CASE REPORT

Tumoral calcinosis: a case report

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Abstract Tumoral Calcinosis is a distinct clinical and histological entity that is characterized by large periarticular deposits of calcium resembling a neoplasm and is found foremost in the region of hip, shoulder and elbow. We report a case of Tumoral Calcinosis in a 25-year-old male who presented to us with gradually increasing swelling of right axilla, and both hips of nearly two years duration. It was diagnostic enigma for the treating surgeons but with the help of an astute pathologist we diagnosed this rare condition and successfully treated it surgically.

Keywords Tumoral Calcinosis · Periarticular

Case report

A 25-year-old male came to our outdoor with complaints of gradually increasing swelling on both hips and right axilla for last 2 years. Two years back he noticed some restriction of right shoulder joint movement for which he had some massage therapy. Couple of days later he noticed a lump of about 2 inches size in right axilla. About 2 weeks later he noticed two similar lumps on the sides of both hip joints. As these swellings were gradually increasing in size he sought local medical help and treating physician thought it to be chronic abscesses and did I & D of these swellings. Since then thick chalky white discharge used to come.

Examination revealed three lumps, one in axilla of about 3 x 2 inches, firm in consistency, nontender, nonmobile with ulcerated surface, floor of which was covered with chalky-white exudates. Two hip masses were about 5 x 3 inches with similar characteristics as that of axillary lump. There were no other significant clinical findings.

Investigation revealed normal hemoglobin, normal total and differential count and normal routine urine. Renal function was normal. Plain X-ray of both hips and axilla revealed spotty calcification in periarticular soft tissues. Serum calcium was 8.4 mg/dL and inorganic phosphorus 4.2 mg/dL. Serum parathormone level was normal. Histopathology showed diffuse and extensive areas of spotty calcification and fibrocollagenous tissue interspersed with macrophages and mixed inflammatory cell infiltrate, appearance suggestive of Tumoral Calcinosis. Chemical analysis showed Calcium Phosphate crystals. Diagnosis of Tumoral Calcinosis was thus made. We performed complete excision of these lumps in 2 settings and patient is doing fine postoperatively. We will follow him up 3 monthly for any evidence of recurrence.

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Fig. 1 Radiograph of right hip showing periarticular soft tissue calcific deposits.

Discussion

Tumoral Calcinosis is a rare benign metabolic dysfunction of unknown etiology. It manifests mainly as large subcutaneous deposits of calcium phosphate near large joints and is also characterized by slow progressive growth [1, 2, 3]. It occurs predominantly in adolescents and young adults and is usually multiple [3]. Tumoral calcification differs from other disorders like Hypervitaminosis D, Milk-Alkali Syndrome, Dystrophic Calcification in that periarticular masses contain amorphous calcium phosphate complexes. Dystrophic calcifications are more superficially located, smaller and unlike Tumoral Calcinosis do not increase in size. About one-third of cases are familial, with both autosomal recessive and autosomal dominant modes of inheritance [4, 5]. The primary defect responsible for the metastatic calcification appears to be hyperphosphatemia from the increased capacity of the renal tubule to reabsorb filtered phosphate [4, 6, 15]. The calcific masses are often painless and become large and bulky [3]. Complications include compression of neural structures and ulceration of the overlying skin with drainage of chalky fluid and risk of secondary infection [3].

The most common laboratory findings are hyperphosphatemia and elevated serum 1,25-dihydroxyvitamin D levels [3, 5, 6, 7]. Serum calcium, parathormone [3, 9, 10, 11] and ALP [8] levels are usually normal. Renal function is also usually normal [4]. Tissue trauma from joint movement may contribute to the periarticular calcifications.

Fig. 2 Intraoperation picture of right hip swelling showing chalky white material.

Radiographic findings are typical multiple rounded opacities separated by radiolucent lines (fibrous-septa) [3, 12].

Microscopically it is firm, rubbery mass, that is not encapsulated and extends into adjacent muscles and tendons. On section, it consists of dense fibrous tissue containing in-spaces calcareous material. Chemical analysis of calcified material reveals mixture of calcium carbonate and calcium phosphate. The microscopic characterization is hydroxyapatite crystals or amorphous calcium phosphate complexes.

Complete excision of the tumor is the treatment of choice [4]. Removal of larger masses is often followed by recurrence, secondary infection and abscess formation. Reduction of serum phosphate by chronic phosphorus restriction in diet also helps [3]. The addition of phosphaturic agent acetazolamide may also be helpful [3, 13].

Conclusion

In 1943, Inchlan described Tumoral Calcinosis as disease of its own. More than 800 cases of this disease is described in the literature under different names. The young age of our patient together with the presence of multiple, slow-growing, periarticular swellings over a span of 2 years, with restriction of joint mobility, chalky-white discharge, spotty periarticular soft-tissue calcification on x-rays, normal serum calcium, PTH and renal function, borderline serum phosphorus and calcium phosphate crystals on chemical analysis allowed us to make the diagnosis of Tumoral Calcinosis and treat the patient successfully. This diagnosis should be kept in mind while dealing with similar periarticular swellings [3].

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