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Case Report

A rare case of mesenchymal hamartoma of chest wall in a neonate

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ABSTRACT

Mesenchymal hamartoma of the chest wall (MHCW) is an extremely rare, benign lesion arising from one or more ribs. Typically, MHCW manifests as a unilateral lesion, yet there have been instances of multiple or bilateral occurrences documented. It is reported to have an increased male predisposition and the common presentation includes a visible chest wall mass. In asymptomatic neonates, conservative management is the preferred approach since there have been no reports of malignant transformation. Surgical intervention becomes necessary in cases where there is an active growth of the lesion or respiratory compromise. Here, we present a case of unilateral MHCW in a newborn being managed conservatively, followed by surgical resection at 2 months of age.

Keywords: Mesenchymal hamartoma, Benign chest wall mass, Neonate

INTRODUCTION

Mesenchymal hamartoma of the chest wall (MHCW) is a rare, benign lesion that arises from the mesenchymal chondro-osseous tissue of the ribs.[1] It is seen in infancy and early childhood with a few cases detected antenatally.[1,2] Prenatal diagnosis is possible but it can only be confirmed by radiological and histological features at birth.[3]

The lesion has a rapid tumor growth period in infancy followed by a self-limited growth period. With the large size of the tumor and aggressive radiological appearance mimicking malignancy, the diagnosis can be challenging.[1] The initial rapid growth prompts clinicians to opt for surgical resection of the lesion irrespective of clinical presentation but MHCW can be managed non-operatively. When MHCW is in the self-limited growth phase, slow regression of the lesion is possible, especially in asymptomatic cases. Regular follow-up is mainstay in such cases to detect increasing size of the tumor or any new symptoms.

To our knowledge, very few cases of MHCW are reported in the neonatal period. Surgical resection of the lesion has been the traditional approach regardless of the presentation. However, resection of a large lesion such as MHCW in neonates increases the risk of post-operative complications. Conservative approach has been increasingly recognized considering the benign and self-resolving nature of the lesion. But there are reported cases of rapidly expanding MHCW which can cause life-threatening respiratory complications. In such cases, surgical intervention should be considered without any delay.[3] We hereby report a case of asymptomatic MCHW being managed conservatively in neonatal period followed by surgical resection when respiratory compromise was noted later in life.

CASE REPORT

Single term baby boy was born to a 28-year-old primigravida mother by caesarean section at 38+6 weeks gestation with a birth weight of 3.225 kg. At 10 hours of life, the baby was referred to our hospital with a mass over the right postero-lateral chest which was noted soon after birth. Besides the obvious swelling, the baby was asymptomatic. The antenatal history including antenatal scans were normal. Birth history was uneventful. On physical examination, there was a hard swelling of 4×3×2

cm present over the right mid-axillary line which was non-tender and immobile. On auscultation, air entry was reduced over the right side. A chest radiograph was obtained, revealing a round opacity over the right lower anterolateral chest wall involving the 6th, 7th, and 8th ribs (Figure 1).

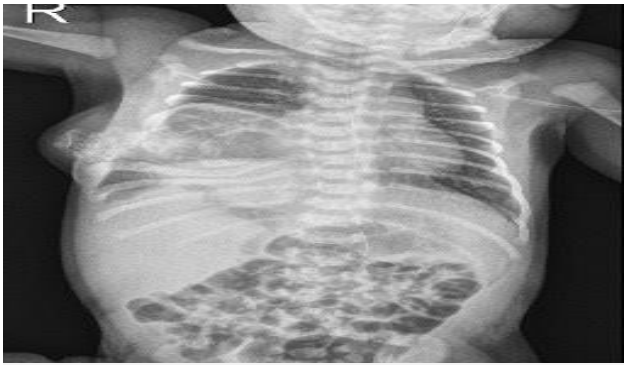

Figure 1: Chest X-ray at presentation showing a large mass arising from the right chest wall causing distortion of 6th, 7th and 8th ribs.

A non-contrast computed tomography (CT) scan of chest was done which confirmed a large soft tissue mass with ossification in the right posterior chest wall centred in the right 7th rib with destruction of the rib and splaying of the 6th and 8th ribs, extending both medially and laterally. The soft tissue component measured approximately 6.4×4.9×4.0 cm and occupied the middle and lower thoracic cavity, displacing mediastinal structures to the left, anteriorly displacing the right lower lobar bronchus, and compressing lower segmental branches causing underlying lung to collapse (Figure 2a and b). A biopsy of the lesion was performed, and the histopathological examination confirmed the diagnosis of a mesenchymal hamartoma of the chest wall. After a multidisciplinary team discussion, including paediatric oncologist, orthopedic oncologist, paediatric cardiothoracic surgeon, and interventional radiologist, conservative management was decided for the baby based on available literature. Screening echocardiography was also done as part of the workup which showed moderate-sized atrial septal defect with left to right shunt. Patient was regularly followed up and no increase in the size of lesion was noted. At 2-months, baby came for regular outpatient follow up when he was noted to have respiratory distress in the form of tachypnoea and chest retractions. A repeat non-contrast CT scan of the chest showed an increase in the size of the large soft tissue component 8.5×6.9×7.1 cm (previously 6.4×4.9×4 cm) with right lower lobar bronchus compression and right lower lobe collapse. The child underwent excision of the mediastinal tumour. The mass was dissected free of all the mediastinal structures along with a part of the right 6th and 7th rib. The procedure was uneventful, and baby was extubated to room air on the same day. The baby remained hemodynamically stable and was discharged on post-operative day 6. Presently, baby is 6 months old and is healthy and growing well.

