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Mesenchymal Hamartoma of the Chest Wall Presenting as Aneurysmal Bone Cyst in Infancy: Two Case Reports

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Abstract **Mesenchymal hamartoma of the chest wall in infancy is a rare condition. The tumor usually presents as a large thoracic mass with surrounding destruction, which can confuse as a malignant bone tumor. Herein, we report two cases that had been treated by surgical resection to demonstrate the clinical, radiological and pathological descriptions.**

Although chest wall tumor is not an uncommon problem in pediatric patients, the majority of such masses are metastatic lesions.[1] Among the primary rib tumors, two-thirds are malignant lesions. Mesenchymal hamartoma of the rib is a rare benign condition, occurring in infants with an incidence of 0.03 per cent.[1,2] To distinguish this surgically curable condition from other malignant neoplasm of the chest wall is of importance because its clinical and radiological presentations are often confused with aggressive bone tumors. Herein, we report the condition in two infants, who presented with large aneurysmal chest wall tumors, to demonstrate their clinical manifestation, radiological feature, histopathology and surgical management.

Case Reports

Case 1

A six-month-old male infant presented with failure-to-thrive and swelling of the left chest wall of three months duration. The pregnancy and birth history were normal. On palpation, a bony-hard mass was found over the antero-lateral aspect of the left thorax. The infant expressed neither signs of respiratory distress nor abnormality of any other systems. The chest x-rays revealed a large extrapulmonary mass, involving the left 3rd to 6th ribs. The adjoining ribs were spread apart and deformed. Thick irregular calcification and compression of the mediastinum to the right side were also visible (Figure 1).

Computed tomography (CT) of the chest showed a multilocular chest wall mass with fluid level, measuring 6×8×10 cm in greatest dimensions. The mass was surrounded by cortical bony rim. Adjacent ribs destruction and mediastinal shifting were also confirmed (Figure 2). An incisional biopsy was initially performed and "benign neoplastic process" was reported. A left thoracotomy with an attempt for en bloc resection was then scheduled. Gross findings were mixed solid-cystic lesions, originating from the expansion of the

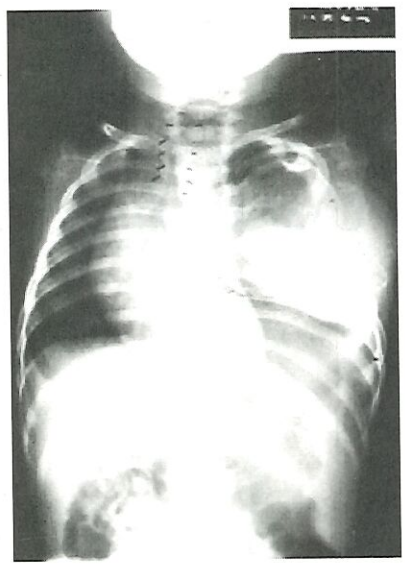

Fig. 1 Chest x-rays revealed a large extrapulmonary mass, spreading the adjoining ribs apart. Thick irregular calcification and compression of the mediastinum to the right were also evident.

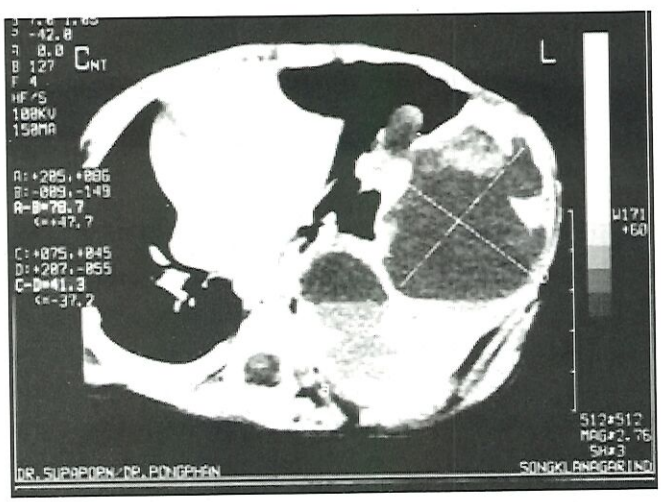

Fig. 2 Chest CT scan showed a multi-locular chest wall mass with fluid level, surrounded by cortical bony rim. Adjacent rib destruction and mediastinal shifting are noted.

