl and her younger brother with multiple calcifications around the elbow and hip joint (1). After studying the disease for more than 20 years, Teutschlaender named the disease as lipocalcinogranulomatosis (2). Inclan was the first to coin the term tumoral calcinosis to describe this condition in 1943 (3). This is, to our knowledge, the first reported case of tumoral calcinosis in a Hispanic male with rheumatoid arthritis.

Case report

A 53-old-Hispanic male with a past medical history of rheumatoid arthritis of more than 10 years' duration was admitted with a chief complaint of painful swelling in the left wrist and left hip for the past 2 weeks. He described the pain as dull and constant. There was no history of any trauma. He also denied any family history of any malignancy. His vital signs were stable. His physical examination revealed left thigh, left elbow (Figure 1A) and left wrist (Figure 1B) non-erythematous, tender and lobulated tumoral masses. His laboratory studies revealed normal renal function and parathyroid hormone level. The calcium level was 9.1mg/dl, phosphate level 3.0mg/dL and vitamin D level 30.3ng/mL. An X-ray of the left elbow (Figure 2A) revealed partially calcified mass with numerous lobulations without any definite evidence of bone destruction. CT scan of the left thigh showed soft tissue calcified masses (Figure 2B). CT guided biopsy of
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the left thigh mass was done. A thick white toothpaste-like substance was aspirated and sent for histopathologic examination that revealed glandular proteinaceous material mixed with degenerating inflammatory cells. The cell block revealed abundant calcium deposits (Figure 3). No neoplastic process was identified in the submitted specimen and the final pathologic diagnosis was tumoral calcinosis syndrome. Surgery was consulted for debulking surgery and patient was started on phosphate binders. Two years after his first presentation, the patient presented for clinical follow-up and his radiologic findings did not improve. The bony lesions got worse and the patient refused any further aggressive treatment measures.

Discussion
Tumoral calcinosis is characterized by the presence of lobulated cystic soft tissue masses which are usually nontender, and firmly attached to adjacent muscle and fascia. They often, containing chalky calcareous material and sometimes these lesions, which are of varied sizes, may ulcerate the overlying skin and drain that chalky, milky fluid (9). The common sites for these masses are hips, elbows and shoulders. Involvement of the hands and knees is uncommon (4). There are no cases of gross bony destruction that have been reported with tumor calcinosis (5). Tumoral calcinosis predominately involves otherwise healthy patients during childhood or adolescence, typically in the first or the second decade of life with no sex predilection (6). The disease is much more common in patients of African American descent unlike our patient who was of Hispanic ethnicity (7-9). Although autosomal dominant cases have been described, this condition may have an autosomal recessive pattern of inheritance as familial cases have occurred in siblings with parents unaffected (10). Familial forms of tumoral calcinosis are linked to mutations of various genes including GALNT3, FGF-23, and Klotho. As illustrated by the current case, the condition can also occur in patients without a positive family history. Usually patients have hyperphosphatemia (unlike our patient), normal serum calcium and a normal PTH (10-13). The tumoral lesions typically measure 5 to 15 cm in diameter and are composed of numerous small deposits of hydroxyapatite calcium salts (1). Margins are variable; some lesions have a well-defined pseudocapsule, whereas some infiltrate the surrounding tissues (8). Microscopy in the active phase demonstrates a foreign body response with a rim of chronic inflammatory cells, giant cells, and macrophages surrounding the calcific deposits. A dense fibrous material is present surrounding the central calcified material in the inactive phase (1). Typical X-ray findings are well-defined calcified para-articular masses that may be multiloculated or lobular. Involvement of the adjacent bone with erosions may rarely be present (14).

The treatment focuses on reduction of serum phosphate through restriction of dietary intake and oral phosphate binders (15). Clinical and radiographic improvement has been reported after phosphorus deprivation therapy (15, 16) although our patient did not improve after these measures. The recommended management of masses caused by tumoral calcinosis is surgical excision which requires a complete removal of abnormal tissue to prevent recurrence (17). Alternative treatment strategies using steroid and radiation
therapy have also been proposed but do not consistently prevent lesion recurrence. Therefore, surgical management appears to be the only curative approach. Also as evidenced in our patient, surgical resection of these masses usually recurs.

Conclusion

In summary, our presentation is the first report regarding tumoral calcinosis in a Hispanic patient with rheumatoid arthritis. Imaging and pathological examinations are the commonly used diagnostic procedures. Although the pathogenesis of the calcification process in tumoral calcinosis is still controversial, surgical removal remains the mainstay treatment with a satisfactory prognosis.

References