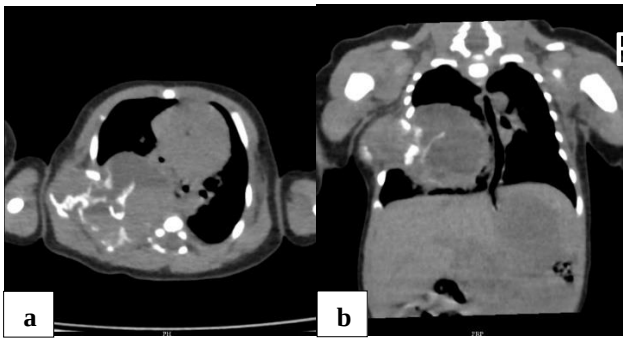

Figure 2: (a) Axial CT image demonstrating mass in the right posterior chest wall involving the ribs; and (b) coronal CT image demonstrating a large mass extending medially into thoracic cavity and laterally into the right chest wall. The intrathoracic component of the mass occupies the middle and lower thoracic cavity reaching till the mediastinum and displacing the mediastinal structures to the left side.

DISCUSSION

MHCW is a rare and benign lesion with an occurrence of <1 per million, mostly diagnosed in neonates or infants.[1,4] Till date, less than 115 cases of MHCW have been documented in medical literature. Typically, the lesion presents unilaterally, often originating on the right side.[4] There is a higher incidence in males, resulting in a male-to-female ratio of 2:1 to 4:1.[1] Majority of the reported cases suggest non-familial occurrence except for two cases that presented in siblings.[4] In majority of instances, the lesions manifested independently, without any accompanying congenital anomalies.

The clinical presentation of MHCW involves swelling in the chest wall accompanied by bone deformity evident on radiological examination. It is a well-circumscribed lesion which originates from the central portion of one or multiple ribs, distinct from the costochondral and costovertebral junctions, leading to erosion of the affected ribs.[2,5] The presentation varies depending on the tumor's size and its impact on the lungs and mediastinal structures, ranging from asymptomatic cases to severe respiratory distress.[3,4] Despite the benign character of MHCW, lesion extension into the thorax can cause respiratory compromise and pose a severe risk of mortality.[4]

Imaging is essential for identifying the condition but is not confirmatory. The lesion presents a range of differentials, including malignant primary bone tumors like Ewing sarcoma of the chest wall, primitive neuroendocrine tumors, and lymphoma, as well as benign conditions such as fibrous dysplasia, hemangioma, and Langerhans cell histiocytosis. MHCW is often mistaken for a malignant lesion, therefore biopsy along with radiographic evidence is necessary for a definitive diagnosis.[6] From a histological perspective, the tumor comprises cartilage, immature

mesenchymal elements, and areas resembling aneurysmal bone cysts, and newly formed bone structures.[5,6]

Two treatment approaches for chest wall mesenchymal hamartoma have been discussed in literature: surgical resection for patients experiencing respiratory compromise and conservative management for those who are asymptomatic. Complete surgical resection has traditionally been the primary treatment option for MHCW, particularly in symptomatic cases. This involves a wide en bloc excision of the affected portion of the chest wall, including the involved ribs, underlying pleura, intercostal muscles, and neurovascular bundles.[3,4] Evidence shows spontaneous resolution of the tumor in asymptomatic patients, and hence the need for regular follow-ups to monitor tumor size and detect any new symptoms. A total of 12 cases of non-operative management have been reported. Out of these, 3 patients underwent subsequent surgical management after an initial period of conservative management. Two of these had bilateral involvement and the lesion was resected only on one side. The first documented case of complete spontaneous resolution of MHCW was reported by Freburn in 2001, after an 11-year follow-up period.[2] Later in the same year, Cameron reported three cases of long-term nonsurgical management, becoming an early advocate for a non-operative approach given the condition's benign nature.[7]

In our case, the infant showed no signs of respiratory compromise in the neonatal period. With regular follow-up, increase in size of the tumor was noted with compressive symptoms and the baby underwent surgical resection of the tumor at 2 months of age due to respiratory compromise. The prognosis after surgical resection is excellent.

CONCLUSION

Mesenchymal hamartoma of the chest wall represents a benign lesion that may manifest asymptomatically. In cases where there is no evident respiratory compromise at birth, immediate surgical intervention may not be an absolute indication. Therefore, a conservative management approach can be pursued. However, surgical resection of the mass is the treatment of choice in life threatening symptomatic cases. Imaging modalities such as CT/MRI can provide useful diagnostic information.

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