right fifth costal bone and cartilage (Figure 3). The adjacent ribs were destroyed by tumor compression, without cortical invasion. Old blood filled the cystic component. Resection involved removal of the antero-lateral portion of the third to sixth ribs. The chest wall defect was then replaced with a four-layer piece of Malex mesh. The histopathology disclosed chondroblast-like mesenchymal stroma with hyaline cartilaginous metaplasia. The large cavernous spaces filled

Fig. 3 Gross finding of the chest wall mass, view from inner aspect.

with blood were similar to that found in an aneurysmal bone cyst. Foci of calcification, enchondral ossification, bone formation and osteoclast like giant cells were also demonstrated (Figure 4). The tissue characteristics were consistent with chondrogenic mesenchymal hamartoma of the ribs. The post-operative course was uneventful and the patient did well up to 12 months of follow-up visit when he began to have mild scoliosis.

Case 2

A two-month old male infant presented with progressive bulging of the left chest since birth. Despite normal growth and development, several episodes of tachypnea were noted by his mother. The physical findings and radiological studies were not different from those of Case 1. A core needle biopsy revealed mesenchymal hamartoma. A left thoracotomy and en-bloc resection of the mass was done. Chest wall reconstruction was made with synthetic mesh. The mass in this case measured approximately 6 cm in greatest diameter, expanding from the anterior portion of the left sixth rib. The cut surface showed a mixed cystic-solid lesion, filled with dark blood. The histopathology confirmed the cartilaginous mesenchymoma. The patient made a good recovery after six days of post-operative ventilatory support. Clinical follow-up detected no recurrence up to nine months.

Discussion

Mesenchymal hamartoma was first reported by Le Ber and Stout in 1962, when they coined the term "Benign mesodermal tumor".[3] The condition was

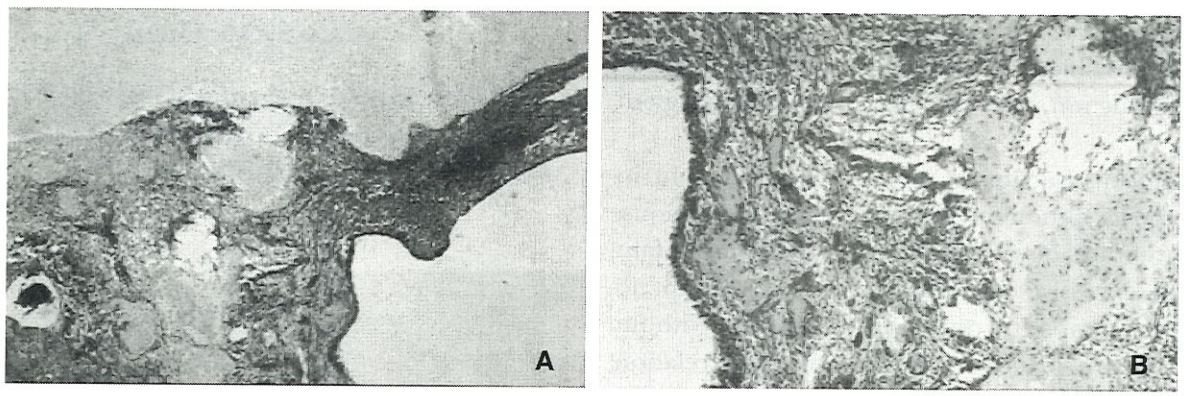

Fig. 4 The histopathology at 10X (A), and 40x (B). Note a large cavernous space filled with blood, similar to that is found in an aneurysmal bone cyst.

then occasionally reported under different terminology such as infantile cartilaginous hamartoma,[2,4] mesenchymal hamartoma,[5-8] thoracic mesenchymoma of infancy[9-11] and chest wall hamartoma.[12,13] To the best of our knowledge, approximately 60 cases have been reported in English literature.

