Scholars Journal of Medical Case Reports

Sch J Med Case Rep 2016; 4(4):209-211 ©Scholars Academic and Scientific Publishers (SAS Publishers) (An International Publisher for Academic and Scientific Resources) ISSN 2347-6559 (Online) ISSN 2347-9507 (Print)

DOI: 10.36347/sjmcr.2016.v04i04.009

Mesenchymal Hamartoma of Chest Wall - A Case Presentation

Dr. Prashant Sadashiv Patil, Dr. Rahul Kumar Gupta, Dr. Paras Kothari, Dr. Abhaya Gupta, Dr. Ravikiran Kamble, Dr. K Vishesh Dikshit, Dr. Geeta Kekre

Department of Pediatric Surgery, LTMMC & LTMGH Sion, Mumbai, India

*Corresponding author

Dr. Prashant Sadashiv Patil

Email: docprashant2010@gmail.com

Abstract: 1 year-old male was presented with complaints of chest wall swelling and features of upper respiratory tract infection. Investigations, including CT chest and trucut biopsy were suggestive of chest wall hamartoma. Left thoracotomy was done and tumor excised into along with involved ribs. The defect was reconstructed with prolene mesh and lattissimus dorsi flaps. Postoperative period was uneventful. Histopathological examination confirmed the diagnosis of chest wall hamartoma. Followup CT chest after 6 months was normal.

Keywords: Chest wall swelling, thoracotomy, chest wall hamartoma

INTRODUCTION

Mesenchymal hamartoma of the chest wall (MHCW) is a very rare, benign tumor with unique location (arising from the ribs), distinct clinical, radiological and histopathologic characteristics [1-4]. It may arise antenatally and typically present at birth or in early infancy as a chest wall mass with or without respiratory distress, and with marked rib deformity. The tumor arises from the ribs and consists of partially mineralized focal overgrowths of normal skeletal elements with no malignant tendency. There is slight male preponderance (Male to female ratio: 2:1), and the age at the time of presentation ranges from birth to 16 years. About half of these patients have respiratory symptoms or signs at presentation.

CASE REPORT

A 1 year old male child was brought with complaints of deformity of chest on left side since 5-6 months and cough, fever, and mild breathlessness since 5 days. There was no history of anorexia /weight loss/contact with T.B.

Examination revealed stable and active child. There was left sided chest wall deformity with protrusion of 6, 7, 8 and 9th ribs laterally and depression posteriorly. Visible veins were present on chest. Left nipple was at a higher level than right. Chest movements were decreased on the left side. Palpation showed a firm to hard swelling on left side with rib deformity. Apex beat had shifted to the right side. Tactile vocal fremitus was decreased on left side. Trachea was in central position. Dull note was present on left side except apical region. On right side resonant note was present. Right side air entry was normal. Left

mid and lower zones air entry was absent .No adventitious sounds.

Investigations including X ray chest and CT chest revealed space-occupying lesion in the left posterior mediastinum with destruction of 6th to 10th ribs. Child was further investigated with Trucut biopsy, which was suggestive of chest wall hamartoma. Metastatic work up was negative. Child underwent surgical exploration through left muscle sparing thoracotomy. It revealed a large, lobulated tumor measuring 10x15 cm arising from inner side of posterolateral chest wall involving 6th, 7th, 8th, 9th and 10th ribs without mediastinal lymphadenopathy. Wide excision of the tumor (margin of about 2-3 cm resected all around) after securing feeding vessels from aorta, along with partial resection of 6th, 7th, 8th, 9th and 10th ribs was done (Fig. 2). Tumor margins were marked with staples after excision. Large defect was closed using Prolene mesh: and Lattissimus dorsi and Serratus anterior flaps (Fig. 3). Post operatively chest wall strapping was given for about a period of one week. Postoperative period was uneventful. Histopathological examination of the resected specimen confirmed the diagnosis of Chest wall hamartoma. Follow-up after 6 months child was asymptomatic. CT chest showed no evidence of recurrence of the tumor. This case was presented because of rarity of the tumor (about 60 cases were reported in the literature).



