Research and Reviews: Journal of Clinical and Medical Case Studies

Primary Aneurysmal Bone Cyst of the Ilium in a Pediatric Patient: Case Report and Review of the Literature

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Case Report

Received date: 17/11/2016
Accepted date: 10/12/2016
Published date: 17/12/2016

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Keywords: Aneurysmal bone cyst, Bone tumor, Orthopaedic surgery, Pelvis, Curettage, Benign tumor, Paediatrics.

ABSTRACT

This article presents the case of a 10 year old male patient who began experiencing pain in the region of the left hip causing claudication. Because the pain continued without improvement, pelvic X-rays were obtained in different projections; these revealed a multilobed osteolytic lesion in the area of the ilium, and MRI showed a hyperintense multilobed image filled with liquid. Based on these results, a decision was made for surgical treatment. Excisional biopsy of the lesion was performed with curettage and placement of a bone graft. To date, no tumor recurrence has been reported. Although the existence of aneurysmal bone cysts has been known for several years, these tumors have not yet been defined properly from a histopathological point of view nor has a consensus been reached on the best methods for diagnosis and treatment. For this reason, various surgical techniques have been developed with their respective indications, contraindications and complications for the management of these lesions. It is of utmost importance that the orthopedic surgeons evaluate the patient thoroughly and, based on the characteristics of the tumor, have available the necessary tools for making decisions on what treatment is indicated in that particular patient. The presence of an aneurysmal bone cyst should be suspected in every patient with bone pain and suggestive images, especially in the pediatric population, to permit proper diagnosis and provide correct treatment with the aim of reducing complications and disease recurrence

INTRODUCTION

Aneurysmal bone cysts are benign osteolytic bone tumors associated with pain, inflammation, the presence of a growing mass and pathological fractures; they primarily affect pediatric patients. These lesions are rare, usually affecting the femur, tibia, fibula, humerus or spine. Their occurrence within the pelvis is rare.

CASE PRESENTATION

A 10 year old male patient began experiencing mild pain in the left hip. The symptoms evolved over one year to stinging, localized pain without radiation and without accompanying symptoms and with claudication. The pain increased with physical activity and remitted with rest.

A month after the start of the symptoms, the pain worsened and increased with physical activity. The pain continued to display the same characteristics, causing claudication and cessation of daily activities.

The patient reported no exposure to animals; his vaccination record was complete, and he had no history of chronic or degenerative disease, traumatic injury, surgical intervention, hospitalization, blood transfusion or allergy.

The family history includes type 2 diabetes mellitus in the maternal and paternal branches.

During review of systems, the patient did not present with nervous, cardiovascular, respiratory or digestive symptoms. He mentioned only pain in the area of the left hip together with limited mobility of the lower left limb.

Physical examination revealed a male who looked his age, with freely chosen attitude, without characteristic facies and who

was cooperative, aware and calm. The vital signs were within normal parameters. Examination of the head, neck, chest, abdomen and genitals revealed no alterations. Musculoskeletal exploration revealed a lower right limb without alterations. The lower left limb showed no skin alterations, appropriate coloring and hypotrophy of the femoral quadriceps muscle. Movements of the hip were limited and there was pain on palpation. The femoral, popliteal, tibial and pedal pulses were normal. Reflexes and neurological examination were without alterations.

Anteroposterior pelvic radiography was performed; it revealed a single lesion measuring 10 cm × 15 cm with well-defined edges and an area of sclerosis between the peripheral region of the tumor and the bone, with a pattern of geographical destruction in the left ilium as well as widening of the cortex compatible with the presence of periosteal reaction and radiopaque images inside compatible with septa **(Figure 1)**. Contrasted pelvic MRI was then performed, revealing a multilobed hyperintense image in the left ilium that was well-defined and hyperenhanced compatible with liquid levels. The laboratory studies are described in **Table 1.**