The pathology is thought to develop antenatally but the age of presentation depends on the size and rate of growth. Most of the cases were diagnosed within the first year of life,[13] however some can be detected in utero by ultrasound and there was a report of teenage presentation.[7] The clinical manifestation varies from an asymptomatic chest wall mass disclosed on the chest x-rays, a visible chest wall deformity, to respiratory distress symptoms.[5,7,12] The respiratory symptoms may be caused by a large mass compressing the airway or thoracic contents. Fatal respiratory failure, shortly after birth has also been described.[5,13,14]

Mesenchymal hamartoma of the chest wall has been reported in males more frequently than females in the ratio of 2:1, and may occur to either left or right side.[13] The majority of tumors are a solitary lesion, ranging in size from 2-8 centimeters.[1,2,4-15] Satellite lesions and bilateral disease exist, although only rarely encountered.[5,6] The radiological features of the lesion are distinctive and was described by Oakley and his colleagues as a large expansile, extrapleural mass involving one or more ribs, with irregular areas of mineralization and ossification, forming an incomplete trabecular network centrally and expanding the involved ribs, with complete or partial destruction. The adjoining ribs appear to be distorted and partially eroded secondary to the mass effect.[16] These characteristic features often cause the lesion to be confused with an aggressive malignant tumor. Complete opacification of a hemithorax with mediastinal displacement to the contralateral side is not uncommon.[11] The chest CT scan provides more precise information regarding intrathoracic extension, compression or destruction of the adjacent ribs and displacement of the mediastinal structures, which is valuable in surgical planning.

Diagnosis by fine needle aspiration cytology may be possible with sufficient supplementary clinical history and radiological findings.[2,7] However, an incisional biopsy or core needle biopsy, as we performed here is safe and provides adequate tissue for a definitive histological diagnosis. The differential diagnosis includes aneurysmal bone cyst, chondroma and other mesenchymal neoplasms such as neuroblastoma and ganglioneuroma.[1,12]

Although malignant transformation occurred to one patient,[13] whether the mass was a true neoplastic process or a developmental mishap is controversial. The latter theory is more popular because of the lack of nuclear enlargement, irregularity and hyperchromatism.[11,13] McLeod and Dahlin suggested the term "hamartoma" be more suitable than the previously used "mesenchymoma". Currently accepted theory explains that the tumor is the result of focal overgrowth of cartilaginous tissue during the ossification process of the ribs, which normally takes place from the posterior to anterior portion.[15] The histological findings consist of three main components which are 1) focal sheets of primitive mesenchymal stroma composing of prechondrocytes without cellular atypia, 2) cartilaginous metaplasia from stoma, and 3) prominent vascularity to aneurysmal bone cyst. The

variable additional findings are foci of calcification, enchondral ossification, fibroblastic hyperplasia, woven bone production, and reactive osteoclast-like giant cells.[1]

Surgical resection is the curative treatment in most reports although there are some exceptions. Blumenthal and his group observed one female infant who had excessive bleeding during an attempt to remove the tumor and only biopsy was done. Without any treatment, the tumor got smaller in size in relation to the thorax with little protrusion beyond the intercostal space.[9] Because the en bloc resection results in a large chest wall defects that may lead to respiratory paradox in the short term and scoliosis in the long term, excision in an asymptomatic patient is debated.[12,13] Growth potential and reported malignant transformation justify the surgical intervention. However, the tumors in some cases stop growing within the first year of life and surgery may be substituted by watchful follow-up. The choices of chest wall reconstruction are personal. Both natural myocutaneous flaps and synthetic materials were selected according to the availability and experience of the surgeons. Although natural flaps are superior in terms of freedom of foreign body, the procedure requires more operating time and may cause more difficulty in cases of recurrence.

One case of malignant transformation was reported by Dounies et al.[13] Primary resection in this case was incomplete due to a technical difficulty and the residual tumor grew rapidly during the sixteen months following the initial resection. Histopathology of the metachronous lesion was poorly differentiated sarcoma. The patient had a seven-year disease-free period after the last operation, which was followed by adjuvant radiation and chemotherapy. Approximately 10 per cent of the cases exhibited local recurrence.[5,10,11,13-15] The longest period from primary resection to the diagnosis of recurrence was 2 years.[5] Scoliosis was reported in 24 per cent and the risk is greater with the larger lesions or when the resection was delayed.[13]

In summary, mesenchymal hamartoma of the chest wall in infancy is a benign tumor that has distinctive radiological features and can be diagnosed by fine needle or core needle biopsy. En bloc resection is curative but has to be followed by careful follow-up to watch for recurrence or scoliosis.

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