J Orthop Sci (2011) 16:665–669
DOI 10.1007/s00776-011-0070-2

CASE REPORT

Primary aneurysmal bone cyst of the proximal tibia crossing the open physis

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Received: 3 July 2010 / Accepted: 27 October 2010 / Published online: 10 May 2011

Introduction

An aneurysmal bone cyst (ABC) is a hyperplastic reactive lesion of unknown etiology consisting of cystic cavities containing blood lined by mesenchymal reactive tissue. It occurs more often in females (2:1), usually before the age of 20 years, and most commonly in long bones [1]. The extension of the primary ABC to the epiphysis after crossing the viable physis is an exceptional finding, and very few such cases have been reported in the literature [2–6]. Although uncommon, ABC can cause severe destruction of long bones in children and young adults. The suggested treatment for such aggressive cases ranges from simple curettage to wide resection and structural reconstruction [7, 8]. Here, we report the follow-up for a 10-year-old girl with large, aggressive ABC of the proximal tibia with transphyseal extension to the epiphysis, which was treated with intralesional excision, curettage and bone grafting.

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Case report

We present a case of 10-year-old girl who suffered a fall from stairs and sustained injury to the proximal aspect of her right leg. However, there was no history of bleeding, and other organ systems were apparently normal. On radiographs, the patient was diagnosed with a pathological fracture through a cyst in the proximal tibia, and was treated with a long leg plaster. The plaster was continued for 8 weeks until healing was demonstrated on repeat radiographs, after which the patient was referred to our institution for further evaluation. She presented to the outpatient department of orthopedic surgery. Additional queries revealed that there was no history of fever, weight loss, anorexia or cough with sputum. There was no other swelling elsewhere in the body. Anteroposterior and lateral radiographs of the right knee showed a healed fracture in the proximal metaphyseal area of her skeletally immature tibia along with a multilocular, large, geographical type of lytic area involving the whole of the metaphysis with minimal periosteal reaction, consistent with Campanacci type II juxtaphyseal ABC [9] (Fig. 1). Hematological parameters were within normal limits. She was put on a functional knee brace and her parents were instructed to follow-up regularly.

The patient reported back after 3 months with increased swelling of the same region. On physical examination, the patient had swelling involving the right knee and the proximal leg. The swelling was slightly erythematous, warm to the touch, firm in consistency, immobile and nonfluctuant. The range of active knee movement was

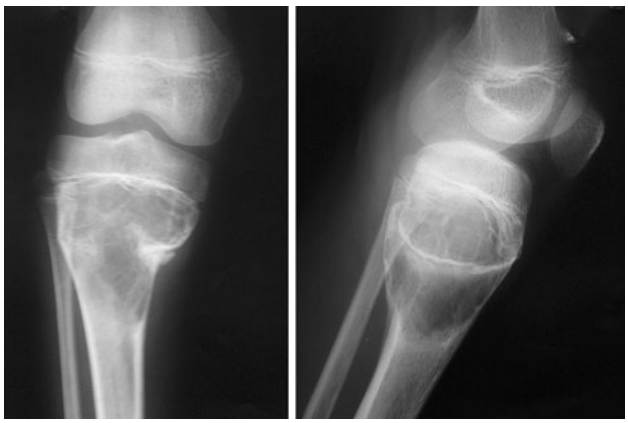

Fig. 1 Radiographs of the right knee with the leg showing a multilocular large osteolytic lesion involving the proximal tibial metaphysis (Campanacci type II ABC) with a healed fracture in a skeletally immature patient

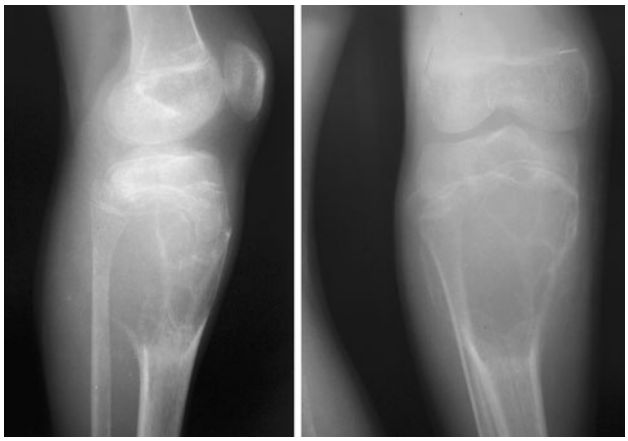

Fig. 2 Radiographs performed after 3 months showing the aggressive nature of the ABC with marked cortical attenuation, especially over the posterior aspect. An osteolytic area is also visible in the proximal tibial epiphysis

