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Multifocal tumoral calcinosis in a 4-year-old girl

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

ABDEF 1 Ilyas Sayar
DEF 2 Kemal Peker
AD 3 Alparslan Kapısız
DE 4 Isıl Esen Bostancı
F 5 Mehmet Gürbüzel
AF 2 Arda Isik
F 6 Necla Aydın Peker

1 Department of Pathology, Erzincan University, Erzincan, Turkey
2 Department of General Surgery, Erzincan University, Erzincan, Turkey
3 Department of Peadiatric Surgery, Rize State Hospital, Rize, Turkey
4 Department of Radiology, Erzincan University, Erzincan, Turkey
5 Department of Molecular Biology, Erzincan University, Erzincan, Turkey
6 Department of Peadiatria, Erzincan University, Erzincan, Turkey

Corresponding Author: Ilyas Sayar, e-mail: drilyassayar@hotmail.com

Patient: Female, 4
Final Diagnosis: Tumoral calcinosis
Symptoms: Hard immobile mass
Medication: —
Clinical Procedure: —
Specialty: Surgery

Objective: Congenital defects
Background: Tumoral calcinosis is an uncommon condition associated with the deposition of painless calcific masses. It is more common in childhood or early adolescence of African-American females.
Case Report: We present a case of a 4-year-old girl with tumoral calcinosis treated surgically. The case is rather rare in terms of the age of the patient and the localization of the masses (gluteal site). In our patient, the biochemical findings were normal, except for hyperphosphatemia and elevated alkaline phosphatase.
Conclusions: Total excision appears to lead to a good clinical outcome and a low incidence of local relapse.

MeSH Keywords: Pathology • Peadiatry • Calcinosis

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Background

Tumoral calcinosis is an uncommon condition associated with the deposition of painless calcific masses. The term tumoral calcinosis was coined by Inclan in 1943 but the condition was identified much earlier [1]. Duret described this condition in 1899 in a young boy and his 17-year-old sister who had multiple calcifications in the vicinity of the elbow joint and the hip [2]. We present a rather rare case of tumoral calcinosis in terms of the age of the patient and the localization of the masses (gluteal site) at different regions in the same patient. The masses were surgically removed.

Case Report

A 4-year-old girl was brought to the pediatric surgery polyclinic by her parents for examination due to swelling in the gluteal site in December 2012. In the anamnesis with her parents, they stated that they had noticed the swelling 3 months earlier. The patient did not have any trauma history or known systemic disease. The physical examination revealed a hard, immobile mass 5 cm in diameter in the gluteal site, as well as a hard, immobile subcutaneous mass at the left hip joint. A superficial examination of the ultrasonograph showed echogenicities with calcified cartilage content and distal acoustic enhancement of the subcutaneous fat tissue (Figure 1). An anteroposterior (AP) radiograph of the pelvis showed calcific masses adjacent to the femur neck and superposed on the midline of the (Figure 2). Pelvic magnetic resonance imaging (MRI) revealed nodular hypointensities with subcutaneous fat tissue calcification at the left hip joint level and the left gluteus (Figure 3). In the axial T2 FSE fat SAT slices, nodular hypointensities were present, as well as hyperintense edema in adjacent subcutaneous fat tissue at the level of the left hip joint and the left gluteus. The ultrasonographic examination of the left shoulder showed acoustic shadowing in a 13-mm segment and the appearance of the same echo as in the hip. In the biochemical tests, results of liver and renal function tests, serum calcium, serum uric acid, erythrocyte sedimentation rate, and hemogram were normal. Only levels of phosphorus (5.9 mg/dl) and serum alkaline phosphatase (342 U/l) were elevated. The gluteal mass was totally excised. Morphologically, the mass had a multinodular structure and was 5×4×3 cm in diameter. Macroscopic analysis revealed a globular yellowish nodule, and the microscopical examination showed amorphous and homogeneous substances, suggesting deposits of calcium. Fibrous connective tissue was found between the nodules (Figures 4 and 5). The nodule was surrounded by scant inflammatory cells and it exhibited focal fat necrosis. The histopathological examination did not reveal any evidence of malignancy. Based on the histopathological evaluation, tumoral calcinosis was diagnosed. The postoperative course showed no complications. No recurrence was observed after a 12-month follow-up period.

