# Aneurysmal bone cyst of rib presenting as a huge chest wall mass

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**Aneurysmal bone cyst is a rare benign tumor of the bone that can be difficult to distinguish from malignant tumors, especially when it presents in an unusual location. Herein, we report a six-year-old girl with a primary aneurysmal bone cyst in an uncommon location. It originated from the 4th rib and she presented with a huge chest wall mass. Despite the large size of the cyst, the only symptom was persistent cough. She was successfully treated with total excision of tumor without any complication. The patient has been followed up for 56 months without any recurrence.**

***Key words: aneurysmal bone cyst, rib, chest wall tumors, childhood.***

Chest wall masses occur very rarely in children and have a wide range of differential diagnoses, including neoplastic and non-neoplastic disorders. In any child presenting with chest wall mass, malignant neoplasms such as soft tissue sarcomas and bone tumors should be considered and ruled out. Benign tumors and tumor-like lesions including simple bone cysts, aneurysmal bone cysts (ABC), giant cell tumors, chondroblastoma and cartilaginous hamartoma of the ribs should also be included in the differential diagnosis.

Aneurysmal bone cyst is a rare benign bone tumor, accounting for 1-2% of all primary bone tumors[1,2]. It is an expanding osteolytic bone lesion consisting of blood-filled spaces that are separated by connective tissue septa containing trabeculae of bone or osteoid tissue and osteoclast type giant cells[3]. Any bone may be affected but it rarely originates from the ribs[3-6]. Herein, we report a child with primary ABC originating from the 4th right rib that presented as a huge chest wall mass.

## Case Report

A six-year-old girl was admitted with a non-productive, persistent cough for two months that was more pronounced in the supine position. There was no history of fever, anorexia, weight loss, night sweating or respiratory distress. She received two courses of per oral antibiotics for three weeks before admission without any regression of her complaints. Her past medical history and family history were unremarkable.

In physical examination, the respiratory sounds were markedly diminished, and crackles and rhonchus were heard over the upper half of the right chest. There was no respiratory distress and other physical findings were unremarkable. Laboratory investigations showed hemoglobin 8.9 g/dl, leukocyte 7,900/mm$^3$, platelet 287,000/mm$^3$, and a differential of 60% polymorphonuclear leukocytes, without any malignant cells. Erythrocyte sedimentation rate, serum electrolytes, lactate dehydrogenase, uric acid levels, and liver and kidney function tests were all in normal ranges.

This case was presented as a poster presentation in the Turkish Pediatric Oncology Group (TPOG), 13th National Pediatric Cancer Congress, 18-22 May 2004, in Cappadocia.

Chest X-ray revealed a huge mass within the upper right thoracic cavity that was minimally displacing the trachea to the left and a lytic bone lesion was observed at the 4th right rib (Fig. 1). A multilobulated cystic mass lesion (8.5 x 8 x 6.5 cm), originating from the 4th rib with bony structure destruction, compression of inferior part of the trachea and the right main bronchi, and compressive atelectatic changes in the adjacent lung parenchyma were seen on computed tomography (CT) scans (Fig. 2). There were no other destructive changes at the other bony structures. The main mediastinal vascular structures were normal, and there was no mediastinal or hilar lymphadenopathy. A T2-weighted magnetic resonance imaging (MRI) scan revealed fluid-fluid levels in this multilobulated cystic lesion (Fig. 2). The internal content of the mass was hyperintense on T1-weighted MRI scans, which referred to a proteinous and hemorrhagic fluid collection (Fig. 2). There was no solid component in this mass lesion.

The presumptive diagnosis for this case was ABC, but other disorders like giant cell tumor, telangiectatic osteosarcoma, chondroblastoma, and cartilaginous hamartoma of the ribs could not clearly be ruled out based only on clinical and radiological findings. She underwent surgery for both diagnostic and therapeutic intents.

