Case Report

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Bilateral Mesenchymal Hamartoma of Chest Wall in a Toddler: An Uncommon Clinical Entity

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ABSTRACT

Aim- Background: Bilateral chest wall mesenchymal hamartoma is a very rare lesion of infancy with distinct clinical, radiologic and pathologic characteristics. Computed tomography of the thoracic cavity is a very useful diagnostic tool. Histopathology examination offers the definite diagnosis.

Report of a case: Herein we present a pediatric case of congenital bilateral mesenchymal hamartoma of the chest wall in a toddler. Both chest masses were surgically removed, when the boy appeared having clinical manifestations and respiratory compromise. As a result, the patient got fully recovered and free of any symptoms.

Conclusions: As far as treatment of this uncommon tumorous condition is concerned, it is strongly supported that asymptomatic cases should be conservatively managed with close follow-up and periodic imaging. On the contrary, in cases presenting with clinical features, complete surgery treatment should be initiated and applied aiming the ultimate cure.

Key Words: *Mesenchymal hamartoma; chest wall; infancy; bilateral; congenital*

INTRODUCTION

Mesenchymal hamartoma of the chest wall is an uncommon benign tumorous condition [1]. It is usually discovered incidentally during infancy when a chest X-ray is conducted for any reason. It is a lesion of uncertain pathogenesis that arises from one or more ribs nearly, always in neonates or in early infancy [2]. It is thought to be a congenital malformation. Clinical features involve a visible by X-Rays chest wall mass, with or without concomitant respiratory distress [3]. Mesenchymal hamartoma usually runs a benign course. It may rarely results to a severe or fatal respiratory compromise [2]. Malignant transformation of the mass is reported to be extraordinary rare [2,4,5]. Multifocal or bilateral lesions are very rarely reported [6]. Treatment of choice is the surgical removal of the mass when there are symptoms such as respiratory compromise and problems secondary to compression of the lung [6].

The purpose of the current report is the presentation of an unusual case involving a bilateral mesenchymal hamartoma of the chest wall in childhood. Diagnostic and surgical treating procedures that have been performed are further described.

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CASE PRESENTATION

We present the case of an 18-month-old male toddler, weighing 15 kilograms, that was diagnosed with mesenchymal hamartoma arising from the chest wall at the age of 5 months.

According to medical history records, the case involves a full-term baby of a 39+4-week gestation age, with a birth weight of 3,510 gr, that was delivered vaginally.

Prenatal and postnatal periods are reported to be uncomplicated and uneventful. Developmental history and clinical condition are also reported uneventful. However, at the age of five months, parents requested medical help because of a progressively enlargement of the left shoulder that they first noticed three months ago. A chest X-Ray showed a large mass extending within the upper left hemithorax. Therefore, the infant was admitted to hospital for further evaluation. Haematological and biochemical parameters were within normal limit values. An ultrasound examination of the left hemithorax confirmed the existence of a large mass measuring 5, 5 cm×5,7cm× 6,5 cm. A computed-tomography scan (CT) of the chest was performed, which revealed the coexistence of two intra-thoracic masses. It indicated a solid mass arising from the first upper six left ribs measuring 7,3cm × 5,8cm × 7, 0 cm, destructing, distorting or partially eroding the ribs, compressing the lung and displacing the mediastinum.

A second mass deriving from the right seventh rib was also indicated. The second mass that measured 2,1cm × 2,7cm × 2,2cm was also deforming and distorting the rib. Atelectatic areas of pneumonic parenchyma were obvious. No lymph nodes were visible or other nearby lesions. Radiologists implied that the above mentioned lesion might consist of a multifocal Ewing lesion. A biopsy procedure of the left mass with Fine Needle Aspiration was applied. The subsequent histology examination revealed tissue elements of mesenchymal appearing fibroblastic hyperplastic cartilaginous lesions consisting of a mixture of spindle cells with little eosinophilic cytoplasm, islands of aneurysmal cyst-like areas with the presence of osteoclast-like giant cells, multi-nuclei giant cells with apparent eosinophilic cytoplasm and areas of woven bone formation, hyalinised material and endochondral ossification. As a result, the diagnosis of mesenchymal hamartoma chest wall was finalised.