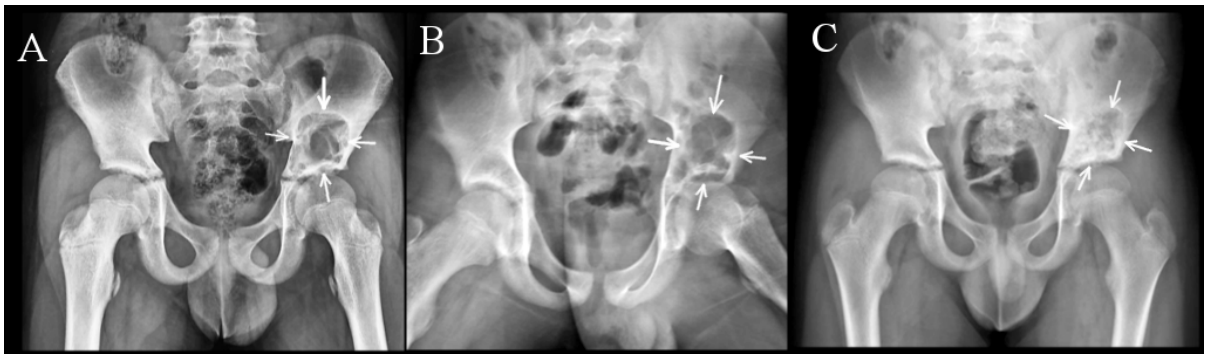

Figure 1. Anteroposterior pelvic radiographs: Preoperative images that show a 10 cm × 15 cm lesion on the left ilium (arrows) with borders well defined by peripheral sclerosis between the tumor and the bone, with geographic destruction pattern as well as thickening of the cortical wall. Findings on this image also suggest interior septae, consistent with an aneurysmal bone cyst (A, B). Postoperative radiographs of the pelvis show curettage and placement of bone graft at the tumor site (arrows). No evidence of recurrence is seen (C).

Table 1. The laboratory studies are described below.

Study	Patient results	Reference range (Pediatric) [1,2]
Glucose	106	75-115 mg/dl
Urea	15	5–25 mg/dl
Creatinine	0.6	0.12–1.06 mg/dL
Leukocytes	9	5.0–14.5 × 10^3/µL
Neutrophils	60	33–76%
Hemoglobin	15	11.5–15.5 g/dl
Hematocrit	45	35–45%
Platelets	224	150,000–450,000/µL
Alanine aminotransferase	42	10–35 U/L
Aspartate aminotransferase	20	10–60 U/L
Prothrombin time	12	11.1–13.1 s
Thrombin time	20	15.0–19.0 s
Partial thromboplastin time	27.4	22.1–35.1 s
Alkaline phosphatase	933	135–530 U/L
Rheumatoid factor	<20	<30.0 IU/ml
C reactive protein	8.02	<1.0 mg/mL
Globular sedimentation rate	10	0–20 mm/h

Based on the clinical, radiological and laboratory findings, a diagnosis of tumor in the left ilium, probably an aneurysmal bone cyst, was established [1,2]. It was decided that surgical treatment would be performed; the patient was taken to surgery, where excisional biopsy was performed with curettage of the lesion and placement of a bone graft. The patient progressed adequately and remained free of disease recurrence until the last medical assessment.

DISCUSSION

Aneurysmal bone cysts were described for the first time by Jaffe and Lichtenstein in 1942. These bone lesions are rare, benign and osteolytic. They are usually associated with pain, inflammation or the presence of a growing tumor [3-5]. In 2002, the World Health Organization defined aneurysmal bone cyst as "a benign bone cyst composed of spaces occupied by blood separated by

septa of connective tissue containing fibroblasts, osteoclast-type giant cells and reactive bone tissue" [6].

In 70% of cases, aneurysmal bone cysts are considered primary, whereas the remaining 30% appear following the development of other tumors; the latter cysts are considered secondary [3,7].

