Case Report

Giant aneurysmal bone cyst of the scapula: A case report☆

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A B S T R A C T

Aneurysmal bone cyst (ABC) is a benign bone tumor affecting mainly children and young adults. It occurs in the metaphysis of the long bones. The scapula is a very rare location. Imaging may be highly suggestive of ABC in cases of an osteolytic, expansive, and hemorrhagic lesion with fluid-fluid levels and thin septa. The diagnosis must systematically be confirmed by performing a biopsy, in order to adopt the best therapeutic strategy. There are several therapeutic means, but wide resection remains the gold standard. The evolution is very variable and can go from spontaneous healing to recurrence with the destruction of the bone. We report a rare case of aneurysmal bone cyst of the scapula in a young patient.

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Introduction

Aneurysmal bone cyst (ABC) is a benign, osteolytic, expansive, and hemorrhagic lesion. It is more frequent in patients under 20 years old. It may be a primary bone lesion or associated with other tumors. Imaging is very characteristic and biopsy remains essential to make the diagnosis, and adopt the best therapeutic strategy. We present a special location of ABC in the scapula, which ended up undergoing a total scapulectomy.

Case presentation

Our case concerns a 20-year-old woman with no remarkable history who has been suffering from painful swelling of the
left shoulder growing aggressively for 2 years before her first consultation. The locomotor examination revealed a mass on the anterior side of the left shoulder, about 8 cm in length, with no inflammatory signs (Fig. 1). On palpation, the mass was painful, adherent to the deep tissues and mobile to the skin. Active mobility of the shoulder was impossible, and arm extension was limited to 90° in passive. No axillary adenopathy was found. The biological examination was unremarkable.

The patient underwent an X-ray of the left shoulder, which showed a deformed scapula by a mixed lytic and condensing process taking the entire piece of bone, respecting the humeral head and the clavicle (Fig. 2). To better characterize this mass, a computed tomography (CT) with and without contrast was performed, showing an expansible osteolytic lesion with thin sclerotic margins and bony septa delimiting varying sizes of spaces filled with multiple fluid lines (Fig. 3). Cortical breaches were found in different places without extension into soft tissues. It measures 116 × 56 mm. The exploration was completed by magnetic resonance imaging (MRI) finding the process of the left scapula involving the coracoid process, glenoid cavity, neck, body, and spine of the scapula, deforming and blowing the cortex with a surrounding rim of low T1 and T2 signal. The lesion was multilocular with fluid-fluid levels and peripheral enhancement (Fig. 4). A biopsy of the mass was performed confirming the diagnosis of ABC. After a multidisciplinary consultation meeting, it was decided to perform an embolization of the scapula’s vascular axes followed by a total scapulectomy with the suspension of the humerus. The postoperative course was simple with no sign of infection.

**Discussion**

ABC is a relatively rare benign bone tumor that occurs mainly in young subjects. It represents about 1% of biopsied bone tumors according to Jaffe and Lichtenstein [1]. It can occur at any age but preferentially affects young subjects under 20 years old as in our case [2]. There is a discrete female preponderance [2–4]. It can affect any bone of the skeleton but mainly the long bones, then the axial skeleton, the flat bones, more rarely the
hands and feet. The scapular location is relatively rare (2%) which makes the particularity of our case [5]. Different theories have been suggested for the pathogenesis of ABC. Some authors have proposed that ABC is more of a reactive lesion, and the vascular disturbances in the bone induce an increase in intraosseous pressure, resulting in local destruction of the bone [6]. Recently, exceptional cytogenetic abnormalities have been described but remain isolated, leading some authors to believe that ABC is a true neoplasm and not just a reactive process [7].

Clinically, ABCs are often revealed by pain or swelling, and more rarely by a pathologic fracture [5,8]. Radiological features of ABCs include eccentric, osteolytic lesion, with periostial calcification, which gives the appearance of “soap bubbles.” Aggressive forms may be manifested by loss of cortical contours or apparent extension into the soft tissues, simulating a malignant lesion [10]. Often, it is difficult to make the diagnosis and additional imaging is necessary [5,9]. CT scan provides lesion mapping, especially in complex areas such as the pelvis and spine [9]. It can show in a third of the cases the typical image of fluid-fluid levels, which are the result of the separation of serum from blood cells [5,9]. MRI is the most powerful method to characterize ABCs. It can typically show a heterogeneous, lobulated or multiseptated mass. Fluid-fluid levels are very suggestive but not specific [10]. They are present in 66% to 84% of cases [11,12]. They appear hyperintense on T2, and hypointense on T1-weighted imaging. Gadolinium injection shows enhancement of the cyst walls and internal septa. Fluid-fluid levels are mainly present in the expansion and stabilization stages but absent in the initial phase [12].

The presence of septation is much more consistent [11]. The differential diagnosis is that of a metaphyseal osteolytic lesion: unicameral bone cyst, giant cell tumor, telangiectatic osteosarcoma, osteoblastoma, hemangioma, chondroblastoma, and chondromyxoid fibroma. Biopsy is essential to establish the diagnosis of ABC, and to eliminate other differential diagnoses and thus adapt the treatment [9].

Once the diagnosis has been established with certainty, it is recommended to wait 4-6 weeks after biopsy before treating ABCs, to allow the trepanation hole to fill and sometimes the cyst to involute [8]. Wide resection is the only therapeutic method that can reliably achieve a cure. Radiotherapy is effective, but it is associated with the risk of malignant transformation, which limits its use to a few exceptional cases, such as recurrent spinal lesions not accessible to any other treatment [13]. Embolization alone is an option to treat some ABCs, especially of the spine and sacrum [14]. Preoperative embolization is useful to decrease the risk of bleeding [8,13]. Other therapeutic methods have been developed. According to a 2010 randomized study by Varshney, polidocanol sclerotherapy would provide comparable cure rates to surgery, with fewer complications [15]. Absolute alcohol has also shown good results [16]. Denosumab can be proposed for the treatment of ABCs, in particular spinal ABCs [17]. Surgical treatment remains the gold standard [8,13,15]. Wide resection guarantees the absence of local recurrence but at the cost of reconstruction issues and potential complications, that are not justified by the benignity of the ABCs [9]. In less aggressive forms, subperiosteal resection reduces the risk of local recurrence compared to simple curettage, and preservation of the periosteum favors reconstruction [9]. The evolution of ABCs is very variable; it can go from recurrences after adequate treatment to spontaneous cures in other cases.

**Conclusion**

ABC is a benign lesion that mainly affects children and young adults. Its prognosis is generally good. The imaging appearance may be highly suggestive of ABC in cases of an expanse lytic process with eccentric metaphyseal topography, presenting fluid-fluid levels and separated by thin septa. When this typical aspect is not complete, the diagnosis cannot be made by imaging alone, and biopsy is the key. The treatment of choice is resection of the tumor with wide excision, followed by immediate reconstruction when stability is compromised.
Patient consent

Informed consent was obtained from the patient.

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