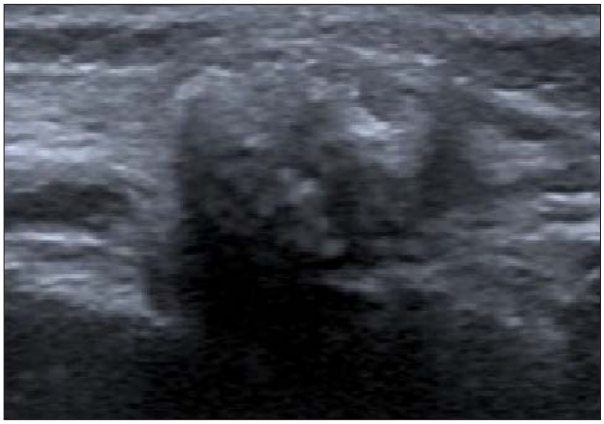

Figure 1. Ultrasonographic view of calcinosis.

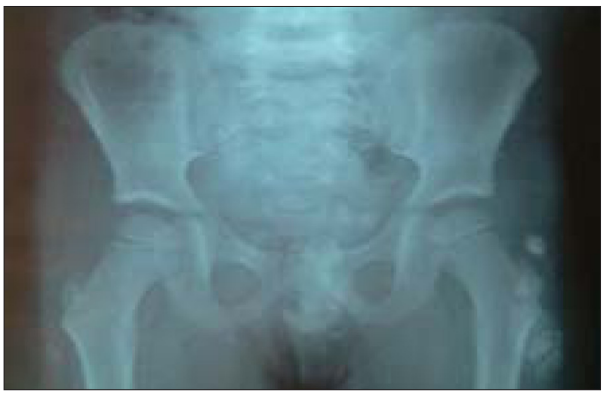

Figure 2. Direct graphy of calcinosis.

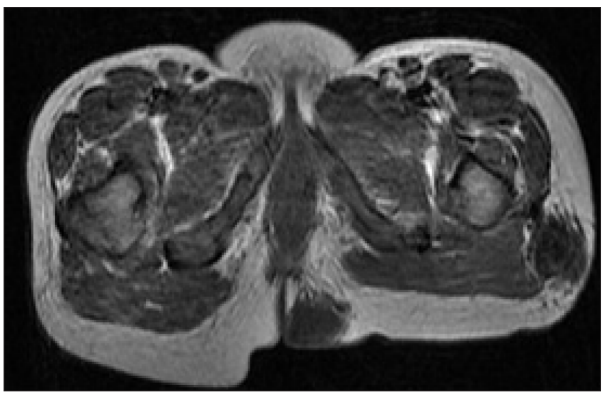

Figure 3. MRI of calcınosis (nodular hypointensities with subcutaneous fat tissue calcification at the left hip joint level and the left gluteus).

Discussion

According to Weiss et al. (2008), tumoral calcinosis is a distinctive clinical and histological entity, which is associated with

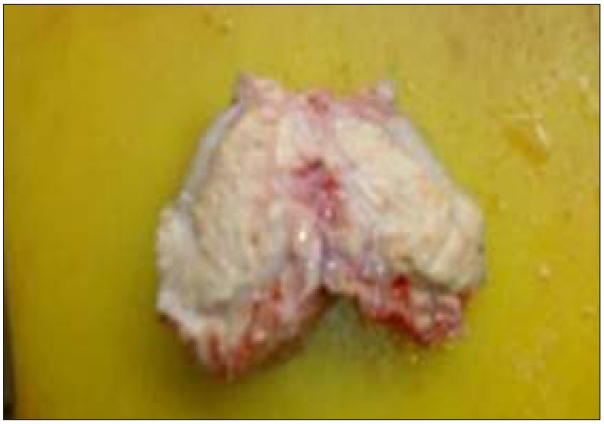

Figure 4. Macroscopic view of calcinosis.