The infant was then evaluated by the pediatric thoracic surgeon team. As the baby was in a fair asymptomatic condition, a conservative treatment was favored. However, in the course of follow up examination at the age of 18 months, it was noticed a progressive respiratory compromise. Another chest X-Ray (Figure 1) and computed tomography scan of the chest were repeated that showed further enlargement of both the masses bilaterally. Under these circumstances the treatment of choice established to be the surgical resection. At first, a right thoracotomy was applied (Figure 2). The whole tumorous mass arising from the right fourth rib was completely excised. The resected

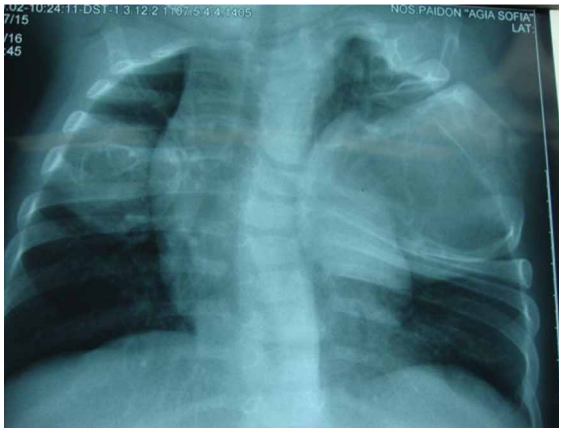

FIGURE 1. Plain chest X Ray shows the bilateral tumorous mass, deforming ribs and compressing lung, heart, vessels and spinal cord.

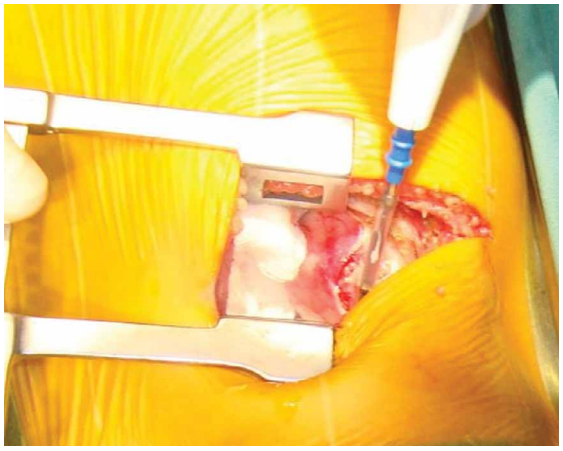

FIGURE 2. Images from the surgical procedure.

mass of the right hemithorax was measuring 5cm X 4,5cm X1,5cm and had a solid and white cystic appearance with areas of hemorrhage mixed with areas of cartilaginous development (Figures 3,4). The histological examination of

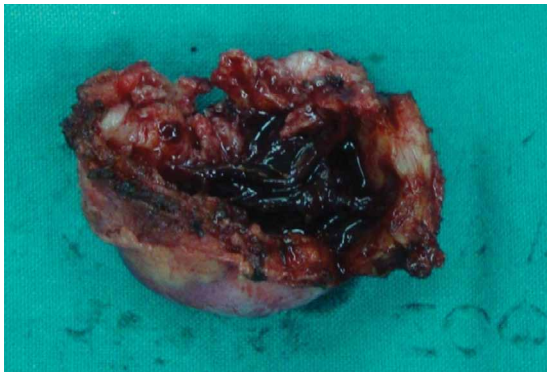

FIGURE 3. The surgically resected right mass.

