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DISTAL FEMORAL ANEURYSMAL BONE CYST, CROSSING THE OPEN GROWTH PLATE AND EXTENDING INTO THE EPIPHYSIS.

Nikolaos A. LALIOTIS	MD, PhD, M. Ch.Orth European Interbalkan Medical Center, Thessaloniki, Greece - 00306944327722
Iordanis G. PETRAKIS*	MD, Orthopaedic Department, General Hospital of Chalkidiki, Polygyros, Greece - 00306974427200 *Corresponding Author
Chrysanthos K. CHRYSANTHOU	MD, European Interbalkan Medical Center, Thessaloniki, Greece - 00306944671049
Panayiotis A. KONSTANDINIDIS	MD, European Interbalkan Medical Center, Thessaloniki, Greece – 006977071744

ABSTRACT We report the case of an aggressive aneurysmal bone cyst, in a 7-year-old girl, crossing the middle of the growth plate and extending into the epiphysis. Plain radiographs, bone scan and MRI were the imaging methods that were used to reach the diagnosis and to rule out other aggressive bone lesions that invade into the epiphysis. The aneurysmal bone cyst was operatively treated with curettage, while preserving both the periosteum and the growth plate. Upon her latest follow-up visit, 3 years postoperatively, the radiographic and clinical examination of the patient yielded satisfactory results, without signs of recurrence and severe growth disturbances.

KEYWORDS : Aneurysmal bone cyst, Benign bone tumor, Children, Growth plate

Introduction

Aneurysmal bone cyst (ABC) is characterized as a benign and expansive osteolytic lesion, active and aggressive, that usually originates at the metaphyseal - diaphyseal area of the long bones [1]. Its incidence is 0, 14/100,000 of the population per year and specifically ranges between 70% - 80% in the second decade of life [2]. An ABC may appear as a primary condition or as a secondary response to other lesions, such as giant cell tumors, chondrob lastomas, chondromyxoid fibromas, chondrosarcomas, telang iectatic osteosarcomas or metastatic disease [3].

Localization of the lesion predominantly involves the metaphysis of a long bone, with assymetrical involvement [4]. Juxtaepiphyseal ABC has been well described, as an aggressive lesion that affects the growth [5]. The invasion of an ABC through the growth plate is extremely rare, with only few cases being reported, affecting the fibula, upper and lower tibia and distal ulna and metatarsals [6-12]. The epiphysis is an infrequent area for an ABC to be located entirely and as a primary condition, with only three cases being reported in the literature [13-15].

We report a 7-year-old girl with a distal femoral aneurysmal bone cyst crossing the growth plate and extending into the epiphysis. We present the clinical and radiological evaluation of the patient. She was treated with curettage and careful preservation of the periosteum and the growth plate. After 3-years of follow-up, she remains asymptomatic and the ABC is inactive.

Case Report

An otherwise fit 7-year-old girl presented to the outpatient department of our clinic complaining of pain and discomfort of her left knee joint. Her clinical manifestations were increased diameter of the distal femur, and an abnormal gait, as a mild limp. On physical examination, her distal femur was painful on palpation, however full knee range of motion could be achieved. On precise evaluation, she was expressing discomfort when performing solely weight standing on the affected side. Despite the swelling, the skin remained normal and there were no obvious signs of a neurovascular compromise on the left lower extremity.

Plain radiographs revealed the existence of a metaphyseal eccentric, radiolucent, expansive lesion, extending through the adjacent physis into the distal femoral epiphysis (Fig. 1), without evidence of a pathologic fracture. There was a clear line of demarcation of the lesion, in the distal femoral epiphysis. This lytic region with "bubbly appearance" indicates bony septae inside the lesion, whereas marked cortical thinning can be noticed. No periosteal reaction was observed. The growth plate appeared intact, apart from the central whole, well formed in both AP and lateral projections.

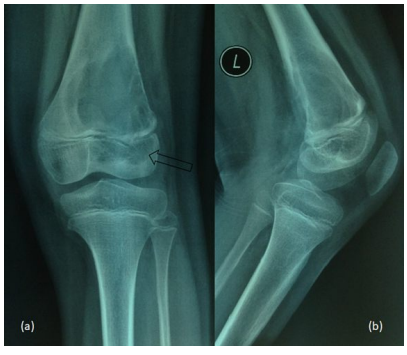

Figure 1. *Initial plain left knee joint radiograph of a 7-year-old female with an expansile, radiolucent eccentric lesion located in the metaphyseal and epiphyseal region of the left distal femoral bone; anterioposterior (a) and lateral (b) projection; note that this radiolucent lesion crosses the growth plate and extends into the distal femoral epiphysis (black arrow)*

The patient underwent an MRI scan (Fig. 2a1, 2a2). The MRI scan clarified the extent of the distal epiphyseal involvement and showed double density fluid - fluid level, distinguishing blood from serum and apparent bony septation. Remarkable feature was the central lesion of the growth plate through which the lesion extended into the epiphysis, with intact articular cartilage. These imaging findings were consistent with an aneurysmal bone cyst. The bone scan indicated increased uptake in the perimeter of the lytic area, with decreased uptake in its center, which is suggestive of an ABC. Blood tests revealed normal values for ESR, CRP, alkaline phosphates.

