

Mesenchymal Hamartoma of Chest Wall in an Infant: Mimicking Persistent Pneumonia

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ABSTRACT

Mesenchymal Hamartoma of the chest wall (MHCW) is a very rare benign tumour. They are usually discovered in infancy. Spontaneous regression is known to occur in this benign condition. Management is surgical removal of mass if respiratory compromise is present. Conservative management is preferred modality in asymptomatic children as malignant transformation is not reported. Herein, we present a case of MHCW in a 5 month old infant presenting with acute respiratory distress with history of respiratory problem at 3 months of age. Child was suspected to have persistent pneumonia in view of radiological findings. Child's respiratory distress improved with antibiotics and bronchodilators. Respiratory symptoms in MHCW are due to extrinsic compression of lung parenchyma. Present case had respiratory symptoms with persistent radiological findings leading to suspicion of persistent pneumonia. His respiratory symptoms and exacerbation on follow up was attributed to hyper reactive airway disease and MHCW was managed conservatively. The non-neoplastic nature, characteristic presentation, histopathology, imaging modality and management options of MHCW are discussed.

Keywords: Benign chest wall mass, Benign tumour, Persistent pneumonia

CASE REPORT

A five-month-old infant presented with complaints of recurrent cough, cold, fast breathing since 2 months of age. At presentation, child had fast breathing, fever, chest retraction and poor oral intake. There was positive history of asthma in the family. Physical examination revealed marked tachypnea with subcostal, intercostal retractions and decreased air entry over right lower lung field with bilateral wheeze. There was a hard mass palpable in the anterior region of chest overlying the 5th, 6th and 7th ribs on right side. Systemic examination was unremarkable except for the respiratory findings. Clinical possibilities kept were community acquired pneumonia with underlying collapse consolidation, pulmonary agenesis and malignant neoplasm arising from rib. As child was symptomatic since two months of age, causes of persistent pneumonia like congenital anomaly of airways, tuberculosis, cystic fibrosis, immunodeficiency was also kept. Chest X- ray revealed opacity of right lung with deformed 5th, 6th, 7th ribs raising the suspicion of a malignant lesion [Table/Fig-1]. USG thorax showed 5 x 6 cm soft tissue mass in the parietal wall.

Child was managed with bronchodilators, antibiotics and oxygen inhalation. There was marked improvement in respiratory distress with these measures. CECT finding pointed towards a diagnosis of mesenchymal hamartoma of chest wall [Table/Fig-2]. As child showed marked response to bronchodilator therapy (salbutamol

nebulization), a possibility of underlying hyperreactive airway disease was also kept. FNAC from the mass showed clusters of spindle cells admixed with richly vascular chondromyxoid stroma. Bone scan showed homogenous radiotracer uptake in right thoracic mass. A biopsy from chest wall showed fragments of cellular cartilage surrounded by spindle shaped mesenchymal cells. Paediatric surgery opinion was obtained. As the child's respiratory distress settled, it was decided to keep him under close observation with periodic imaging. The child has been followed up for one year with two episodes of exacerbation of respiratory symptoms responding to medical measures and growth percentiles between 25th and 50th for the normal population.

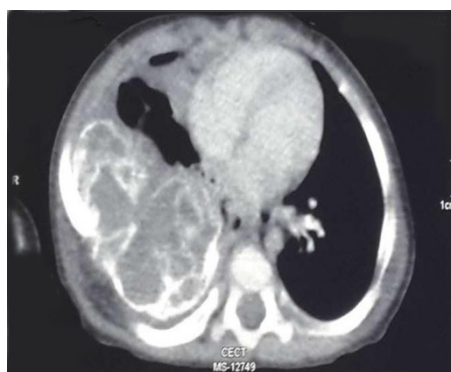
DISCUSSION

McLeod and Dahlin in 1979 gave the present accepted nomenclature of Mesenchymal hamartoma [1], Mesenchymoma, Infantile cartilaginous hamartoma [2] and infantile osteochondroma are other names used for this entity in the past.