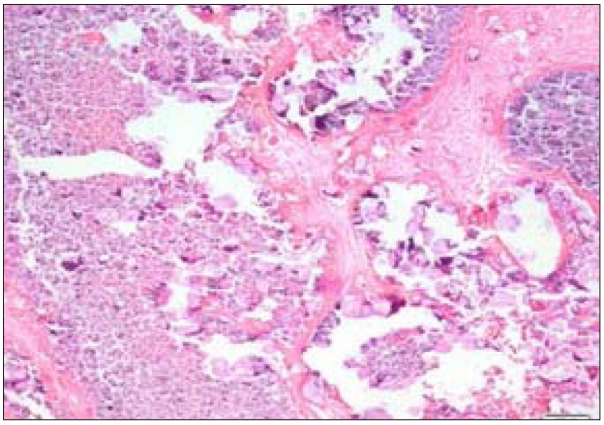

Figure 5. Microscopic view of calcinosis.

tumor-like periarticular deposits of calcium that are mainly found in the regions of the hip, shoulder, and elbow. The disorder occurs generally in otherwise healthy children, adolescents, and young adults, is more often multiple rather than solitary, and frequently affects 2 or more siblings of the same family. Unlike similar calcifications associated with other diseases (e.g., hypervitaminosis D, renal insufficiency, and milk-alkali syndrome), there are no abnormalities in calcium metabolism [3].

The exact cause of tumoral calcinosis is still not known. Smack et al. [4] proposed a pathogenesis-based classification of tumoral calcinosis, with 3 types recognized: primary normophosphatemic tumoral calcinosis, primary hyperphosphatemic tumoral calcinosis, and secondary tumoral calcinosis. In the first type, patients have no known disorders of phosphate or calcium metabolism. In the second type, they have normal serum calcium and raised serum phosphorus. This condition is thought to be caused by a defect in phosphate resorption. In the third type, patients usually have a concurrent disease, such as chronic renal failure, with secondary hyperparathyroidism, hypervitaminosis D, milk-alkali syndrome, and bone destruction capable of causing soft-tissue calcification. The recurrence rate is very high in this type. Typical clinical findings, radiology, and histopathology showing calcification help in the diagnosis [5,6]. Previous studies showed that calcium levels did not increase, but that slight to moderate hyperphosphatemia was present in many clinical cases [7]. Although serum alkaline phosphate and uric acid levels are normal, calcitriol (1,25-dihydroxyvitamin D_3) levels may be increased. A previous study observed a subcutaneous conglomerate of multiple rounded opacities separated by radiolucent lines (fibrous septa) imparting a “chicken-wire” pattern of lucencies with distinct fluid levels in some nodules in CT, radiography, and MRI examinations [8].

In our patient, the biochemical findings were normal, except for hyperphosphatemia and elevated alkaline phosphatase. There was no past trauma history or family history of tumoral calcinosis. In the histopathological evaluation, the presence of rough calcific nodules of various sizes separated by thick fibrous bands of subcutaneous tissue made it possible to distinguish them from other types of subcutaneous metastatic calcification, dystrophic calcification, and intraepidermal calcific nodules, all of which should be considered in the differential diagnosis. Only a few cases of tumoral calcinosis have been reported in children in the literature [9,10]. This is the first case in terms of age, sex, and localization.

Although in our case total resection of the mass was done, various other operations can be done. Lykoudis et al. [11] used wide excision and reconstruction with bilateral V-Y advancement gluteal fasciocutaneous flaps in their case.

Conclusions

We conclude that although a diagnosis of tumoral calcinosis is possible with imaging techniques, a histopathological study is essential to make a definitive diagnosis. Medical treatments have been attempted but been reported to have limited success. Early surgical removal of the mass appears to be essential [4].

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