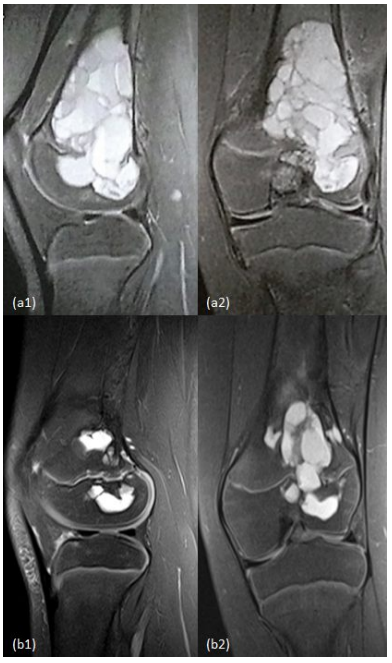

Figure 2. *MRI showing the changes in the aneurysmal bone cyst appearance in the left distal femoral bone region before and after curettage treatment;*

(a) initial appearance of the lesion on the sagittal (a1) and coronal (a2) MRI views showing an area of diffuse high signal appearance in both the left distal femoral metaphyseal and epiphyseal region;

(b) sagittal (b1) and coronal (b2) MRI views after curettage treatment of the aneurysmal bone cyst, clearly showing the reduction of fluid depiction in the involved areas and the intact peripheral zone of the growth plate, ten months post operatively.

We made the differential diagnosis including chondroblastoma (in our patient, the main lesion affected the metaphysis, so it was exceptional), subacute osteomyelitis, eosinophilic granuloma and mainly the possibility of a telengietatic osteosarcoma, due to the disappearance of the cortices in the posterior area of the metaphysis.

Fine needle biopsy revealed blood and we continued to surgical exploration of the lesion. With lateral approach on the metaphysis and appropriate fenestration, we cleared the lesion that consisted of blood filled spaces, with a peripheral membrane. We found the central lesion of the growth plate and through that we carefully cleared the epiphysis. We tried not to increase the lesion of the growth plate. In order to protect the growth plate, we did not use phenol.

The striking finding was the absolute absence of cortical bone. The periosteum was intact, surrounding the lesion.

Due to uncertain diagnosis we did not use autografting which is the method we use in typical ABC lesions.

The tissue elements were sent for histological examination. Histology showed that the cavity lesion was filled with blood. The stroma is characterized from firm fibrous connective tissue that separates the cystic cavities. We found multinucleated giant cells and hemosiderin laden macrophages that were located in places in the stroma. Small foci of osteoid matrix were also observed. No atypical cells were noted. These are elements of aneurysmal bone cyst.

A short-leg cast was applied for 4 weeks postoperatively and no weight-bearing was allowed. After that period, the patient was allowed to initiate a range of active motion exercises to the knee joint. She was advised to avoid weight-bearing for another six weeks. Gradually increasing weight bearing was allowed after that period, initially with the use of two crutches, then with one. After a 3 months period, the patient was capable of full weight-bearing.

The girl was followed up regularly with plain radiographs (Fig. 3), where impressive reformation of the cortices became obvious.

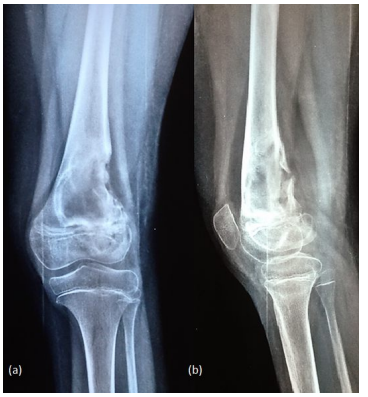

Figure 3. *Plain left knee joint radiographs, three months post operatively, after curettage of the lytic lesion, without the use of autografts; anterioposterior (a) and (b) lateral view demonstrating the reformation of cortical bone on the lateral and posterior distal femoral metaphysis.*

The metaphysis and epiphysis were gradually increasing in bone content (Fig. 4). Slight valgus deviation was seen after 6 months but this did not progress over the following years. Residual elements of the cyst are still remaining, with no further increase, as seen in the latest MRI scan (Fig. 2b1, 2b2).

