Solid variant of aneurysmal bone cyst of the rib presenting as a left intrathoracic mass without radiological bone destruction

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An aneurysmal bone cyst (ABC) is a benign but often rapidly expanding osteolytic multi-cystic osseous lesion that occurs as a primary, secondary, intra-osseous, extra-osseous, solid, or conventional lesion. A 15-year-old boy presented with a left-sided intrathoracic mass displacing the lung without bone destruction. The mass was totally resected without rib resection, and the pathological diagnosis was ABC. The clinical manifestations, etiology, management, and pathology are discussed, with a brief discussion regarding the difficulty in the preoperative differential diagnosis.

Key words: aneurysmal bone cyst, rib, child.

An aneurysmal bone cyst (ABC) is a benign, non-neoplastic proliferative tumefaction of bone characterized by the presence of channels and spaces of varying size surrounded by delicate walls. It often expands rapidly and causes a locally destructive cystic bone lesion. Although ABC affects all age groups, the peak incidence is in the first and second decades of life. It accounts for about 1-2% of biopsied primary bone tumors. It may be of conventional or solid type[1,2]. The solid variant is a rare sub-type and more difficult to recognize. The metaphyseal region of long bones, including the distal femur and proximal tibia, are the most common sites; rib involvement is very rare[2].

Herein, we present a 15-year-old boy who underwent surgery for a left thoracic mass, which was proven to be an ABC by pathologic examination.

Case Report

A 15-year-old boy was admitted to our hospital with the complaint of left-sided chest pain of one-month duration. The chest pain was vague at first, but grew more intense in time. The medical history of the patient did not reveal any trauma or remarkable disease. He did not experience any other pain or fatigue, weight loss or fever before the present complaint. On the physical examination, he had regular respiration with a slight decrease in the respiratory sounds on the left side, and he had no palpable mass on the thoracic wall. The laboratory investigations including blood biochemistry tests and lactate dehydrogenase value were within normal limits.

The patient admitted to our clinic with a nonopaque computerized tomography (CT), which was taken at the referring hospital. Plain and lateral chest radiographs showed a well-circumscribed mass with irregular borders on the left side (Figs. 1a, b). CT showed a round mass, measuring 77x68x70 mm, with cystic, solid, and calcified components within the left thorax, compressing the left upper lobe. No significant destruction of adjacent ribs was detected (Fig. 2); however, it was considered inadequate for detailing the mass due to unenhanced contrast. Magnetic resonance imaging (MRI) of the chest revealed a round, multiseptated mass in the left hemithorax compressing the upper lobe of the left lung. Axial T2A MRI showed the lobulated and multiseptated mass with fluid-fluid levels representing blood degradation products within cystic spaces. Contrast enhancement at the periphery and septae were seen (Figs. 3a, b). Atelectasis of the adjacent lung parenchyma was noted. The bones and soft tissues composing the chest wall were normal.

A left posterolateral thoracotomy was performed.

An 8x7 cm, hard, lobulated, well- circumscribed mass was seen adhered to the inner surface of the fourth and fifth ribs, intercostal muscles, and parietal pleura. The mass was loosely adhered to the visceral pleura but did not invade the lung parenchyma. Atelectasis of the adjacent lung parenchyma was noted. The mass was excised en bloc without bone resection, but the periosteum was injured during the excision of the mass.

Macroscopically, an irregular soft tissue mass was seen. A thin fibrous capsule was seen on the outer surface of the nodular mass. On cross-section of the mass, blood- and mucin-filled cystic areas with peripheral calcifications were seen. The microscopic examination demonstrated multiple blood-filled cavernous spaces separated and surrounded by fibrous connective tissue, multinucleated osteoclastic giant cells, and well-developed vascular spaces with hemorrhages in the septae lining the cystic spaces (Fig. 4). At the periphery of the mass, inflammatory and osteoclastic cells were seen. There were focal areas of calcification and osteoid. No malignant component was observed and the surgical margins were free. These pathological findings were consistent with solid ABC. The patient was discharged after an uneventful one-week postoperative course. During the two-year follow-up, the patient did not experience any complaints and the chest radiographs did not show any lytic rib lesions.

Discussion

Rib tumors are rare entities in the pediatric population; however, a significant number of rib lesions are malignant[3]. Benign primary tumors of the ribs, such as fibrous dysplasia (30%), enchondroma (2.8-12.2%), osteochondroma (2.7-8.5%), Langerhans cell histiocytosis (eosinophilic granuloma), osteoid osteoma (1-1.4%), ABC (1-2%), osteoblastoma (0.9%), giant cell tumor (0.5-1.4%), and cystic angiomatosis[4] are rare, comprising 5-7% of all primary bone neoplasms. It is also difficult to distinguish these diseases radiologically. Although mesenchymal hamartoma typically presents prenatally or within six months of age as an extrapleural mass arising from the ribs, it should be considered in the differential diagnosis of ABC with consideration of the age of the patient[5]. Precise diagnosis is made only by pathological examination.