Aneurysmal bone cysts mainly affect persons in the first two decades of life, i.e., they have a high prevalence primarily in the pediatric population [3]. The sites most frequently affected by this pathology are the femur, tibia, fibula, humerus, skull and spine, but any other bone can be affected [3,8-10]. With regard to the spine, the lumbar region is most often affected, followed by the thoracic, cervical and sacral regions. The presence of these tumors in the spine can result in pathological fractures and neurological complications of the spinal cord [3,8-10]. Aneurysmal bone cysts of the iliac crest and ilium are extremely rare. Agarwal and colleagues reported the case of an 11 year old female patient in whom the disease began with increased volume and paresthesia in the region of the right hip. A clinical-radiological diagnosis of aneurysmal bone cyst was made and the tumor was subsequently removed by intralesional curettage and comprehensive resection of the soft tissue. At the 18-month follow-up, the patient remained asymptomatic without recurrence of the tumor and without postoperative complications [11].

Primary aneurysmal bone cysts have a low incidence, ranging from 0.14 to 1.4 cases per 100,000 persons [12]. Aneurysmal bone cyst can also appear in adults, although the highest prevalence of this lesion occurs between 12 and 13 years of age, and 80% of cases occur before 20 years of age [8,13,14]. Males are more commonly affected than women, with the ratios ranging from 1:1 04 to 01 8:1 [3,9]. Aneurysmal bone cysts constitute 1 to 6% of benign primary bone tumors [4,7,8,12]. In approximately 8.4% of cases, tumors can be found in multiple bones in the same patient, and only 8 to 12% of cases present with a tumor in the pelvis [7,11,14].

The pathophysiology of these benign tumors is still unknown, although there are several theories about possible vascular, traumatic or genetic origins [8,12]. Authors such as Campanacci describe the development of this lesion as a response to intraosseous bleeding, which leads to the development of the cyst. Lichtenstein proposed that the aneurysmal bone cyst is a reactive lesion in which vascular alterations in the bone cause an increase in intraosseous pressure, causing local destruction and bone distension [8,12]. The traumatic theory is based on the idea that this type of lesion may develop after a fracture because the fracture can alter intraosseous circulation, with the lesion forming secondarily, as was proposed by Ratcliffe and Grimer. Recent genetic studies have proposed aneurysmal bone cyst as a primary tumor of bone and not as a reactive process secondary to a local bone problem. Genetic mutations, especially T(16;17) have been detected, and these could confirm the tumor etiology of the lesion [3,8,15]. A familiar pattern of pathology of these lesions has been described by some authors [12].

The progression of aneurysmal bone cyst is divided into four phases. The first of these is the initial phase, which is described as osteolysis of the marginal part of the bone with discrete elevation of the periosteum. The second phase, called the growth phase, is characterized by progressive destruction of the bone. This begins the stabilization phase, which features an expansive lesion with intratumor bone separations. The last phase is the healing phase, during which progressive ossification of the lesion occurs [8,12].

Patients with primary aneurysmal bone cyst generally present initially with pain in up to 65% to 93% of cases, with soft tissue inflammation in 15% to 18.1% of cases and with a tumor that grows over time, although in 3.4% to 25.9% of cases, the initial symptom is a pathological fracture or alterations in growth due to effects of the tumor on growth cartilage [4,8,12,13]. Constitutional symptoms such as fever, weight loss, nausea, changes in general status and vomiting are uncommon. As mentioned earlier, the most commonly affected sites are the femur, tibia, humerus and fibula; these account for 52% of the affected body regions. Less commonly affected areas include the small bones of the hands and feet [15]. In the pediatric population, the femur and tibia are the most affected, especially in the metaphyseal regions [3]. Aneurysmal bone cysts affecting the pelvis can extend to the endopelvic tissues, causing abdominal and urinary symptoms [7].

The multiple classification systems for aneurysmal bone cysts include the classification of Enneking, who divides these lesions into three stages: 1) inactive, the stage at which the tumor is most benign because the lesion is well demarcated, there is little tumor expansion, and there is minimal inflammation and periosteal reaction; 2) active, the stage at which the lesion produces more symptoms, there is tumor expansion with cortical thinning, and a layer of bone appears that separates the healthy bone from the tumor and shows the classic "soap bubble" sign; and 3) aggressive, in which the lesion expands rapidly, destroying the surrounding tissue and producing severe symptoms [3,14]. Another classification based on morphology is that proposed by Capanna, who divides the lesions as follows: type I lesions, which are centrally located and well contained; type II lesions, which are more expansive and involve cortical thinning with involvement of the entire bone segment; type III lesions, which are eccentric, metaphyseal and generally affect only one cortex; type IV lesions, which are subperiostic and expand away from the bone; and type V lesions, which develop in the periosteum and expand to the periphery, subsequently entering the cortical bone [3,14].

