TUMORAL CALCINOSIS*
A Case Report

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SUMMARY: Özkan İ, Türeli C, Çullu E, Karaman C, Şendur F, Alparslan B. (Department of Orthopedics and Traumatology, Adnan Menderes University Faculty of Medicine, Aydın, Turkey). Tumoral calcinosis: a case report. Turk J Pediatr 1999; 41: 375-379.

Tumoral calcinosis is a rare disorder with the calcified masses in subcutaneous tissues. We report herein a nine-year-old girl, in whom the calcified lesions bilaterally involved the soft tissues in the anterior part of the knee joint. Serum calcium and phosphorus levels were in normal ranges and there was no family history. Surgical excision was performed and recurrence was not observed in early follow-up. Review of the literature shows that only clinical and radiological appearance of tumoral calcinosis are generally agreed while its epidemiology, etiology and treatment are still under discussion. *Key words: tumoral calcinosis, ectopic calcification syndrome.*

Tumoral calcinosis is an uncommon disease of uncertain origin, which is rarely seen in Europe but is much more common among black Africans[1]. This ectopic calcification syndrome is characterized clinically by para-articular soft tissue calcifications. Various locations and types of calcium deposits have been defined. Irregular and painless calcifying masses were previously reported arround the shoulder, hip, elbow, temporomandibular joint, paraspinal soft tissues and eye[2-6]. Cases of tumoral calcinosis in association with hyperphosphatemia have been reported, and it seems likely that hyperphosphatemia may play a role in the pathogenesis[4,7,8]. However, numerous underlying factors are thought to increase susceptibility to this disease. Genetical disorders, recurrent soft tissue microtrauma and terminal renal failure have been cited as causes of tumoral calcinosis[7,9]. In most cases, local factors are probably involved as well.

Histologically, there are fibrous walled cystic spaces containing structureless calcific debris in association with a variable inflammatory reaction.

We report herein a case of a nine-year-old girl with idiopathic tumoral calcinosis on the anterior part of both knees.

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Case Report

A nine-year-old girl was admitted suffering from multiple, painless masses on the anterior part of both knees since 10 months of age. The masses were irregular in shape and located on the anterior part of the patella and patellar tendon (Fig. 1). Radiographs revealed several calcific subcutaneous masses around the knee joint (Fig. 2).

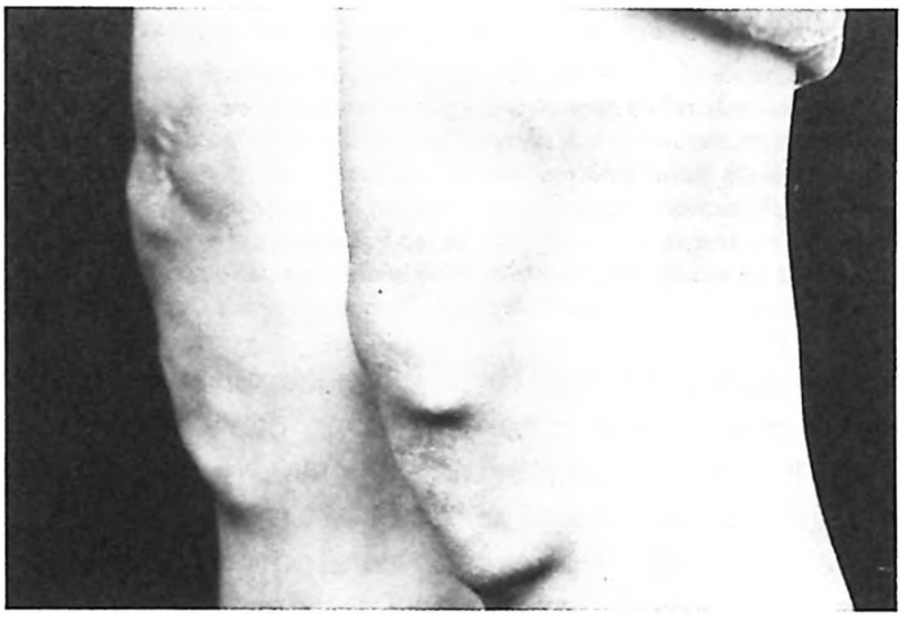

Fig. 1: The appearance of the patient's knees.

Fig. 2a: Lateral radiography of the right knee.

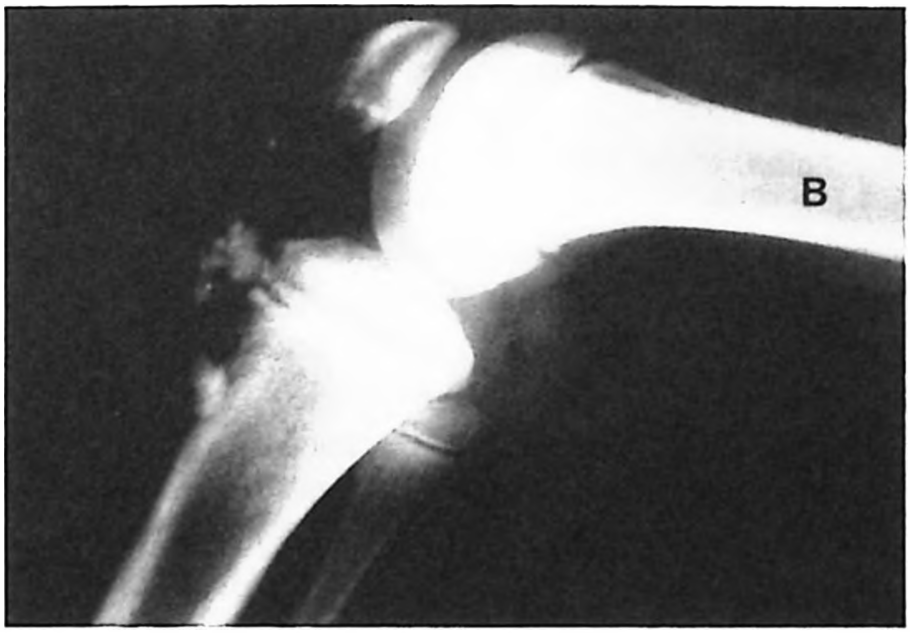

Fig. 2b: Lateral radiography of the left knee.

