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CASE REPORT Open Access

ANEURYSMAL BONE CYST OF DISTAL FEMUR PRESENTING WITH PATHOLOGIC FRACTURE IN A PEDIATRIC PATIENT.

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Abstract

Aneurysmal bone cysts are benign but locally aggressive tumors, with 80% diagnosed in patients younger than 20 years. We report a case of a 7-year-old male who presented with pain in his right thigh due to a minor injury and was diagnosed with a pathologic fracture in the distal femur. The fracture was fixed with plate and screws augmented with antibiotic-impregnated bone cement, and the histopathological analysis confirmed the diagnosis of aneurysmal bone cysts.

Keywords: Bone Cyst, Aneurysmal, Pathologic Fracture, Paediatric.

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Introduction

Aneurysmal bone cysts (ABCs) are benign but locally aggressive tumors, with 80% of cases diagnosed in patients younger than 20 years of age. Approximately 36% of patients will present with a pathological fracture, usually in active lesions. Pathological fractures should be suspected in pediatric patients when there is minimal trauma, unusual location, or abnormal processes in the bone. [1] Because ABCs primarily manifest in pediatric patients, growth plate involvement and permanent limb length deformities are of great concern.[2,3]

The treatment concepts of ABCs have evolved over the years, with resection being the standard of care in most cases. Adjuvant treatments have been used due to more than 50% local recurrence rates, such as polymethyl methacrylate (PMMA) bone cement, argon beam, phenol, ethanol, and cryotherapy. Less invasive methods such as aggressive biopsy, selective arterial embolization, sclerotherapy with ethic or polidocanol, and systemic therapy with Receptor activator of nuclear factor kappa-B ligand (RANKL) inhibitors have also been tried. This study compared the results of a series of patients treated by curettage with and without adjuvant phenol treatment and less invasive interventions.[3]

Case Report

A 7-year-old boy presented with significant pain in his right thigh caused by a trivial injury (he fell and stumbled at home). Before this injury, the patient had no complaints in his right leg.

He had a closed injury over his right lower thigh on clinical examination. A palpable defect in the shaft femur and crepitus was present. The thigh was tender, and the transmitted movements were absent. The neurological examination was unremarkable, and distal pulses were present. A plain radiograph of the right femur showed a solitary, ill-defined, expansile lesion with thin-walled cavities on anteroposterior and lateral views (Figure 1a).

The patient was managed by open reduction and curettage of bony defects for histopathological analysis. The fracture was fixed with a plate and screw and filled with antibiotic-impregnated bone cement (Figure 1b). The sample was sent for histopathological examination. Histology showed blood-filled cystic spaces and multinucleate osteoclastic giant cells separated by fibroblast and reactive woven bone, thus confirming the diagnosis of ABCs (Figure 2). The patient was mobilized non-weight bearing until three weeks post-operative. The Range of Motion rehabilitation was done on the first day after the operation to reduce the risk of knee stiffness. Serial radiographs were done in the 1st month, 3rd month, 6th month and 12th month after operation. The patient showed no recurrence at 1-year follow-up.

Figure 1: (a) Pre Operative radiograph showing right distal femur with bone cyst and pathological fracture and (b) Post Operative Radiograph showing nail and plate fixation. (PA and Lateral view)

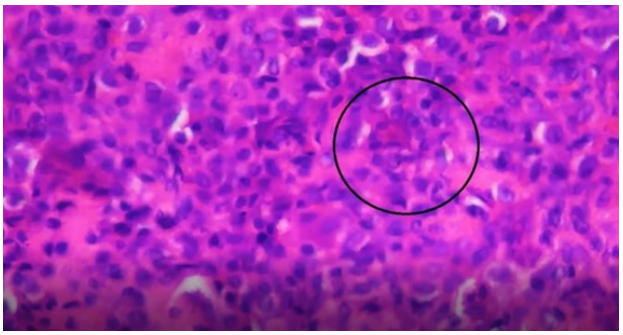

Figure 2: Histopathology Analysis showed bone tissue with osteoblastic rimming accompanied by extensive bleeding areas and osteoclast type giant cell (Black circle)(H&E Stain, 100x Magnification).
Click on image to enlarge)

Discussion

ABC is a benign, locally aggressive tumor in childhood and early adulthood. It usually affects the metaphysis of long bones but can also occur in the spine and pelvis. [4] The etiology of ABCs is still controversial. The pathophysiological process of aneurysmal bone cyst formation is not fully understood. Multiple theories have been proposed, including local vascular alteration, failure in bone maturation, and chromosomal modifications. Local vascular alteration may be due to preexisting bone tumors, failure in vascular repair due to traumatic injury, or vascular malformation in bone. In the 1990s, a study by Kransdorf and Sweet proposed that ABCs are secondary changes to a vascular malformation caused by the evolutionary alteration of a preexisting bone tumor.[5] This leads to the formation of a second lesion in the bone, masked by the primary bone pathology. This process consists of four phases: osteolysis, growth and expansion, healing, and ossification. Another study in the late 1990s found that some ABCs have a genetic component that triggers the appearance of these tumors.[6] The ABCs can develop as a primary or secondary lesion associated with other bone diseases.[5]