20–100°. There was no distal neurovascular deficit. Radiographs taken at this time showed an expansile cyst with cortical attenuation; the cortex was eggshell thin over the posterior aspect of the tibia. Radiolucency was also noticed in the proximal tibial epiphysis (Fig. 2). Magnetic resonance imaging (MRI) was performed, which revealed a multilocular lesion with fluid levels confirming its vascular nature. The cyst was found to be extending into the epiphysis, crossing the open physis (Fig. 3). A differential diagnosis of telangiectatic osteogenic sarcoma was considered. Fine needle aspiration cytology revealed blood only. Biopsy of the lesion showed multiple blood-filled sinusoidal spaces separated by fibrous and osseous septa of varying thicknesses lined with fibroblastic cells, spindle cells, multinucleated giant cells and strands of mature osteoid, which was suggestive of an aneurysmal bone cyst.

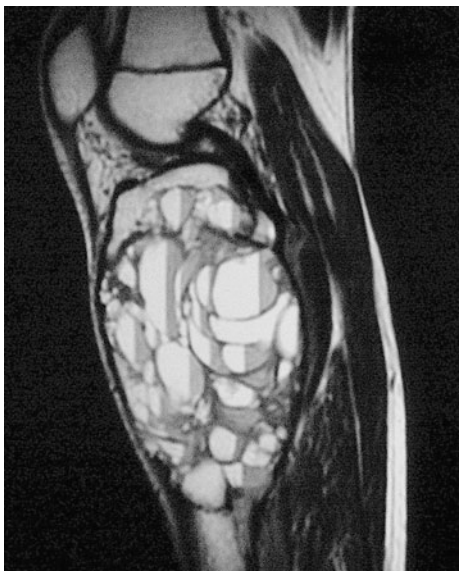

Fig. 3 MR imaging of the proximal tibia shows high signal intensity in the proximal tibia, with multiple fluid levels. Erosion through the proximal tibial physis and extension into the epiphysis is clearly visible

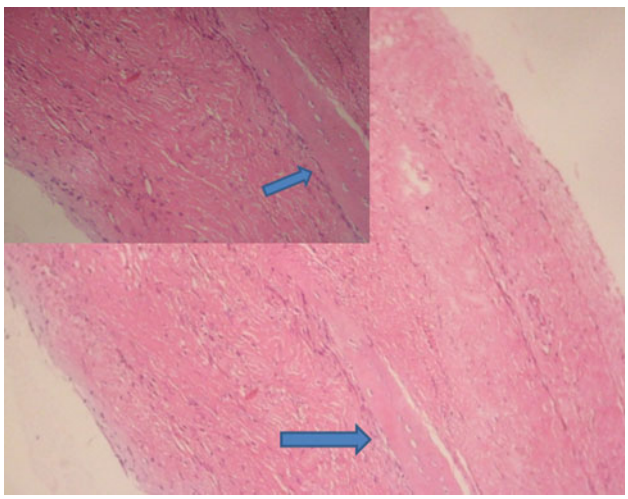

Fig. 4 ABC septa with fibrocollagenous tissue and bony trabeculae (marked with an *arrow*) (H&E, ×10). *Inset* shows a magnified view of the area marked with an arrow in the original figure (H&E, ×20)

There were no features that were suggestive of malignancy (Figs. 4, 5).

The patient was scheduled for intralesional excision, curettage and bone grafting. On exploration, hemorrhagic aggregates of tissue with cortical attenuation were found. The extension of the cyst cavity into the epiphysis was confirmed peroperatively. Since a large defect had to be filled with the graft, an autogenous corticocancellous bone graft from the iliac crest was mixed with an allograft. All of the excised tissue was submitted for histopathological examination. Multiple sections were studied after

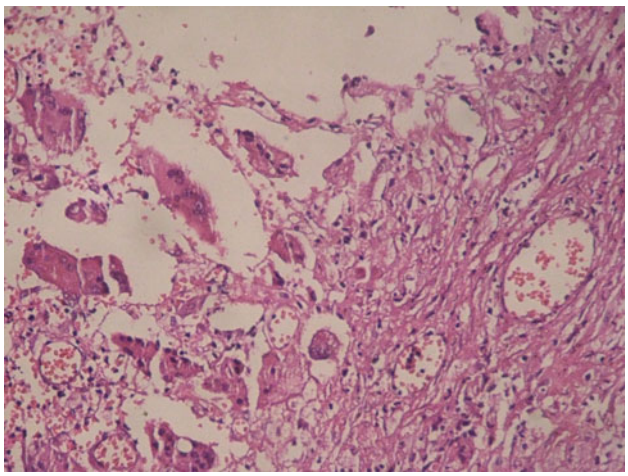

Fig. 5 Fibrous septa made up of fibrocollagenous tissue, red blood cell filled spaces, clusters of osteoclastic giant cells, and foam cells (H&E, ×20)

