Case Report

Mesenchymal (chondromatous) hamartoma of chest wall in a neonate: A case report and review of literature


* Correspondence Info:
Dr. Sahil I. Panjvani, MD Pathology
D-2/12 Officer’s quarters, Sangam cross roads,
Near Vaibhav Society, Harni Road. Vadodara-390022. Gujarat-India
Email- dr.sahilpanjvani@gmail.com

Abstract
Mesenchymal hamartoma of chest wall (MHCW) is a rare benign chest wall tumor mostly presenting at birth or in the first year of life with typical clinical, radiological as well as histopathological features. Here we report a case of this rare entity which can be easily mistaken for a malignant tumor. In this case preoperative probable diagnosis of some malignant tumor was made on clinicoradiological features and the final diagnosis of MHCW was given histopathologically.

Keywords: Benign; malignant; mesenchymal hamartoma; tumor.

1. Introduction
Mesenchymal hamartoma of chest wall is a very lesion of early infancy and childhood. It always arises in the rib and constitutes benign proliferation of skeletal tissue characterized by a prominent cartilaginous component and haemorrhagic cavities (secondary aneurismal bone cyst). Although the most common presenting manifestation at birth is a deforming chest wall mass, many cases are discovered incidentally on chest radiographs which may be obtained during the evaluation of the patient with respiratory symptoms. Patients with large intrathoracic mesenchymal hamartomas may rarely end with severe and fatal outcome like respiratory compromise.1-2 Despite the clinical, radiological and some histological features are highly suggestive of a malignant process, MHCW is a benign condition. The recommended treatment is controversial, with some favor early surgical intervention while others prefer conservative approach.3-5

2. Case report
A full term 2 days old male neonate presented with a large lump on the anterior aspect of right side of chest wall. The baby was born to a 28 years old primi mother with uncomplicated pregnancy. No resuscitation was required at birth. The patient was admitted for the evaluation for the chest wall mass. Physical examination revealed a healthy newborn with normal vital signs. There was approximately a 5X3 cm nontender hard lump in the region of middle part of right side of chest wall. Other findings were unremarkable. A chest x ray revealed a calcified mass arising from 4th and 5th right sided ribs. A CT was ordered and showed an expansile mass with 5X3.5X2.5cm in size, arising from right fourth and fifth anterior ribs with some calcification. Based on these finding a probable diagnosis of some malignant tumor of bone was made. The patient had undergone for a complete resection of the chest wall mass for further evaluation for histopathology. The specimen was sent to our department.

Specimen consisted (figure-1) of two ribs along with cystic mass. The heamorrhagic cystic mass was measuring 5X3.5X2.5cm. outer surface was spongy and brownish with area of cartilage measuring 2X2cm in size. Multiple sections (figure-2,3,4,5) revealed the cystic spaces were filled with blood and walls were composed of fibrous tissue, reactive woven bone with osteoclast like giant cells. The lumens of the cysts were lined by fibroblasts, collagen and occasional macrophages. There was also presence of hyaline cartilage with enchondral ossification. No evidence of malignancy was seen. The final diagnosis of mesenchymal (chondromatous) hamartoma of chest wall was made.
3. Discussion

Mesenchymal hamartoma of chest wall (MCWH) is a rare lesion of infancy and childhood. This entity was probably firstly described by Nash and stout in 1961. The estimated incidence is about 1/3000 among primary bone tumors. Other names includes mesenchymoma, infantile osteochondroma and infantile cartilaginous hamartoma. The currently accepted nomenclature was initially proposed in 1979 by McLeod and Dahlin. This name best reflects the benign nature and multiple histologic component of this lesion. MCWH is not a true neoplasm and is composed of maturing, proliferating and normal skeletal elements, with no propensity for invasion or metastasis.

The natural history is not well understood. Prenatal diagnosis is probably done by ultrasonography, but as shown the most MCWH are diagnosed postnatally. Lung hypoplasia is not common. Usually regression starts within the first year of life after a period of further growth.

Mesenchymal hamartoma is a well circumscribed lesion arising from the central portions of ribs associated with erosion of adjacent ribs. Adjacent structures are compressed by virtue of lesion size, expansion extrapleural mass effect. However, no invasive characteristic are seen.

Patients with MCWH are usually diagnosed at or shortly after birth with respiratory distress or a palpable mass. Typically the lesion appears as hard, immobile subcutaneous and extrapleural chest wall mass with deformation of one or more ribs. Most of all MCWH are diagnosed within the first six months of life. Later and incidental diagnoses have also been reported.

On chest radiography MCWH presents as an ipsi- or bilateral mass with calcific density. The lesion erodes the ribs and leads to destruction of the chest wall. Reactive bone formation is common. Chest CT reveals heterogenous rib lesions, extrapleural soft tissue masses with mineralization and regions of soft tissue attenuation. MRI can help to differentiate aneurismal bone cysts from MCWH.

The various histopathological of MCWH are described by many authors. These are highly cellular small
round, oval or spindle mesenchymal cells and fragments of hyaline cartilages with blood filled cavities. Immunohistochemical staining may demonstrates presence of S-100 protein in cartilaginous areas. The clinicoradilogical features with some histological findings like actively proliferating fibroblastic and chondroid elements may suggest malignant rather than a benign process and provoke potential dangerous over-treatment. The main differential diagnosis of MHCW includes aneurysmal bone cysts and primary bone tumors. In contradiction to the malignant tumors, which require surgery, radiation and/or chemotherapy with a relatively poor prognosis, mesenchymal haemartomas of chest wall are typically cured with complete surgical excision. Recurrences have been noted with incomplete resection. The most important post surgical complication is scoliosis, which appears in about 20% patients. In conclusion, mesenchymal hamartoma of chest wall is an unusual rib lesion which commonly affects infants. The clinical features and radiological appearance may suggest a more aggressive malignant lesion unless one is familiar with this entity. Appropriate diagnosis allows proper treatment which may consists of either close clinical follow-up or surgical resection in symptomatic cases or lesions causing chest wall deformity.

Acknowledgement

We are thankful to our senior histopathology technicians Mrs. Bhamini M McEwan and Mr. Moses McEwan for their dedication in their work by providing good quality of sections, slides and staining, so that we could diagnose this entity.

References