Case Report

Primary Aneurysmal Bone Cyst of the Epiphysis

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Abstract Aneurysmal bone cysts are benign active or aggressive bone tumors that commonly arise in the long bones, especially the femur, tibia, and humerus and the posterior elements of the spine. Aneurysmal bone cysts affect all age groups but are more common before skeletal maturity (first two decades of life). They usually involve the metaphysis or metadiaphyseal region of long bones. Although juxtaphyseal lesions abutting the growth plate and extending into the epiphysis have been described, there is no report of an aneurysmal bone cyst entirely and primarily located in the epiphysis. We report on a 3-year-old boy who presented with an entirely contained aneurysmal bone cyst to the proximal tibial epiphysis. We discuss the clinical presentation, diagnosis, including imaging and

pathology, and treatment. A review of the pertinent literature also is presented.

Introduction

An aneurysmal bone cyst (ABC) is a benign but often locally aggressive bone lesion. It occurs more often in the skeletally immature, although there is a wide range of age at diagnosis [6, 12, 13]. An ABC can involve any bone, but it is more commonly located in the metaphysis of long bones and the posterior elements of the spine. It can present as a primary or secondary lesion (eg, associated with chondroblastoma, osteoblastoma, and others). Radiographically, an ABC is a lytic, expansile lesion that often presents with cortical thinning and septations, characteristically showing fluid-fluid levels on MRI. The natural history is one of continued growth and surgical treatment is recommended [7, 18].

Although ABCs typically are located in the metaphysis, owing to the aggressive nature of this tumor, physeal involvement or extension may occur, resulting in growth plate closure or development of angular deformity [4, 14, 17]. However, to our knowledge, primary epiphyseal involvement has not been reported in the literature.

Case Report

A 3-year-old boy, previously healthy, presented to the outpatient clinic with complaints of right knee pain of a few weeks' duration. There was no clear initiating factor, such as trauma. There were no other symptoms or complaints. On physical examination, there was pain and tenderness located over the right aspect of the proximal

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Each author certifies that his or her institution has approved the reporting of this case report, that all investigations were conducted in conformity with ethical principles of research, and that informed consent for participation in the study was obtained.

This work was performed at The Children's Hospital of Philadelphia.

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tibia, just below the articular joint line. There was essentially full ROM of the knee and no masses were palpable. The overlying skin was intact with no changes. The neurovascular examination was normal. The patient walked with a slight limp favoring the contralateral extremity. The remaining physical examination was unremarkable.

The initial radiographs showed a well-defined, eccentric, radiolucent lesion in the proximal tibia epiphysis. CT showed intralesional septations and absence of joint or physal plate disruption. MRI showed fluid-fluid levels caused by the different densities of the cyst fluid owing to the setting of red blood cells (Fig. 1).

The patient underwent open incisional biopsy with intraoperative frozen section. After diagnosis of ABC was confirmed, with no signs of any other associated tumors, including chondroblastoma, he underwent a four-step approach and bone grafting during the same anesthetic event. The four-step approach was described previously [8] and consists of intralesional curettage, high-speed burring, electrocauterization to find any residual cyst wall lining, and 5% phenol painting of the cyst wall. Care was taken to avoid the physis and the joint. Loupes for

magnification, headlamps for illumination, and fluoroscopy for guidance were used during the surgical procedure. The growth plate and articular surface were cautiously avoided during resection. All lesional tissue was sent for pathologic evaluation intraoperatively, which on final histopathologic analysis, showed bland spindle cell proliferation with uniformly scattered multinucleated giant cells, hemosiderin pigment accumulation (confirmed by the Prussian blue stain), fragments of trabecular bone and hyaline cartilage, and abundant blood and fibrin. Mitoses were fairly numerous in the spindle cell proliferation, but no atypical forms were seen. Linear strands of woven bone also were noted, suggesting a cyst lining (Fig. 2). The histologic features were most consistent with those of an ABC. After careful examination of all submitted pathology material, no signs of any associated lesion were seen in the pathology specimen, particularly no features of chondroblastoma, which was high on the differential diagnosis owing to the intraepiphyseal nature of this tumor.

Three months into the postoperative period, the patient had increasing pain develop over the surgical area. New

Fig. 1A–G (A) Anteroposterior and (B) lateral radiographs of the knee show a well-defined, eccentric, radiolucent lesion confined in the proximal tibial epiphysis. (C) Coronal and (D) sagittal CT scans show the extent of primary involvement and close proximity to the articular cartilage and growth plate. (E) Axial, (F) sagittal, and (G) coronal T2-weighted MR images show characteristic fluid-fluid levels caused by the different densities of the cyst fluid attributable to the setting of red blood cells, preservation of the physis, and articular surface.