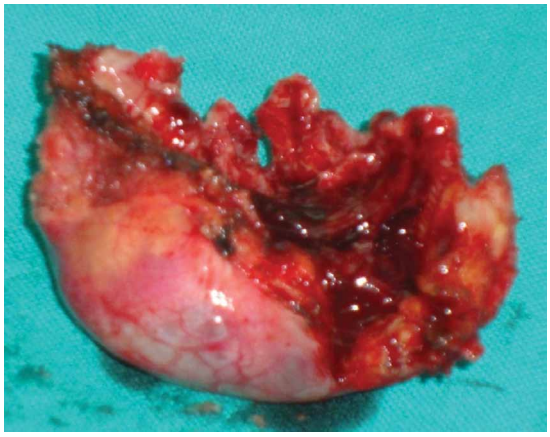

FIGURE 4. Images of the resected right mass.

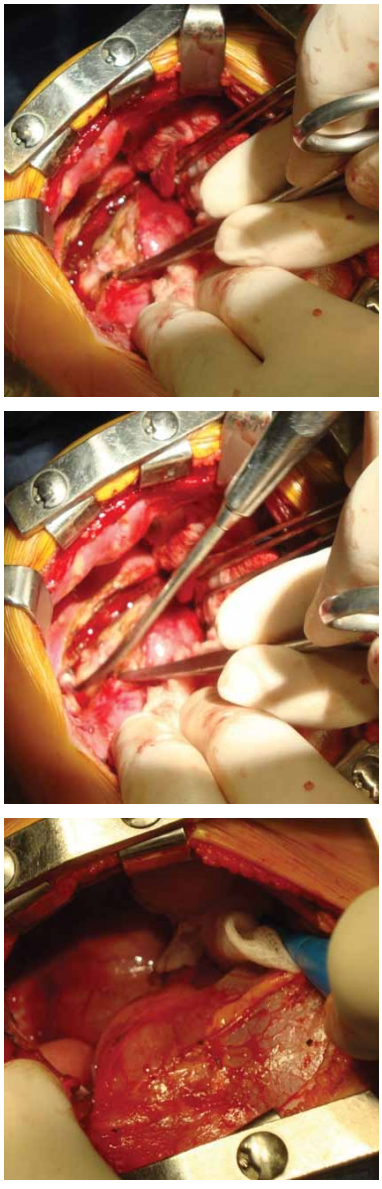

FIGURE 5. Images from the second surgical procedure.

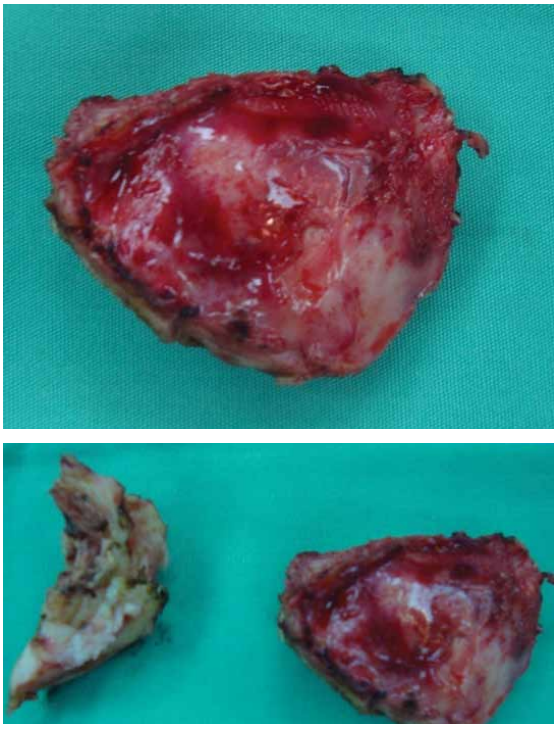

FIGURE 6. Images of the resected left tumorous tissue.

the resected tumour confirmed once again the diagnosis of mesenchymal hamartoma deriving from the chest wall. In a second surgical time, the other concomitant mass was also completely resected along with a part of ribs (Figures 5,6). The mass was located in the left hemithorax arising from the 2nd, 3rd and 4th rib. It measured 12cm X 7cm X7cm deforming the ribs neighboring heart and big heart vessels, lying till the spinal cord, compressing lung and diaphragm. Despite the size of the bilateral tumour and the pre-existent clinical features, the toddler underwent an uneventful operative and postoperative course. The boy was fully recovered and free of symptoms. Post-operative follow-up at one month, three months, six months, one year and two years showed no evidence of recurrence clinically or radiologically.