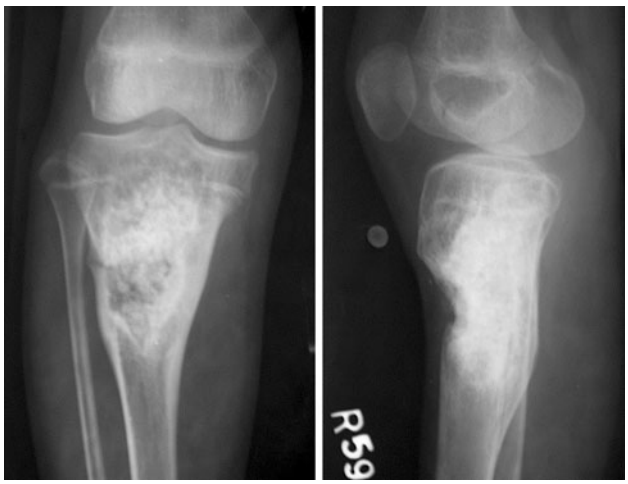

Fig. 6 Follow-up radiographs at 6 months after surgery showing the cyst in healing phase

processing the whole tissue (Figs. 4, 5). Histologically, areas typical of an ABC were identified, along with some broad areas of fibroblastic proliferation, hemosiderin-laden macrophages, and osteoid proliferation (suggestive of fracture healing). Features like a thin fibrous wall lined with flat to slightly plump layers of mesothelial-like cells were not seen, thereby excluding the possibility of a simple bone cyst. There was no evidence of malignancy in any of the sections examined. Hence the diagnosis of primary ABC was established.

The patient was assigned to regular clinicoradiological follow-up in order to check for any shortening, angular deformity or recurrence, as well as to monitor the extent of healing and physeal growth. Radiographs taken at 6 months (Fig. 6) and 48 months (Fig. 7) after surgery

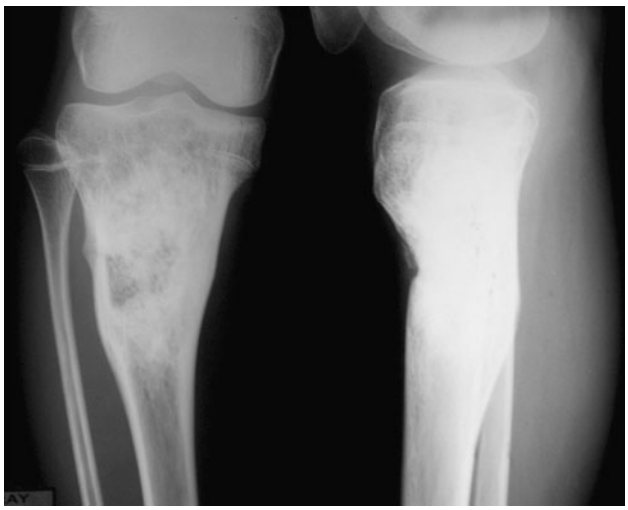

Fig. 7 Follow-up radiographs at 48 months after surgery showing the healed cyst with a residual radiolucent area. No signs of growth disturbances and recurrence can be seen

showed healing with a residual radiolucent area but without any deformity, shortening, or recurrence. At the last follow-up visit, the patient was free from pain and disability (deformity or shortening). Written, informed consent was obtained from her parents (who had authorized treatment, radiological and photographic documentation). They were also asked if data concerning the case could be submitted for publication, and they consented.

Discussion

Lichtenstein [10] first described ABC as a separate pathologic entity, and rejected previous suggestions that the lesion was an unusual presentation of a giant cell tumor or atypical hemangioma of bone. Nowadays, ABC is considered a hyperplastic lesion of unknown etiology consisting of cystic cavities containing blood lined by a mesenchymal reactive tissue. Histologically, the lesion bears no similarity to either a cyst or an aneurysm [11]. It has been identified as a pseudotumoral condition resulting from a local alteration in hemodynamics, leading to increased venous pressure and the production of a local hemorrhage, with consequent proliferation of a reactive osteolytic tissue, increasing the hemorrhagic process [1].