The most perplexing subject in the presented patient is the absence of a lytic rib lesion accompanying the intrathoracic mass in the radiological images; therefore, ABC of the rib was not considered initially in the differential diagnosis.

Aneurysmal bone cyst (ABC) was first described by Jaffe and Liechtenstein in 1942 and accounts for only 1.3% of all bone tumors[6]. It is an expanding osteolytic bone lesion consisting of blood-filled spaces that are separated by connective tissue septae containing trabeculae of bone or osteoid tissue and osteoclast type giant cells[7]. In 1983, Sanerkin et al.[8] described a variant of ABC in which the predominant histological features were solid, and they used the term "solid variant of aneurysmal bone cyst"

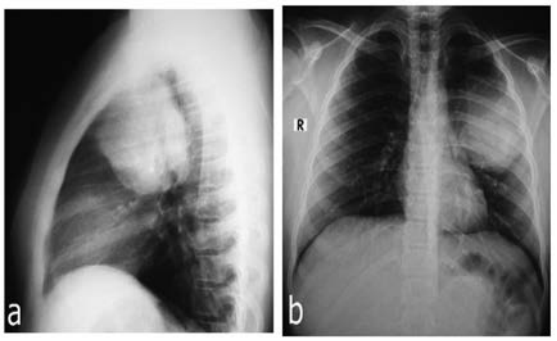

Fig. 1a, b. Plain and lateral chest radiographs showed a well-circumscribed mass with irregular borders on the left.

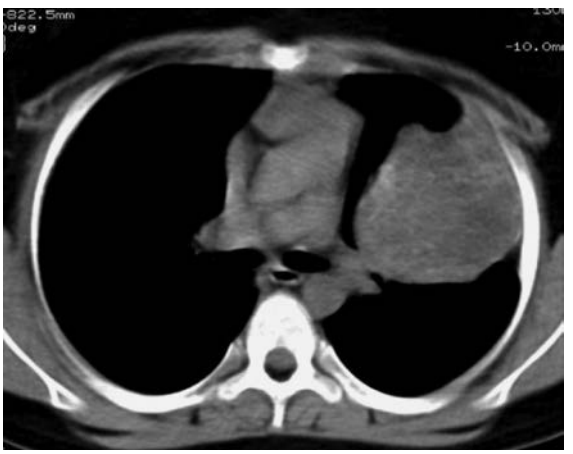

Fig. 2. Computed tomography (CT) showed a round mass, measuring 77x68x70mm, with cystic, solid, and calcified components within the left thorax compressing the left upper lobe; no significant destruction of adjacent ribs was detected.

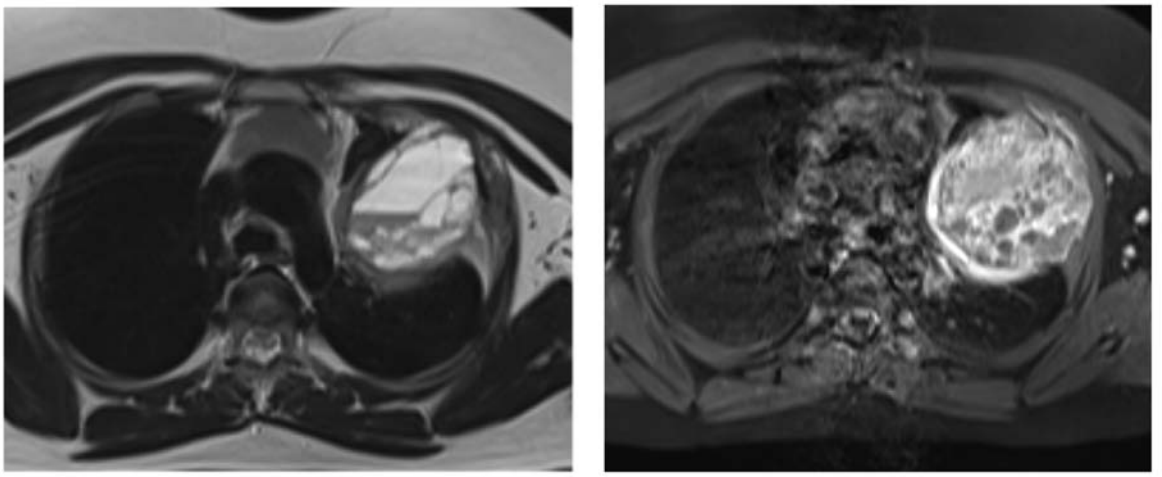

Fig. 3 a, b. Magnetic resonance imaging (MRI) of the chest revealed a round, multiseptated mass in the left hemithorax compressing the upper lobe of the left lung. Axial T2A MRI showed the lobulated and multiseptated mass with fluid-fluid levels representing blood degradation products within cystic spaces (Fig. 3a); contrast enhancement at the periphery and septae were seen after intravenous contrast administration (Fig. 3b). Atelectasis of the adjacent lung parenchyma was noted. The bones and soft tissues of the chest wall were normal.