Fig. 1: Clinical photo of patient with left chest deformity



Fig. 2: X-ray chest lateral view

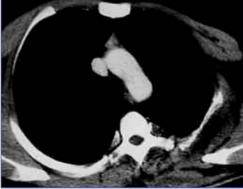


Fig. 3: CT thorax showing mass in left post.

Mediastinum

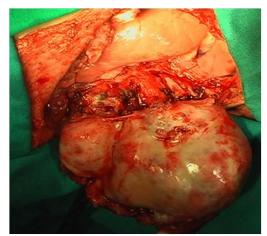


Fig. 4: Intra-op photo showing tumor



Fig. 5: Wound closure after wide local excision of tumor

Diagnosis

The chest radiograph show a large partially calcified well-defined extrapleural mass arising from one or more than one ribs. The involved ribs may be expanded, distorted and partially destroyed. CT scan is the optimal method of imaging and helps to determine the site of origin, tumor density, enlargement and effect on adjacent structures. MRI has also been used as an alternative imaging modality. They show the pathognomonic features of rib origin, mineralized matrix, and hemorrhagic cystic components associated with mesenchymal hamartoma [5].

The definitive diagnosis of MHCW is established only by histopathological examination. MHCW are well circumscribed, and the cut surface shows both cystic and solid components, and composed of two or more cell types of mesenchymal origin not ordinarily found together. Microsopically, this tumor is composed predominantly of chondroid tissue with large endothelium lined blood spaces and primitive mesenchyme with osteoclastic giant cells, endochondral ossification and maturation to trabecular bone [6, 7].

Differential diagnosis

The tumor should be differentiated from chondroblastoma, osteo-blastoma, aneurysmal bone

cyst, and other mesenchymal neoplasms. Radiologically this lesion can sometimes be confused with malignant tumor: congenital neuroblastoma, Ewing's sarcoma and malignant teratoma.

Surgical excision of mesenchymal hamartoma of the chest wall is recommended if compressive effects of the mass result in cardiac or pulmonary compromise or if the mass is physically deforming. Small asymptomatic lesions may not require surgical resection. Close observation and imaging may represent a more appropriate course of management, since spontaneous lesion regression has been reported [8].

In conclusion, mesenchymal hamartomas of the chest wall are unusual rib lesions most commonly affecting infants. The clinical manifestation and radiographic appearance may suggest a more aggressive malignant process unless one is familiar with this diagnosis. CT and MR imaging findings clearly reflect the underlying histopathologic characteristics of these lesions, demonstrating pathognomonic features of rib origin, mineralized matrix, and hemorrhagic cystic components. Accurate diagnosis allows appropriate treatment avoiding unnecessary chemotherapy and radical surgeries.

REFERENCES

- 1. Dounies R, Chwals WJ, Lally KP, Isaacs H, Senac MO, Hanson BA, Mahour GH, Sherman NJ; Hamartomas of the chest wall in infants. The Annals of thoracic surgery, 1994; 57(4):868-75.
- 2. Shamberger RC, Grier HE; Chest wall tumors in infants and children. Semin Pediatr Surg, 1994; 3(4):267-276.
- 3. Odell JM, Benjamin DR; Mesenchymal hamartoma of chest wall in infancy: natural history of two cases. Pediatr Pathol., 1986; 5(2):135-146.
- 4. McLeod RA, Dahlin DC; Hamartoma (mesenchymoma) of the chest wall in infancy. Radiology, 1979; 131(3):657-661
- Schlesinger AE, Smith MB, Genez BM, McMahon DP, Swaney JJ; Chest wall mesenchymoma (hamartoma) in infancy: CT and MR findings. Pediatr Radiol., 1989; 19(3):212-213.
- 6. Castellano VM, Fiano C, Vargas J, de Agustin P; Mesenchymal hamartoma of the chest wall. Cytopathology, 1997; 8(3):210-219.
- Nicholson SA, Hill DA, Foster KW, McAlister WH, Davila RM, Dehner LP; Fine-needle aspiration cytology of mesenchymal hamartoma of the chest wall. Diagn Cytopathol 2000; 22(1):33-38.
- 8. Brand T, Hatch EI, Schaller RT, Stevenson JK, Arensman RM, Schwartz MZ; Surgical management of the infant with mesenchymal hamartoma of the chest wall. J Pediatr Surg., 1986; 21(6):556-558.