Mesenchymal hamartomas are not true neoplasm, at times they have been classified as non-neoplastic development anomaly. It generally presents in neonates and infants [3], arising antenatally. Most of the times, a diagnosis on prenatal USG is possible. Origin is from ribs usually and may be multifocal [4]. It is almost always of congenital origin [5]. It has M:F ratio of 2:1. Chest wall deformity or mass and respiratory distress are usual presentation [6].

Recurrent respiratory symptoms with persistent radiological finding may raise the possibility of persistent pneumonia. At the same time if underlying respiratory symptoms are due to any other underlying condition, the respiratory symptoms may be wrongly attributed to mesenchymal hamartoma compressing the lung parenchyma.

Present case was a five-month-old infant with mesenchymal hamartoma of chest wall. He had respiratory symptoms which were explained by hyper reactive airway disease. We were able to manage it conservatively



[Table/Fig-1]: Chest X-ray right lung opacity with distortion of 5th, 6th, 7th ribs
[Table/Fig-2]: CECT Chest-large chest wall based heterogeneous lesion with matrix calcification and remodeling of underlying ribs with multiple enhancing septations

with a follow-up period of one year. The management of MHCW depends on respiratory symptoms. Surgery is preferred for children who are symptomatic secondary to compression by mass. Cameron D et al., in their case series of three patient reported conservative management of 2 patients with one of the patient requiring surgery [7]. Pawel BR et al., in their case report described a partially resected MHCW which had a benign course on further follow up [8]. Freeburn et al., reported a case of spontaneous regression of infantile MHCW by age of 11 years [9]. Their case also had respiratory symptoms attributed to asthma like in our case MHCW.

Mesenchymal hamartoma are well-circumscribed lesions arising from the central portions of ribs, associated with erosion of adjacent ribs at gross examination [10]. Radiographic appearance of well circumscribed mass with involvement of multiple ribs is diagnostic of MHCW in an infant.

CT scan helps to delineate site of origin, tumour density, effect on lung parenchyma and adjacent structures [11]. CT best demonstrates matrix mineralization, whereas MRI best demonstrates haemorrhagic cystic lesion [6].

Definitive diagnosis is by histopathology examination [4]. Microscopically, the lesion is composed of cartilage, bone and spindle proliferation. Cystic spaces suggest diagnosis of an aneurysmal bone cyst. Spindle cells are arranged in loose fashion. Cartilage is in form of plates maturing into trabecular bone, simulating the appearance of epiphyseal plates.

Other differentials for such mass lesions of chest are fibrous dysplasia and haemangiomas arising from ribs. Langerhans cell histiocytosis, Ewing sarcoma or PNET with large soft tissue mass and involvement of ribs (destruction, sclerosis) is a close differential. But these can be differentiated on basis of rib origin, presence of secondary aneurysmal bone cyst component with characteristic radiological finding. Because of morbidity associated with surgery, conservative management can be tried with surgical resection in symptomatic patient at presentation or follow up [7]. Management is guided by clinical presentation with surgery being curative. Surgery is indicated if compression by the mass causes pulmonary or cardiac compromise. Conservative management by close follow up and periodic imaging is advocated as spontaneous regression can

occur and large residual chest wall defect after en bloc resection can frequently result in a scoliosis. Surgery may also be complicated by large amount of blood loss due to haemorrhagic elements. Recent reports continue to support attempted total resection. Long term data on conservative management outcomes are lacking.

CONCLUSION

MHCW with its clinical presentation and radiological features may be alarming and may lead to suspicion of malignant lesion unless clinician is aware of this entity. With better understanding of natural history of this benign condition, a trend towards conservative management especially in asymptomatic cases is likely. There is need for data on long term follow up on cases managed conservatively. Malignant transformation in primary mass or after resection is rare.

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