When aneurysmal bone cyst is suspected, the first study to perform is a simple X-ray of the affected area. The classic radiographic aspect of aneurysmal bone cyst is a radiolucid cystic lesion in the metaphyseal portion of the bone. The lesion can be osteolytic, and it is possible for it to extend into the surrounding cortical bone; the lesions are generally contained within the cortex of the bone but can also elevate the periosteum. Lesions in the epiphysis should raise suspicion of a malignant tumor [3,7,9,12]. Another effective means of detection is CT, which contributes particularly to preoperative planning, the delimitation of the lesion and

the identification of pathological fractures. We find that contrasted MRI can show the internal separations and the presence of intratumoral liquid, which is indicative of the presence of blood, and that these features are better observed in the T2 sequence. Although this finding is not pathognomonic, it is highly suggestive of the presence of an aneurysmal bone cyst [3,7,9,12].

Histopathological study of the lesion with the aim of ruling out other tumor lesions, especially malignant ones, is of utmost importance. The classic histopathology of aneurysmal bone cyst is a cavernous vascular tumor of variable diameter with intratumoral communicating cavitations, without clots and without an epithelial coating [11]. Microscopic study of the lesion can reveal hemorrhagic tissue with cavitary spaces separated by fibrous septa containing inflammatory cells and giant cells [5]. Currently, incisional biopsy is recommended as the standard for diagnosis because in some cases, fine needle aspiration biopsy yields an insufficient sample because the material obtained consists only of blood elements [3,15].

Differential diagnosis of aneurysmal bone cyst must be performed to distinguish it from other benign bone tumors, among which unicameral bone cysts, fibroid chondromyxoid, giant cell tumor, osteoblastoma, chondroblastoma, hemangioma, non-ossifying fibroma, fibrous dysplasia and eosinophilic granuloma, among others, should be considered. Similarly, aneurysmal bone cyst should be differentiated from bone tumors or malignant soft tissues, especially telangiectatic osteosarcoma [3,15].

Once the diagnosis is established, treatment usually consists of intralesional curettage that may be accompanied by graft, bone cement or bone substitutes and may or may not include adjuvant therapy; the latter aims to eliminate microscopic deposits where the tumor was located to reduce the risk of recurrence [3,13]. A new treatment is so-called "curopsy" ("biopsy with intention to cure"); this procedure is similar to a biopsy and has the diagnostic and therapeutic goal of limited percutaneous curettage, which removes the membrane coating multiple quadrants of the cyst and leads to consolidation [6,15]. Another modern technique is endoscopic curettage; the use of this technique in cases of aneurysmal bone cysts of the patella, proximal humerus, proximal tibia and calcaneus has been described [16]. The authors did not place a bone graft and did not observe recurrence in any of the cases during follow-up. The main advantages of this procedure are aesthetics (small incisions are used), preservation of the bone structure (the bone cortex is minimally affected) and the fact that intracystic observation is better, allowing more complete curettage and thereby reducing the risk of recurrence and causing less disruption of the soft tissue, which allows earlier rehabilitation [16]. Broad resection with reconstruction is indicated for lesions that have destroyed the metaphysis or are located in periarticular areas. In large lesions located in the axial skeleton, selective preoperative arterial embolization minimizes intraoperative bleeding. The spontaneous resolution of aneurysmal bone cysts has been reported, even if the treatment is merely surgical [3].

The treatment of aneurysmal bone cysts in the pelvis represents a major challenge due to the complexity of the anatomical region, the proximity to neurovascular structures, the vulnerability of the sacroiliac and coxofemoral joints and because tumors in this region are generally large and extremely vascularized. During the surgical treatment of aneurysmal bone cyst in this region, attempts should be made to preserve the stability of the joints and the structural integrity of the area, especially of the acetabulum [7,11]. Some authors recommend curettage of the lesion in cases of primary aneurysmal bone cysts and reserve the use of broad resection for cases of recurrence or for lesions located in expendable bones that can be surgically removed without causing high morbidity or without the need for a reconstructive procedure [13,17]. Lesions in the proximal or medial segment of the fibula, clavicle or body of the scapula can be treated only with resection, whereas lesions of the foot can be treated with resection and arthrodesis using an autograft [15].