Fig. 2A–B (A) The initial biopsy specimen shows predominantly solid areas with a few cyst walls (upper left) and abundant blood and fibrin (lower right) (Stain, hematoxylin and eosin; original magnification, $\times 2$). (B) A higher-power view of the tumor shows the bland

spindle cell background and evenly distributed multinucleated giant cells. There are areas of hemorrhage in the tumor with hemosiderin pigment accumulation (middle of picture) (Stain, hematoxylin and eosin; original magnification, $20\times$).

Fig. 3A–E Followup images show recurrence of the ABC. (A) Anteroposterior and (B) lateral radiographs show resorption of the bone graft and recurrence of the lytic lesion. (C) Axial, (D) sagittal, and (E) coronal T2-weighted MR images confirm a recurrent lesion without any evidence of articular cartilage or growth plate arrest.

imaging, including plain films and MRI, were obtained and confirmed recurrence of the lesion (Fig. 3). The patient underwent a more aggressive four-step approach, again avoiding the growth plate and the articular surface. The final pathology report after the second surgery confirmed the recurrent ABC (Fig. 4). Again no features of

chondroblastoma or other processes were noted. Cultures obtained showed negative results. At last followup, 1 year after the second procedure, radiographs showed no signs of recurrence or physal arrest; the patient was pain-free, with full ROM, and had returned to all his previous activities (Fig. 5).

Fig. 4A–B (A) The resection specimen shows even more classic features of an ABC with variably sized cyst spaces, which are lined by lesional cells instead of vascular endothelium. In the walls of the cysts is a bland spindle cell proliferation with uniformly scattered multinucleated giant cells, immature bone and cartilage, hemorrhage,

and hemosiderin deposition (Stain, hematoxylin and eosin; original magnification, $\times 4$). (B) A higher-power view of the lesion shows bland spindle cells and scattered multinucleated cells (Stain, hematoxylin and eosin; original magnification, $\times 10$).

Fig. 5A–B (A) Anteroposterior and (B) lateral radiographs obtained at the 1-year followup show resolution of the lesion with no signs of residual cysts or recurrence. Clinically, the patient was pain free, had full ROM, and had returned to activity without limitations.

Discussion

ABCs occur with an estimated incidence of approximately 0.14 to 0.32 per 100,000 individuals [12, 20]. The tumor most commonly occurs in the metaphyseal region of long bones. Spine lesions also frequently are seen. Although epiphyseal extension has been reported in association with metaphyseal ABCs [17], to the best of our knowledge, this is the first report of a primary ABC entirely contained in the epiphysis [6, 12, 13, 18]. In 1999, a review of 15 juxtaepiphyseal ABCs showed no growth arrest before or after surgical treatment, which consisted of excision, curettage, adjunctive cauterization (in two cases), and bone grafting. Of these 15 cases, three children had a recurrence and the lesion “grew away” from the physal plate [17].

The recurrence rate of ABCs from various surgical treatments reportedly is between 10% and 59% [1–3, 5, 7–10, 15, 16, 19]. It is unclear whether any factors such as

age, gender, location, or histologic characteristics predispose to recurrence. One study suggests a high recurrence rate among very young children [20]. In a review of seven children with an average age of 5.5 years (range, 2.9–10.6 years), recurrence occurred in five of the seven patients. The recurrences appeared rather rapidly after surgical treatment (average, 8 months) [9]. Proximity of the bone cyst to the physis also might be a risk factor for recurrence owing to the difficulty in complete cyst removal in this area [8].

Young children with epiphyseal lesions typically present with pain and gait abnormalities as the initial findings. The differential diagnosis of an epiphyseal tumor depends on the age of the patient along with the findings seen on imaging studies. Among the differential diagnosis in a child presenting with an epiphyseal lesion are chondroblastoma, subacute osteomyelitis (Brodie’s abscess), and giant cell tumor. Subchondral cysts also may be seen in

association with Perthes' disease and other conditions such as inflammatory arthritis. Given our patient's age, the diagnosis of a giant cell tumor seemed highly unlikely, as this tumor is seen more often in the older adolescent age group. Infectious processes likewise can be ruled out based on the pathologic findings seen in our case. The cystic nature of the lesion seen in the diagnostic imaging studies and intraoperatively would point toward an ABC. A primary ABC of the epiphysis has not been reported. In 1986, Ippolito et al. [11] reported on atypical cases of chondroblastoma, which included three cystic types that resembled bone cysts on appearance; final pathology reports for their cases, however, revealed findings consistent with chondroblastoma tissue [11]. In our patient there were no histologic features consistent with a chondroblastoma. Given, the diagnostic, clinical, and histologic findings seen in our patient, we conclude this was indeed a primary ABC of the epiphysis.

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