

## Case Report

# Standing in the shadows - rare case of aneurysmal bone cyst of chest wall presented with massive hemothorax

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## ABSTRACT

Chest wall tumours of bones in childhood are rare in general. Chest wall aneurysmal bone cysts (ABC) are even rarer. If happened, they affect spine and long bone, those affect chest wall are extremely sparse. We are presenting a case of 8-year-old girl who presented with spontaneous hemothorax. We share our experience and diagnostic dilemma we encounter in emergency situation as well as challenging in the management. Aneurysmal bone cysts, despite being benign, but, rarity and unusual presentation added to atypical radiological features may lead to catastrophic consequences unless diagnosed on time.

**Keywords:** Aneurysmal bone cyst, Hemothorax, Thoracotomy, Bone tumours, Chest wall

## INTRODUCTION

Chest wall tumours are rare in children. They are usually highly malignant and mostly consist of malignant small round cell tumors e.g. Ewing's sarcoma, primitive neuroectodermal tumor (PNET), rhabdomyosarcoma, osteosarcoma, chondrosarcoma, and other sarcomas.<sup>1</sup>

Aneurysmal bone cysts (ABC) are primarily seen in adolescents, with 80% occurring in the patients less than 20 years of age.<sup>2</sup> They are usually secondary to an underlying lesion such as fibrous dysplasia, giant cell tumour, chondroblastoma and osteosarcoma.<sup>3</sup> An ABCs without a primary lesion is a rare entity.<sup>4</sup> Primary ABCs occurs most commonly in teenagers.<sup>5</sup> ABCs are mostly benign however, it can be locally aggressive leading to erosion and weakening of the bone structure.<sup>6</sup> They are cystic expansile bone lesion that can occur in any bone in the body, rarely involves the ribs.<sup>7</sup> Presentation is usually due to mass effect on the adjacent soft tissue, causing pain, swelling, deformity, disruption of growth plates, neurologic symptoms depending on the location of the

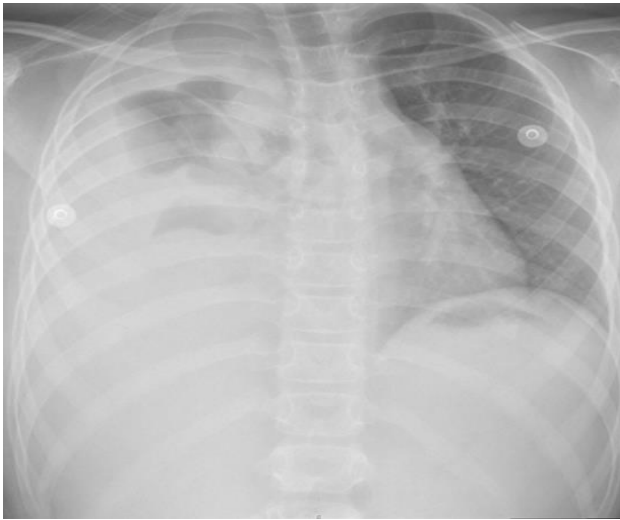
lesion, and pathologic fractures.<sup>8</sup> We are presenting a rare case of ABC of the chest wall in a child, highlighting its atypical presentation and diagnostic challenges we encountered while managing this patient.

## CASE REPORT

We are presenting a case of 8-year-old girl who came with worsening chest pain that associated with shortness of breath for few days. No constitutional symptoms. Clinically, she was pale and baseline investigations shows microcytic hypochromic anemia. Chest radiograph (Figure 1) showed massive right pleural effusion up to middle zone. Pleurocentesis revealed hemorrhagic fluid. Based on this findings, a computed tomography scan was performed (Figure 2), revealed extensive hemothorax with extravasation of contrast which was attributed to bleeding from posterior intercostal vessels, resulting in mediastinal shift to the left. While closely examining images of bone setting (Figure 3), we noticed a faint opacity adjacent to the inner surface of the posterior eighth rib with minimal cortical erosion suspicious of

arterial extravasation. However, until that moment, intercostal vessels were incriminated as bleeding source. The minimal cortical erosion was not linked to the clinical situation.

Based on the provisional diagnosis of vascular bleeding, selective angiogram was performed (figure 4), surprisingly, a tumour blush rather than vascular malformation or pseudo aneurysm clearly manifested. This indicates that the hemothorax is due to bleeding from a chest wall tumour-like lesion rather than vascular lesion. Embolization was not performed as decision to surgically resect the tumour was made.