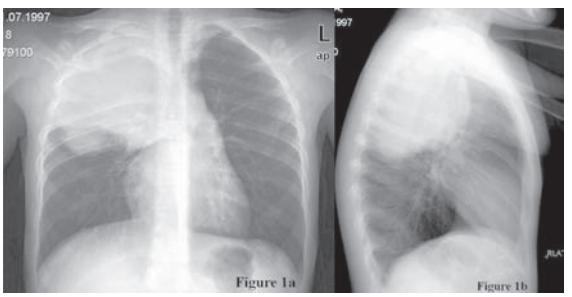


**Fig. 1.** A large mass lesion within the upper right thoracic cavity and lytic bone lesion at the 4th rib were seen by the posteroanterior (a) and lateral (b) chest radiographs.

The mass was completely removed and the 4th rib was partially resected without any need for reconstruction of the chest wall. Histopathologic examination was consistent with ABC. She has been followed up for 56 months without any recurrence.

## Discussion

An ABC is an expansile osteolytic lesion with a thin wall, containing blood-filled cystic cavities[3]. The term aneurysmal is derived from its radiographic appearance. ABCs are difficult to distinguish from malignant lesions in some locations. It may mimic a sarcoma in the ribs, scapula, or sternum, especially when associated with a large soft-tissue component. A CT is usually necessary for differential diagnosis, which also provides information on the extent of involvement before surgery or other treatment. An expansive, multilobulated, cystic chest wall mass presenting with multiple fluid – fluid levels arising from the right 4th rib was detected in our case. Fluid-fluid levels also are seen in many other bone lesions, and this finding is not specific to ABCs. These levels may be seen in malignant and benign lesions, such as giant cell tumors, and in telangiectatic osteosarcomas. In this case, clinical, laboratory and radiological findings were suggestive of benign primary rib tumors, which are rare and account for 5-7% of all primary bone neoplasms[6]. These tumors include fibrous dysplasia, enchondroma, osteochondroma, ABC, osteoid osteoma, osteoblastoma, giant cell tumor and cystic angiomatosis[6]. Besides the benign primary tumors of the ribs, malignant

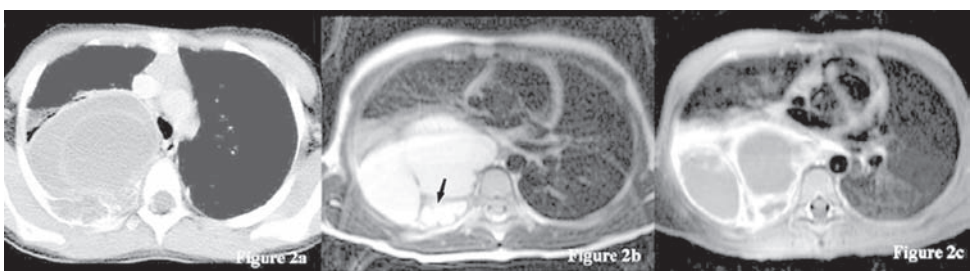


**Fig. 2.** (a) Chest CT scan showed an expansive, multicystic, chest wall mass lesion, compressive atelectatic changes at the adjacent lung parenchyma, cortical destruction and lytic changes at the 4th right rib. (b) T2-weighted MR images revealed fluid-fluid levels in the multilobulated cysts. (c) The internal content of the mass was seen hyperintense on fast spin echo T1-weighted MR images, which refers to proteinous and hemorrhagic fluid.

tumors such as soft tissue sarcomas should also be considered in the presumptive diagnosis, but absence of a solid component and lack of radiological evidence for malignant features were unlikely for a malignant tumor. However, resection of the mass was required both for diagnosis and treatment of this huge cystic tumor. Definitive diagnosis could only be made after histopathological examination.

Aneurysmal bone cyst usually occurs in the first two decades of life, and the median age at diagnosis is 10.2 years[4]. Both sexes are equally affected. The most common sites are long bones, spine and pelvis[1-5]. It rarely originates from the ribs, but when it does, posterior or lateral aspects are primarily involved, as observed in our case[6]. The review of some selected large ABC series showed rib primaries in 1.3-2.7% of the patients[3-5].

