CASE REPORT

Cytohistomorphological identification of congenital bilateral mesenchymal hamartoma of chest wall

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Abstract

Cases of Congenital hamartoma of the chest wall in infants are rare and their bilateral involvement is extremely rare. An 18 month-old child presented with bilateral chest wall swellings since birth which were gradually increasing in size. The X-Ray of the chest wall revealed bilateral involvement of 1st, 2nd and 3rd ribs with involvement of extra-pleural soft tissues. The cytological diagnosis given was round cell tumor of the chest wall keeping in view the presence of round cells in a hemorrhagic background. The biopsy revealed areas showing mature cartilage, bone, fibrous tissues along with hemorrhagic cystic cavities. Basing on the biopsy report, final diagnosis of chest wall hamartoma was given. Following complete excision, the child was doing well and there was no recurrence of the lesion as observed in the follow-up sessions.

Key words

Chest wall, Child, Hamartoma

1 Introduction

Congenital mesenchymal hamartoma is a rare condition involving newborn infants, accounting for 0.03% of primary bone tumor cases [1]. It was first described as intrathoracic mesenchymoma [2] which was later on replaced by hamartoma due to its benign nature [3, 4]. It generally comprise of mixture of cartilaginous component, fibrous tissues and cystic cavities. In most of the children, the lesion noticed at birth gradually increases in size as seen in our case. But the child in the present report had bilateral masses involving both the sides of the chest wall which gradually increased in size, leaving her parents panicking. The increase in size of the chest wall mass can lead to respiratory distress in few rare cases [3], but it was not seen in our case. Very few such cases have been reported in the English medical literature. Only two cases have been reported stating more than one site involvement of the congenital hamartoma in newborns & infants [5, 6]. In the present case report, we have enumerated the cytological and histopathological features of the bilateral involvement of this entity which is extremely rare.

2 Case report

An 18 month-old female child presented with bilateral chest wall masses since birth which gradually increased to a size of 3 cm on right side and 2 cm on left side (Figure 1). The parents, being worried about the gradual increase in size, came to the clinician for consultation. These masses were not tender, rather were hard in consistency and fixed. The chest X-Ray of the child revealed bilateral expansive masses involving 1st, 2nd and 3rd ribs with cortical destruction and erosion (Figure 2). This was accompanied by extra pleural soft tissue mass. The hematological parameters of the child were within normal limits. Repeated fine needle aspiration cytology from different areas of the swellings revealed few clusters of round cells in a hemorrhagic background. The provisional cytological diagnosis given was round cell tumor of the chest wall keeping in view the clinical diagnosis of Askin's tumor and the patient was advised to go for biopsy.