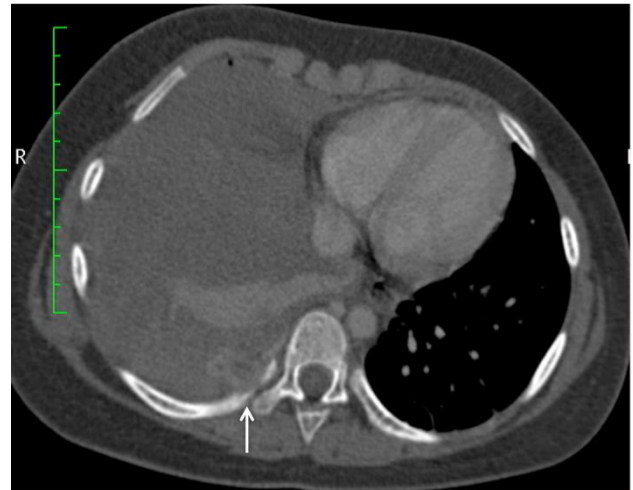
**Figure 1: Postero-anterior plain chest radiograph showing massive right pleural effusion up to middle zone.**



**Figure 2: Axial view of CT scan of the thorax showing massive right hemothorax with shifting of mediastinum to the left (white arrows).**

Emergent thoracotomy was performed, where a small tumour, around 2x2 cm in size was excised en-bloc and homeostasis was secured. Chest drain was inserted. Histopathology of the resected tumour came as ABC evident by the presence of circumscribed mass composed of numerous blood-filled cystic spaces separated by

fibrous septa containing fibroblast, capillaries, inflammatory cells, benign osteoid and scattered multinucleated giant cells. Post-operative period was uneventful, patient discharged home after 7 days.



**Figure 3: Axial cut of bone window of CT of the chest showing faint opacity (white arrow) adjacent to the inner surface of the eighth right rib.**



**Figure 4: Selective cannulation of posterior right eighth rib where artery angiogram showing tumour blushes.**

## DISCUSSION

ABC is not a true cyst or aneurysm. The etiology is unknown but the theory of arteriovenous malformation is widely accepted.<sup>10</sup> There is no race or sex predominance. Approximately 80% of ABC occurs in teens and below age of 20, rarely below 4 years.<sup>11</sup> ABCs may involve any bone, mostly spine and long bone.<sup>3, 9</sup> Grossly, ABCs consists of a paper-thin cortex and multiple blood filled cavities. The lesion should be designated as a primary ABC if it has a uniform histological pattern or as a secondary ABC if the lesion contains another bone tumor

such as fibrous dysplasia, non-ossifying fibroma, osteblastoma or chondromyxoid fibroma.<sup>6,10</sup>

Patients may present with pain, which may be of insidious onset or abrupt due to pathological fracture, with a palpable lump or with restricted movement.<sup>3,9</sup> Hemothorax is an unusual presentation for aneurysmal bone cysts. Rangachari et al proposed that aneurysmal bone cysts have raised intra-cystic pressures which are dynamic and diagnostic in nature.<sup>8</sup> Raised intra-cystic pressures in aneurysmal bone cysts are maintained as long as the periosteum over the cyst is intact even in those with pathological fractures.

The radiographic features of ABCs vary from a unilocular radiolucency to a “ballooned out” multilocular radiolucency with a honeycomb or soap-bubble appearance, rarely, radiopaque.<sup>12</sup>

ABCs are divided into five morphologic types, as described by Capanna et al<sup>7</sup>: Type 1 is the central metaphyseal manifestation, the lesion is well contained within the bone, and the profile of the bone is either intact or expanded slightly. Type 2, the cyst involves the entire segment of bone, with cortical expansion and thinning. Type 3, the cyst has an eccentric metaphyseal location, showing no or minimal expansion of the cortex. Type 4, the cyst has a subperiosteal expansion with no or minimal cortical erosion. Lastly, type 5, the lesion displaces the periosteum toward the soft tissues and at the same time penetrates the cortex, extending into the cancellous bone.

ABC can be further classified based on their behaviour in to: 1) Aggressive type, with no evidence of reparative osteogenesis, no periosteal shell, and an ill-defined endosteal margin. 2) Active type, with an incomplete periosteal shell and a sharply defined intraosseous border. 3) Inactive, with a complete periosteal shell and the intraosseous margin defined by a sclerotic rim of reactive bone.<sup>6</sup>

Traditionally ABCs have been treated by curettage and bone grafting with a recurrence rate of between 11 and 31%. En-bloc resection showed to reduce the recurrence rate. Other options include selective embolization of feeding artery and intralesional injection of fibrosing agent such as phenol or liquid nitrogen either in isolation or as a precursor to surgical excision, this can further reduce the recurrence risk up to 20%.<sup>8</sup>

Back to the patient, the acute presentation with dyspnoea and hemothorax, together with unusual radiological features made the initial diagnosis difficult. Hemothorax can be explained either due to spontaneous rupture of the ABC following perforation of its outer shell, most likely, or due to possible penetration of an intercostal artery by the expanding tumour. Interestingly, despite CT scan

showed bleeding from the chest wall, however, suspicion of tumour-like lesion was raised only by angiography, which actually intended for therapeutic embolization of the bleeding. Surgically obtained tissue histology was the definitive diagnostic tool for this challenging lesion.

ABC is a rare, benign pathology, which should be considered in the differential diagnosis of bleeding chest wall tumors. Complete surgical excision can be the best treatment for cure.

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