Block resection has proven to be the best method to reduce recurrence; however, this procedure cannot always be performed, mainly due to the location of the lesion. To avoid the risk of pathological fracture and instability, it is important to take into account the integrity and stability of the site from which the tumor was removed, as there is the possibility of internal fixation [3].

The use of cryotherapy has been reported in the treatment of aneurysmal bone cyst, and this method has been shown to reduce the recurrence of these lesions. Sclerotherapy, which is based on the vascular theory previously mentioned, has also been used successfully; if hemodynamic alterations of the tumor can be controlled, it can be resolved. Because sclerotherapy is a non-invasive method, patients treated with this technique have better functional results and fewer complications, although the recurrence rate is the same as with surgery. Another recently used method that has given excellent results is ablation by the injection of radionuclides into the lesion. Similarly, coagulation with an argon laser has been used to reduce the recurrence of this type of cyst [6]. Chemical ablation with phenol, hydrogen peroxide, liquid nitrogen, polyvinyl alcohol, cyanoacrylate, percutaneous injection of steroids, calcitonin and fibrosing agents such as Ethibloc (Ethicon) has also been reported, as well as the placement of aspired bone marrow [4,6-8,13]. The problem with percutaneous injections of these agents is the high risk of recurrence and partial healing because of the presence of intracystic septa [12].

It has been reported that recurrence of aneurysmal bone cysts occurs in 10% to 59% of cases within the first 24 months after surgery [8,13]. The risk factors for recurrence are as follows: a young patient (age <12 years), the presence of open physis, lesions adjacent to the growth cartilage, the predominance of certain cell types on histopathological analysis and the mitotic index [8,13]. With respect to curettage, there is a lower rate of recurrence when a high-speed drill is used [12]. A small number of malignant transformations of aneurysmal bone cysts have been reported, especially when radiation is used in addition to other treatment. There have been cases of malignant transformation to telangiectatic osteosarcoma and fibroblastic osteosarcoma [3].

CONCLUSION

Aneurysmal bone cysts are benign osteolytic bone lesions that predominantly affect the pediatric population. Because these tumors have a low incidence, doctors often lack clinical suspicion of their presence. Aneurysmal bone cysts can affect any bone but mainly affect long bones such as the femur, tibia, fibula and humerus. Structures such as the spine, pelvis and small bones of the hands and feet can also be affected. Although several theories of the physiopathology of these lesions have been advanced, to date there is no consensus on the development of these tumors because no theory has been verified. The symptoms are usually nonspecific, with patients initially presenting with pain, inflammation, increased volume and/or pathological fractures. The diagnosis of these lesions should begin with clinical suspicion based on knowledge, followed by simple radiographs in which a radiolucid cystic lesion, which may be osteolytic and expand into the surrounding cortical bone, is observed. Subsequently, other imaging studies such as CT scanning and MRI should be conducted with the aim of performing preoperative planning. Next, it is desirable to obtain an incisional or excisional biopsy for histopathological diagnosis and to exclude other benign or malignant tumors, as the therapeutic plan and prognosis would differ. After obtaining diagnostic confirmation, the doctor must choose the most appropriate treatment for the patient based on the characteristics of the tumor. Depending on the treatment performed, recurrence in these cases can be high; thus, patients with aneurysmal bone cysts should receive long-term follow-up. More research must be performed with the aim of establishing the best diagnostic and therapeutic methods for these lesions and to help determine the physiopathological basis for the development of these tumors. It is equally important that all medical specialties know about this topic and that physicians have a high level of suspicion in patients presenting with the previously mentioned symptoms and refer them to an orthopedic surgeon for precise diagnosis and appropriate treatment. In the case presented here, the patient was subjected to a surgical procedure with adequate outcome and without presenting any recurrence.

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