The ABC primarily occurs in the second decade of life and may not be included in initial differential diagnosis, especially in older patients, as over 90% of ABC occurs before 20 years, and younger age can complicate management with increased recurrence risk.[7,8] The patient's neurological examination was unremarkable, and distal pulses were present. Pain and swelling of variable duration at the site of involvement are the most common symptoms in patients. Rarely does the initial presentation involve pathologic fracture, specifically in the significant long tubular bones of the extremities.[9]

The diagnosis of ABCs requires the correlation of clinical, radiographic, and histologic findings to distinguish the primary from a secondary form of the disease. The radiographic features of ABCs are pretty distinct and aid in diagnosing the condition.[9] The patient's plain radiograph of the right femur showed a solitary, ill-defined, expansile lesion with thin-walled cavities on anteroposterior and lateral views. The other modality that can be used is a CT scan, which would be a more appropriate imaging investigation for every patient because of specific characteristics of their medical history that helped to restrict the diagnosis of a bone mass. ABCs show up as aggressive, expansile, lytic lesions with a sclerotic ring resembling an "eggshell" on a CT scan.[8]

In 1986, Campanacci criteria were used, a radiographic classification system for ABCs.[10] These criteria differentiate five types based on their radiographic appearance: Type 1, which is centrally arising and can spread to the bone's cortical edge; Type 2, which is centrally located and replaces the entire bony segment; Type 3, which occurs eccentrically; type 4, which is subperiosteal and lifts the periosteum circumferentially, and Type 5, which is subperiosteal and elevates the periosteum circumferentially but destroys adjacent cortical and cancellous bones as they enlarge. These criteria provide a comprehensive approach to managing ABCs and predicting outcomes.[10]

The ABC treatment method has developed over time. The standard of care for ABC has been curettage with or without bone grafting of the defect. However this method of care is burdened by recurrence rates of approximately 25%–31%.[11] The usage of local adjuvants has been introduced in an attempt to reduce the risk of recurrences, such as the usage of cryotherapy with liquid nitrogen, phenol, PMMA cementation, doxycycline foam ablation, or aggressive curettage by using a high-speed burr. Our case demonstrates Type 1 ABC, comprising 23% of ABCs, using curettage through a bony window as recommended treatment, retaining the Campanacci criteria as a helpful classification method.[10] The patient was managed by open reduction and curettage of bony defects for histopathological analysis. The sample was sent for histopathological examination. Histology showed blood-filled cystic spaces and multinucleate osteoclastic giant cells separated by fibroblast and reactive woven bone, thus confirming the diagnosis of ABCs. ABCs are almost always received as curetted material rather than an en-bloc resection. As a rule, the curetted material must be entirely submitted for histologic evaluation. The cysts generally lack any lining, but some flattened endotheli-

al-like cells can be present. Osteoclast-type giant cells are found in clusters with increased numbers within the cyst wall.[9] Giant cell tumors, fibrous dysplastic tumors, chondroblastomas, chondromyxoid fibromas, and non-ossifying fibromas are among the additional lesions that are frequently linked to ABCs.[8]

Although the overall prognosis of ABCs is good, the goal of any treatment modality is to slow down the disease progression, symptom relief, and fixation or prevention of pathologic fracture[8]. Our patient underwent open reduction and curettage of bony defects, fixation with a plate and screw and filled with antibiotic-impregnated bone cement. Traditional ABCs treatment is intralesional curettage with or without bone grafting, sometimes enhanced by additional techniques, such as cauterization, phenol, argon seed coagulation, and cryotherapy.[12] Despite this technique, local recurrence of the disease is relatively high.[13] Regarding treating pathological fractures, some authors suggest that a formal histopathological diagnosis should be obtained before further intervention.[7] Since more severe pathologies may not be excluded initially, even after an open biopsy or needle of suspected ABCs. Treatment options for pathological fractures primarily include intramedullary nails, plates, or prostheses with respective advantages and disadvantages based on the region of bone lesion.[14]

In the last procedure, the patient was mobilized non-weight bearing until three weeks post-operative. In the study by Mendon et al., after surgery, the patient was instructed to bear no weight for four weeks, at which point full weight bearing and mobilization were initiated. The patient underwent routine follow-ups 1, 3, 12, and 18 months after surgery to check for recurrence.[15] Post-operative weight-bearing (WB) is restricted in lower-extremity fracture treatment to prevent failure and poor fracture fixation. Early weight bearing is recommended in patients to restore walking ability and avoid muscle mass loss. However, post-operative WB restriction is associated with pneumonia and prolonged hospital stay, particulary in older patients.[16]

Conclusion

ABC of distal femur can be managed successfully by curettage of the lesion, internal fixation and filling of defect by bone cement. Long term followup is essential in children to detect any recurrence or growth disturbance early.

Abbreviations

ABCs	Aneurysmal bone cysts
CT	Computerised tomography
PMMA	Polymethyl methacrylate
RANKL	Receptor activator of nuclear factor kappa-B ligand
WB	Weight bearing

Declarations

This study received no external funding. All data are available in the article, and no additional data are required. The authors declare that there is no conflict of interest.

Consent

The patient's parents provided written informed consent to be published as a case report.

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