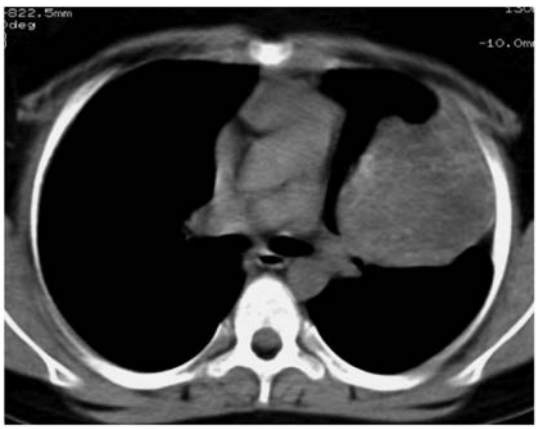

Fig. 4. Cystic spaces containing red blood cells separated by septae and composed of spindle-shaped fibroblastic cells and multinucleated giant cells are shown.

(ABCs) to describe it. Histopathologically, it is a non-neoplastic bone lesion[6]. It may involve any bone, mostly the spine and long bones, but localization in the ribs is unusual, involving 2.7% of all cases[9]. ABC is predominantly a disease of the first three decades of life, and occurs equally in both sexes[10].

The etiology of ABC is unknown, but circulatory disturbances and trauma have been proposed to be factors in the pathogenesis[10]. The medical history of our patient did not reveal any trauma or remarkable disease. They may originate as a localized arteriovenous malformation, and may have a pre-existing lesion such as fibrous dysplasia, nonossifying fibroma, giant cell tumor, osteoblastoma, angioma and chondroblastoma, or chondromyxoid fibroma[4], and in the presence of such a lesion, it is designated as 'secondary ABC'. If a pre-existing lesion is not identified, then it is classified as 'primary ABC', as defined in our case.

The most common symptoms in a patient with ABC of the rib are chest pain (46%), swelling of the chest (21%), dyspnea (7%), paraplegia (7%), and pathologic fractures (2%), similar to all chest wall tumors[10]. Our patient had only chest pain, which was intermittent and vague at first, but had gradually increased and became more intense over a period of one month. There was no respiratory distress.

Although the radiological appearance of ABC of the rib is not specific enough to establish a definite diagnosis, the typical radiological features are a purely lytic, eccentric, expansile, and sharply circumcised lesion, which usually arises in the metaphysis[11]. CT and MRI are useful in the diagnosis. MRI typically demonstrates a lobulated, multiseptated lesion with fluid-fluid levels and blood degradation products surrounded by a thin, well-defined low signal intensity rim on both T1- and T2-weighted images. Fluid-fluid levels may also

be a feature of other lesions, including giant cell tumor and chondroblastoma[6]. The value of scintigraphic characteristics of such cysts is realized when complete radiographic imaging studies of solitary bone lesions are obtained preoperatively. The diagnosis is confirmed if the scintigram features are typical[12].

Although the patient's MRI showed a multiseptated mass with fluid-fluid levels representing blood degradation products within cystic spaces, we considered it to be an intrathoracic extrapulmonary lesion, particularly an atypical presentation of hydatid cyst, instead of ABC due to the absence of any bone destruction[13]. Hydatid cyst is still a substantial public health problem worldwide, including in Turkey. In this setting, a biopsy was not considered before surgery. However, perioperatively, we found a well-defined capsulated hard mass adhered to the fourth and fifth ribs that was totally separated from the inner margin of the ribs with sharp dissection.

The current popular methods of treatment of ABC include curettage and bone grafting, radiation, curettage and cryosurgery, or an en bloc resection[11]. We performed an en bloc resection of the mass without rib resection. The histopathological diagnosis was solid variant of ABC. Therefore, we reevaluated the presurgical chest radiograph and CT for an unrecognized lytic rib lesion, and followed the patient with periodic chest radiographs for two years. The patient did not experience a recurrent mass or a lytic rib lesion during the follow-up.

Aneurysmal bone cyst (ABC) has been of great interest to pathologists because they may be mistaken for malignant tumors, mainly in cases of giant cell tumors or osteosarcomas, on the basis of cellularity and variable mitotic activity[1].

In conclusion, we report this case because of its unique features. This case was extraordinary due to the absence of any bone destruction, and was thus considered an intrathoracic extrapulmonary mass until the operation, when we determined that it was adhered to the ribs without destruction. A search of the literature revealed no prior publication regarding ABC without bone destruction. The pathological diagnosis was solid variant of ABC, which is a very rare form.

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