Serum calcium was 9.5 mg/dl and serum phosphate 4.0 mg/dl. Urea, creatinine, alkaline phosphatase, sodium, potassium, magnesium, parathormone, free and total T3 and T4, testosterone and cortisone values were in normal limits. Excretions of calcium and phosphate in 24-hour urine were also normal. Laboratory examinations for serum calcium and phosphate were repeated at monthly intervals three times, each time revealing normocalcemia and normophosphatemia.

The masses on the right knee were removed under general anesthesia. During surgery, a chalky semifluid material extruding through the masses was observed. Histological examination revealed fibrous walled cystic spaces containing calcific debris and various inflammatory reactions (Fig. 3).

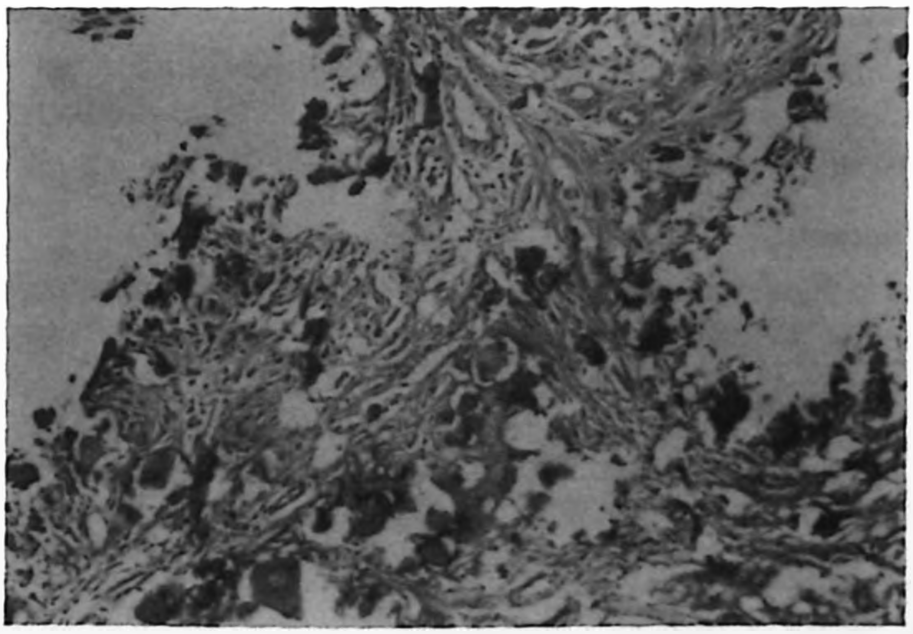

Fig. 3: Photomicrograph of calcifying masses (hematoxylin eosin x 200).

Discussion

Idiopathic tumoral calcinosis should be diagnosed by eliminating the other diseases in which similar calcifying masses are seen. Idiopathic synovial chondromatosis must be differentiated, especially when the lesions in tumoral calcinosis are seen around big joints as in the case reported here. Tumoral calcinosis may be confused with synovial sarcomas when the synovial sarcomas present with dense and conglomerate calcifications. Malignancies such as extraskeletal osteogenic sarcoma, extraskeletal chondrosarcoma and mesenchymal chondrosarcoma must also be considered in the differential diagnosis[10].

Pathogenesis-based classification of tumoral calcinosis was made by Smack et al.[11]. They suggested three pathogenically distinct subtypes of tumoral calcinosis: 1) primary normophosphatemic tumoral calcinosis, 2) primary hyperphosphatemic tumoral calcinosis, and 3) secondary tumoral calcinosis. The presented case is a group 1 tumoral calcinosis with normal serum phosphate level and no family history. Soft tissue calcifications are a frequent complication in patients with chronic renal failure[7]. These patients are accepted as secondary tumoral calcinosis. Familial cases of tumoral calcinosis have also been reported[9]. Some authors suggest that tumoral calcinosis should be included among the clinical presentations of calcium pyrophosphate dehydrate crystal deposition disease[12]. However, we believe that is difficult to explain this with normal serum calcium and phosphorus levels as in the presented case.

The medical treatment of tumoral calcinosis is symptomatic. At present the valid medical treatments are surgical excision and/or a low-phosphorus, low-calcium diet with phosphate-binding antacids[8,13]. Acetozolamide appears useful in treatment of tumoral calcinosis, which is resistant to phosphorus deprivation by aluminum hydroxide alone[8]. Complete surgical excision of lesions has been recommended[13,14], but recurrence is common[16,14]. Although there has been only eight months of follow-up in the present case, there has been no early recurrence of the lesions around the knee.

Review of the literature shows that only clinical and radiological appearance of tumoral calcinosis are generally agreed while its epidemiology, etiology and treatment are still under discussion.

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