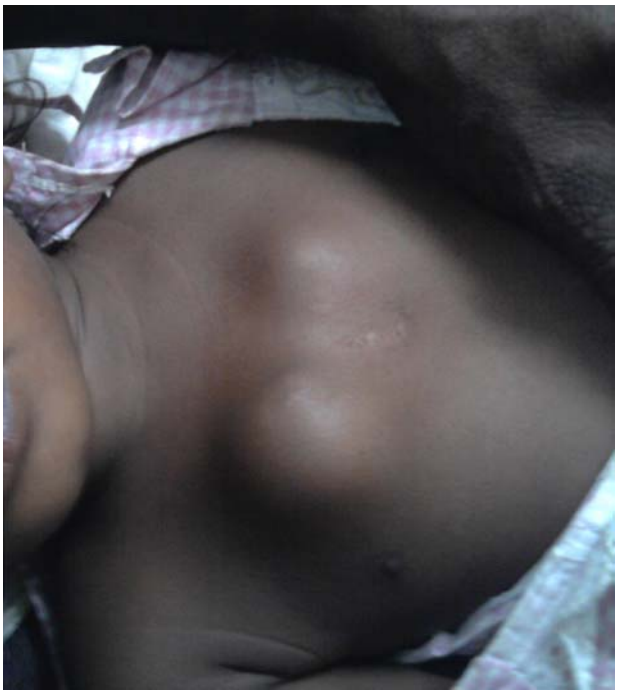

Figure 1. Child presenting with bilateral chest wall swellings

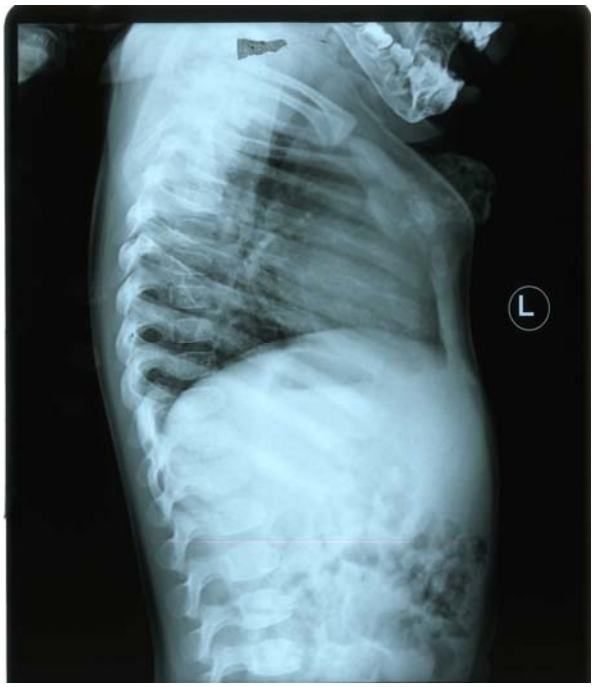

Figure 2. Chest X-Ray showing multiple ribs cortical destruction

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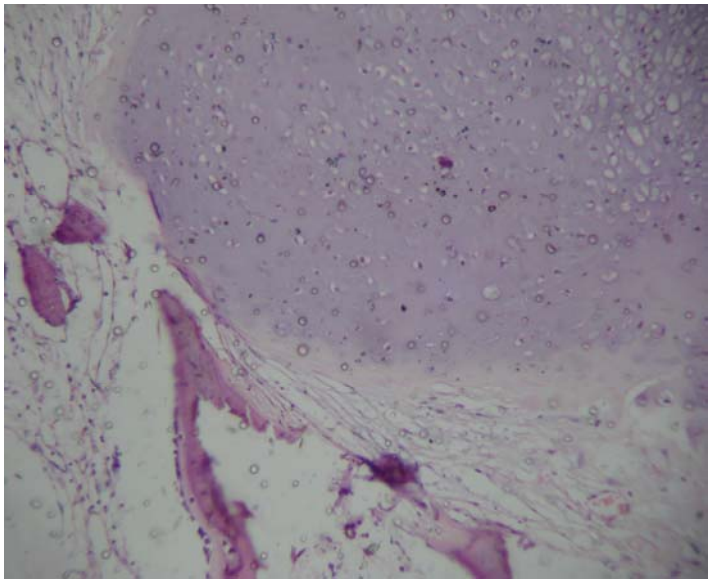

Figure 3. Microphotograph showing areas of cartilage, bone and fibrous tissues(100X, H&E)

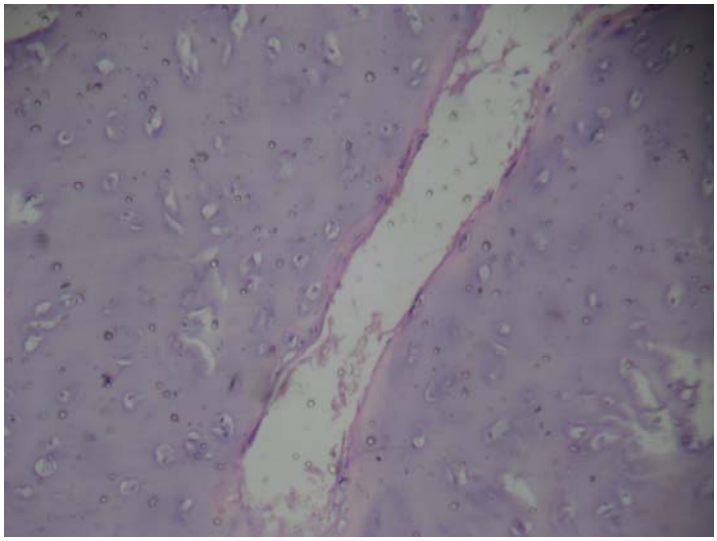

Figure 4. Microphotograph showing blood filled cystic space and cartilage(400X, H& E)

Both the masses were excised and the gross received showed irregular greyish white cartilaginous bony structures with attached fat measuring 6×3×2 cm^3. Cut-section was gritty, whitish with greyish areas in between. Microsections revealed areas of cartilage, fibrous tissues, bony tissues, giant cells and few blood-filled spaces (Figure 3 & Figure 4). The final histopathological diagnosis given was congenital mesenchymal hamartoma of the chest wall (B/L).