Campanacci and Capanna [9] reviewed the data from 198 cases with ABCs who presented between 1940 and 1982, and proposed a classification system based on the anatomical location of the lesions, and a staging (aggressive, active, inactive) to assess their activities:

- Type 1: the well-contained cyst occupies the center of the bone with little or no expansion of cortex

Table 1 Details of the reported cases of primary ABC that extended into the epiphysis after crossing the viable physis (juxtaepiphyseal primary ABC)

Reports	No. of cases	Age	Location of open physis invaded by ABC	Type and nature of ABC	Treatment	Follow-up
Carlson et al. [4]	One	Not mentioned	Capital femoral and greater trochanter physis	Type 1 or 2; aggressive	Not mentioned	Not mentioned
Dyer et al. [5]	One	9 years	Invasion of head of fourth metatarsal	Type 2; active	Excision with fibular graft placement	Not mentioned
McCarthy et al. [6]	One	10 years	Proximal fibular physis	Type 2; aggressive	Excision of proximal fibula	No evidence of proximal migration of fibula
Capanna et al. [2]	Nine	6–16 years (seven cases were 13 or more years old)	Proximal humerus (3), metacarpal (2), proximal tibia (2), distal tibia (1), proximal fibula (1)	Type 1 (one case); type 2 (six cases); type 3 (two cases) Aggressive (6 cases); active (3 cases)	Curettage or resection with bone grafting	Healing (all cases), growth disturbances (5 cases)
Rizzo et al. [3]	Five	2–14 years (range of age group across the whole series of 15 cases)	Separate details of cases with epiphyseal extension of the ABC are not mentioned; at least one of them had crossed the proximal tibial physis	Type 2 (four cases); type 3 (one case)	Excision, curettage, and autogenous bone grafting	Separate details of these cases were not provided by the authors
Present case (2010)	One	10 years	Proximal tibial physis	Type 2; aggressive	Intralesional excision, curettage, auto + allogenic bone grafting	Healed

- Type 2: the lesion substitutes the entire segment of the involved bone, which is greatly inflated with cortical attenuation
- Type 3: the cyst has an eccentric metaphyseal location, showing no or minimal expansion of the cortex
- Type 4: the cyst is subperiosteal in a diaphyseal location on long bones, with no or minimal cortical erosion
- Type 5: a lesion of the metadiaphyseal area of the long bone inflates the periosteum towards the soft tissues and simultaneously penetrates the cortex to extend into the cancellous bone.

A primary ABC is defined histologically as anastomosing fibrous-walled channels containing blood. These channels may have a complete or incomplete lining of endothelial cells, but unlike true blood vessels they contain no elastic lamina or muscle layer. Typically, the fibrous walls contain red blood cells, granules of hemosiderin, foreign-body giant cells, and spicules of reactive bone. A secondary ABC has the same microscopic characteristics, but with additional histological findings of a coexisting lesion, either benign or malignant [11]. It has been suggested that the ABC element in such cases is secondary to a pre-existing lesion [12].

Direct extension of metaphyseal lesions across a viable physis is a rare phenomenon, as it acts as a relative—but not complete—barrier to tumor extension. Though the invasion of the physis by osteogenic sarcoma, and metaphyseal osteomyelitis of the granulomatous type or caused by low-grade pyogenic organisms are well known, such an invasion by a benign metaphyseal tumor is considered exceptional [2, 13]. Invasion of the viable physis by a primary ABC has been reported only vary rarely in the literature [2–6] (Table 1). We therefore performed a literature search of such cases, and are now adding a new case.

ABCs that cross the physis may lead to growth disturbances in the form of limb length discrepancy or angular deformities. Thus, such cases pose a therapeutic challenge as physeal plate damage may occur due to invasion of the cyst, or it may be damaged iatrogenically during excision and curettage. Capanna et al. [2] reported growth disturbances in five of the nine of these cases in their series. Rizzo et al. [3] did not notice growth disturbances in any of the patients and concluded that the lack of growth disturbances in their series probably reflects that the patients sought medical attention at an early stage. The results of our case seem to be in tune with the observations of Rizzo et al. We suggest that juxtaepiphyseal primary ABC should

be surgically treated at an early stage, because such cases are likely to produce growth disturbances if allowed to follow their natural histories.

Acknowledgments We deeply appreciate the digital work support of Mr. Abhishek Talwar, and thank him for contributing his valuable time to the completion of this article.

Conflict of interest They did not receive grants from any commercial entity in support of this work. There are no conflicts of interest.

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