Figure 4. *Plain knee joint radiographs, three years post operatively, after curettage of the distal femoral aneurysmal bone cyst; anterioposterior comparative right and left knee joints views (a) and (b) lateral left joint view where the bone density in both the metaphysis and the epiphysis has increased and is radiographically noticeable.*

We plan to perform a new curettage if the cyst will further increase and later correct the valgus deviation using an 8 plate. The growth plate remains intact in its peripheral zone, on the MRI studies (Fig. 5).

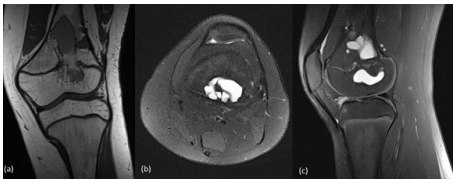

Figure 5. *MRI of the left knee joint after curettage of the distal femoral aneurysmal bone cyst. three years post operatively, showing that the cyst has no further been increased; coronal T1 (a), axial T2 (b) and sagittal T2 (c) MRI views; note the intact growth plate in its peripheral zone.*

The girl has fully returned to her normal everyday life activities and she complaints neither about pain nor about any other functional impairments.

Discussion

Aneurusmal bone cyst (ABC) is a benign but on occasion locally aggressive bone tumor. Campanacci and Capanna classified ABC with a system, based on the anatomical location of the cyst and on the stage of activation (aggressive, active, and inactive) [16]. They usually are located eccentric on the metaphyseal and diaphyseal area of long bones, usually during the 2nd decade of life. Capanna et al further analyzed the juxtaepiphyseal location of the cyst, reporting nine cases [8]. They stated that these type II cases, when aggressive, have the potential to cross the growth plate. The growth plate acts as a barrier to physeal involvement and invasion into the epiphysis. There is no clear explanation for this [17, 18]. It has been suggested that production of an antiangionesis factor or an inhibitor of tumor collagenenase is the possible reason why the growth plate acts as a barrier [19].

The location of the lesion that involved both metaphysis and epiphysis was described in the proximal and distal tibia, proximal and distal humerus, metacarpus, distal ulna and fibula, but not in the distal femur. Rizzo et al reported 15 patients between 2-14 years, with juxtaepiphyseal involvement, with one of them affecting the distal femur, but none had growth plate disruption [9]. Further case reports have been published sporadically affecting the proximal fibula (McCarthy and Ogden 1982) [12], proximal femur (Carlson et al 1972) [10] and the distal ulna (Kapila et al 2015) [15]. We present a 7 year old girl with a lesionextending from the distal femoral metaphysis to the epiphysis, through a central hole in the open growth plate.

Metaphyseal lesions that appear to cross the epiphysis on our imaging methods require thorough investigations in order to reach and confirm an adequate diagnosis of being either a benign condition or malignant.

The differential diagnosis should include pathologies that are well known for their ability to invade through an open physis [20]. We must consider low grade osteomyelitis and eosinophilic granuloma that appear as osteolytic lesions that may extent to metaphysis and epiphysis. Giant cell tumors usually present later in life and more commonly with a closed growth plate. Chondroblastoma is located in the epiphysis but may extend to the metaphysis. It presents with different histological findings.

Telangiectatic osteosarcoma was our main consideration during the procedure, as it can be mistaken for ABC. In our patient there was no tissue out of the femur and there was no periosteal reaction, despite the disappearance of the cortices. This was the reason why we proceeded only to curettage and not to bone grafting.

Treatment of an ABC requires a thorough curettage, in order to remove all elements of the tumor [21, 22]. Dormans et al have reported a 4 step approach for treatment of ABC. In our patient, we tried to remove all elements of the cyst without further damage to the open growth plate. This was difficult for the lesion in the epiphysis, since we cleared the cyst through the existing hole in the growth plate. We did not use phenol, to protect the growth plate. The management of a cyst, with an intralesion removal of the cyst, is described by Grimer and collegues with the term curopsy [24]. This is the treatment we provided to our patient.

Three years later, we observed no recurrence of the lesion and the alignment of the leg is acceptable, but we continue regular follow up of our patient.

Conclusion

In our patient, the osteolytic lesion of the metaphysis that was crossing the growth plate and extending in the epiphysis required appropriate investigation. Differential diagnosis included both benign and malignant diseases. We used radiological, MRI and bone scan examinations. Intraoperatively, our surgical technique focused in the removal of all elements of the cyst. We aimed to avoid any further damage to the growth plate. Preservation of the periosteum was essential for the bone reformation.

The final result is satisfactory one. We continue to follow up our patient for possible recurrence of the cyst or axial deviation of the affected limb.

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