The child was followed up for a period of 12 months and it was found that she was doing well and there was no further recurrence of the disease.

3 Discussion

Although mesenchymal hamartoma of the chest wall is rare in infants and children, its bilateral presentation, as noticed in our case, is extremely rare. Other synonyms of this entity include mesenchymoma [7], infantile osteochondroma [8] and infantile cartilaginous hamartoma [9]. But, the currently accepted name of mesenchymal or chest wall hamartoma of chest wall proposed by Mc Leod and Dahlin in 1979 [3], is benign in nature with multiple histological components.

Mesenchymal hamartomas are not considered as true neoplasms because they are composed of benign, mature cartilage, bone, fibrous tissues and hemorrhagic cavities and lack invasion or metastasis [12].

Clinically, mesenchymal hamartomas are well circumscribed, painless mass arising from the central portion of the ribs which can involve and also erode adjacent ribs [6]. There is compression of the adjacent structures due to the size, expansion and extrapleural effect [10]. The chest X-Ray reveals large expansive lesion affecting multiple ribs with cortical destruction, erosion and extrapleural mass.

As many of the chest wall masses are malignant, accurate diagnosis of mesenchymal hamartoma is very important. The fine needle aspiration cytology (FNAC) of this lesion reveals lobules of hyaline cartilage, few spindle cells in an abundant chondroid matrix [11]. But, in our case, the cytology of the swellings revealed only few round cells found in a hemorrhagic background on repeated aspirations from different areas of the swellings. Thus, it was misinterpreted as small round cell tumor of the chest wall. Later on, the diagnosis was corrected in line with the biopsy findings. Many small round cell tumors including Ewing sarcoma, primitive neuroectodermal tumor (PNET) known as Askin tumor, can commonly occur as primary rib tumors [10]. In cytology, these reveal round cells in singles and sheets with rossetes formation.

Microscopic examination of mesenchymal hamartoma reveals mixture of proliferating cartilage, bone, spindle cells along with hemorrhagic cystic cavities representing secondary aneurysmal bone cyst (ABC) formation [12]. In our case, presence of mature cartilage, bone, spindle cells in sheets with cystic cavities have led to the histopathological diagnosis of mesenchymal hamartoma of the chest wall. Sometimes, the hypercellular proliferation of solid areas of proliferating mature cartilage and bone could mislead to the diagnosis of osteoblastoma, chondrosarcoma or osteosarcoma [13]. Chicken wire calcification seen in osteoblastoma and the malignant features like increased N: C ratio, hyperchromasia and nuclear atypia present in the latter two entities are notably absent in mesenchymal hamartoma [12]. The common benign lesions affecting ribs are fibrous dysplasia, and hemangioma. Though secondary ABC component can be seen in these conditions, but these are uncommon in newborns and Chinese letter pattern seen in fibrous dysplasia, lobular capillary proliferation in the latter dissect these from hamartoma.

Small, asymptomatic lesions do not require any surgical intervention which can be recommended in physical deformitity caused by hamartoma seen in our case and in cardiac or pulmonary compromise due to compressive effects of the mass [14]. Surgical intervention should be avoided as it could lead to severe blood loss and postoperative scoliosis. With close observation and proper investigations to diagnose mesenchymal hamartoma accurately, it can lead to the spontaneous regression of the lesion [2]. Cytopathologists should be aware of this rare lesion, as seen in our case in which we mistook this as round cell tumor in cytology for which the patient unnecessarily went through surgical excision. Histopathologists sometimes can misdiagnose the benign spindle cell elements in hamartoma as sarcoma in biopsy [15]. On histopathology, our case was correctly diagnosed as bilateral congenital hamartoma of the chest wall and on follow up, was seen to be doing well with no recurrence. While dealing with this type of presentation in a child, accurate diagnosis allows appropriate treatment which either consists of close clinical follow-up or surgical resection in symptomatic patients or lesions disfiguring the chest wall.

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