DISCUSSION

Mesenchymal hamartoma chest wall is a rare benign tumorous lesion. This abnormality is estimated to be rather a congenital situation seen mostly in neonates and infants. A percentage of 80% of reported cases involve early infancy. Very few cases involving adults have been reported [3,7,8]. It is more common in males in an estimated frequency 2:1 to4:1 compared to females [2,3,9]. It is mostly unilateral, located favoring the right hemithorax [2]. Bilateral mesenchymal chest wall hamartomas occur extremely rarely [6]. Very few cases have been reported in

literature. Its incidence is estimated to be 1 in 3000 among primary bone tumors and <1 in million in the general population [10,11]. To this date, nearly 80 cases have been reported usually in the form of case presentation [6,7]. Therefore, MHCW is an uncommon, benign clinical entity with multiple histological components and rare possibility of malignant transformation [12].

In the vast majority of cases, the mass is large and well-delineated, arises from the central part of ribs and compresses surrounding organs. It causes deformity of the thoracic wall [7]. It may be asymptomatic and as a result an incidental finding in a chest X-Ray. Other times, it may also present as a palpable or visible chest mass. In certain cases, it may cause respiratory symptoms and compromise because of the subsequent lung compression or cardiovascular compression [13,14,15]. Sometimes, neurological symptoms may present as a result of spinal compression.

Diagnosis is based on imaging methods like chest X-Ray, computed tomography or MRI. Typically the lesion is well-circumscribed deriving from ribs. Final diagnosis is made by histological findings. The combination of spindle mesenchymal cells with no atypia or abnormal mitotic activity, the cartilaginous proliferation, the islands of aneurysmal cysts, the presence of osteoclast like giant cells, fragment of hyaline cartilage, the woven bone formation and endochondral ossification consist histological findings diagnostic of the MHCW [13,15]. Differential diagnosis might include other mass lesions such as fibrous dysplasias, haemangiomas, congenital neuroblastoma, Ewing's sarcoma, malignant teratoma, Langerhans cell histiocytosis, PNET osteosarcomas, chondrosarcomas, osteochondromas [15].

Surgical resection of the mass is the only final curable therapeutic process. However, a few authors describe in their papers that conservative management should be firstly preferable. They claim that in some cases MHCW stops growing within the first year of life. There has been registered the possibility of a spontaneous regression [15]. Under this spectrum, conservative management that includes close follow up and periodic imaging should be applied in advance. On the other hand, it seems that management is guided by clinical manifestation. When there are symptoms such as respiratory or cardiovascular compromise as a result of the mass causing compression phaenomena to neighboring tissues, radical surgical treatment is obligatory and considered therapy of choice [16,17]. The prognosis after surgery appears to evolve excellently. Postsurgical complications have been reported, the most important of which is postsurgical scoliosis that affects almost 20% of patients undergoing the surgical resection of the intra-thoracic tumour [3,15,16]. Chemotherapy or radiotherapy do not constitute part of treatment guidelines. Whether surgical treatment should be applied immediately after diagnosis is established or if conservative management should be first favored is still a matter of controversy among researchers, as long term data on conservative approach outcome is lacking. Therefore many reports continue to support early and total resection as the only adequate therapy to avoid life threatening respiratory complications.

CONCLUSION

To conclude, bilateral chest wall mesenchymal hamartoma is a very rare lesion of infancy with distinct clinical, radiologic and pathologic characteristics. Computed tomography of the thoracic cavity is a very useful diagnostic tool. Histopathology examination offers a definite diagnosis. As far as treatment is concerned it is strongly supported that asymptomatic cases should be conservatively managed with close follow-up and periodic imaging. On the contrary, in cases presenting with clinical features, complete surgery treatment should be initiated and applied aiming at the ultimate cure.

Conflict of Interest: *The authors declare that they have no conflict of interest. The authors state that they have full control of all primary data and agree to allow the journal to review their data if requested.*

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