Chest wall tumors usually grow into the thoracic cavity, and they can reach huge sizes when they are clinically diagnosed, as in the reported case. ABC is a rapidly growing, destructive benign bone tumor that can show extension into adjacent tissues. Our case had a Stage 3 ABC according to the staging system described by Enneking et al.[1], due to the presence of progressive growth with cortical bone destruction. These tumors may cause pain, palpable mass, dyspnea or pathological fracture[3,6]. Despite the large size of the tumor, the only symptom in our patient was persistent cough. She also had anemia, which could be related to the hemorrhage of this multilobulated lesion into the cystic spaces.

Histopathological examination is mandatory for the definitive diagnosis. A needle biopsy is sometimes not sufficient, so an open incisional biopsy may be required for differential diagnosis, particularly of telangiectatic osteosarcoma, which is a highly neoplastic disease[2,5].

The main treatment strategy is primary surgery, although spontaneous healing has been reported in a few cases[2,7]. Follow–up without any treatment could be offered for inactive and asymptomatic lesions, until they increase in size and become painful[2,7]. A complete local excision should be performed if possible, especially in ABC arising in nonessential bones such as proximal fibula, clavicle, rib, or pubic ramus[1,2,5,7]. Selection of the treatment method depends on the age of the patient and location of the mass, and also its nature as active or inactive or clinically aggressive[2,7]. The treatment of pelvic or spinal ABCs, or those in close contact with an epiphyseal plate, is especially problematic. Aggressive surgery should be avoided. Our patient underwent primary surgery electively. The mass lesion was completely resected with partial resection of the 4$^{th}$ right rib without any need for reconstruction. Successful treatment of ABC by curettage of lesions with or without bone grafts has also been reported[2,5,7]. The intralesional procedures such as percutaneous sclerotherapy, embolization of the lesion or intralesional implantation of bone particles may be suggested not as an initial treatment but an alternative to surgery, especially in patients with unresectable ABC[2,7]. Local recurrence is the main problem, and the recurrence rate of ABC ranges from 5% to 40% depending on the type of treatment[4]. The local recurrence risk may be reduced by adjuvant therapies including phenol instillation, cryosurgery, or laser coagulation in patients with aggressive and/or recurrent ABC[1]. Radiotherapy may also be considered for some selected inoperable patients with incompletely resected, aggressive, and/or recurrent ABC[1], but should be avoided whenever possible because of significant late effects[2,7]. Two or more years of follow–up has been recommended to evaluate the efficacy of treatment, because recurrences are more commonly seen within the first two years of diagnosis[1]. Our patient has been followed for 56 months without any recurrence.

Aneurysmal bone cyst can occur either as a primary or secondary lesion. The etiology and pathogenesis are unclear. A secondary type of ABC constitutes 20-30% of cases and develops in an underlying bone lesion such as giant cell tumor, unicameral bone cyst, hemangioma, hemangioendothelioma, chondroblastoma, osteoblastoma, telangiectatic osteosarcoma, chondrosarcoma, nonossifying fibroma, fibrous dysplasia, chondromyxoid fibroma, or eosinophilic granuloma, where it may occur as a consequence of alterations in hemodynamics and vascular hemorrhagic degenerative processes[1-5,8]. Many different theories have been suggested for the pathogenesis of the primary ABC. The previously proposed theories about the reactive nature of the primary ABC are being challenged today[8]. The demonstration of recurrent chromosomal abnormalities, t(16;17)(q22;p13), discovery of the role of the ubiquitin protease

USP6 oncogene, characterization of the clonality, and demonstration of insulin-like growth factor-I overexpression in primary ABC cells have led investigators to suggest that primary ABC is a neoplastic disorder[9,10]. Since this is an uncommon tumor, collaborative studies including genetic analysis should be performed to further identify the pathogenetic mechanisms.

In conclusion, ABC is a rare entity, but should be considered among the presumptive diagnoses of chest wall masses during childhood. Clinical and radiological investigations are helpful for the diagnosis; however, definitive diagnosis depends on histopathologic examination. A complete local excision should be attempted whenever possible, avoiding any permanent morbidity.

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