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Case Report

A case of mesenchymal hamartoma of the chest wall of a child*

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ABSTRACT

Mesenchymal hamartoma of the chest wall is a rare benign tumor that usually occurs in infants and children. The clinical presentations and imaging features are atypical and difficult to differentiate from malignant tumors. In this article, we present a case with a large mesenchymal hamartoma tumor of the chest wall. A large right-sided chest wall mass was discovered in a 6-month-old boy by his mother. Chest X-ray revealed a thoracic mass with well-defined margins on the right side that expanded into the right ribs. Chest computed tomography showed that the mass originated from the thoracic wall. The patient underwent complete removal of the mass, and histopathology results confirmed a mesenchymal hamartoma.

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Introduction

Mesenchymal hamartoma of the chest wall (MHCW) is a tumor that arises from one or more ribs [1], which typically

appears during the neonatal period and in childhood [2]. MHCW generally presents as unilateral and solitary, but multiple or bilateral MHCW have been reported [3]. Patients can be asymptomatic, but large MHCWs can compress the lung parenchyma and trachea, leading to respiratory distress [4].

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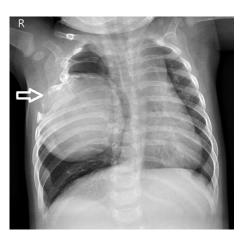


Fig. 1 – Chest X-ray revealed a large mass in the right chest. The mass expanded and disrupted the fourth rib (arrow) and displaced the mediastinum to the left. The mass had well-defined margins

MHCW is a benign tumor, and malignant transformation is extremely rare [5]. Observation and resection are two approaches typically applied for the management of MHCW [6]. Here, we report a case of large MHCW in a child that was discovered by his mother.

Case report

A 6 month-old boy presented with a mass in the right chest that was discovered by his mother. The patient had no other medical history. Physical examination revealed a hard, immobile mass in the right posterior chest wall. Breath sounds were markedly decreased on the right lung. On admission, the patient was in mild respiratory distress, with a respiratory rate of 57 breaths/min. Chest X-ray showed a right-sided, large chest wall mass causing expansion and destruction of the fourth rib and displacement of the mediastinum to the left (Fig. 1). Chest computed tomography (CT) scans revealed a large mass, measuring 5.7 cm imes 4.1 cm, arising from the right posterior chest wall (Fig. 2). The mass presented with well-defined borders and heterogeneous density. The fourth rib was involved and undergoing destruction without periosteal reaction. Coarse calcification was observed. The tumor compressed the right bronchus. Chest CT with contrast agent showed heterogeneous enhancement of the mass.

The patient underwent complete resection, and histopathological results confirmed an MHCW. Chest X-ray performed after surgery showed complete tumor removal, and the right lung volume was normal (Fig. 3). Six months after surgery, the patient had normal development without respiratory systoms.

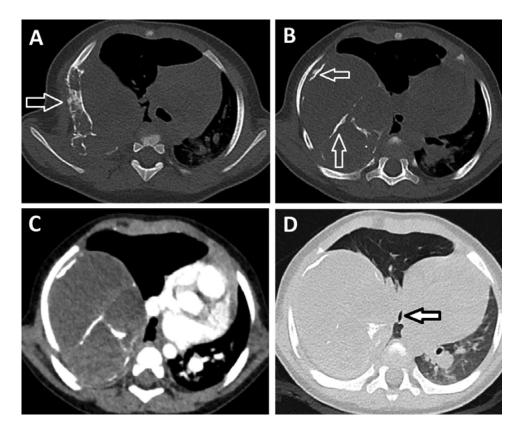


Fig. 2 – Chest computed tomography scans of the patient. The mass originated from the right thoracic wall destructing the fourth rib without any periosteal reaction (A, arrow). The mass contained coarse calcifications (B, arrow) and enhanced heterogeneously (C). The mass compressed the right bronchi and the heart (D, arrow)



Fig. 3 – The tumor was completely removed, and the right pulmonary volume returned to normal

Discussion

MCHW is a rare and benign entity, also known as benign mesenchymoma, infantile osteochondroma, and infantile cartilaginous hamartoma [5]. To date, more than 100 cases of MCHW have been reported in the literature [3]. Most MCHW cases present unilaterally and commonly arise on the right side, with a male predominance and a male-to-female ratio ranging from 2:1 to 4:1 [5]. The patient in this report was a boy, and the tumor arose from the right thoracic wall.

Although MCHW is benign, it can erode the ribs and enlarge, leading to thoracic deformity and displacement of the airways, lungs, heart, and major vessels [6]. Clinically, patients present with a subcutaneous chest wall mass that is hard and immobile, causing a chest wall deformity [7]. Other patients may present as asymptomatic or with life-threatening symptoms associated with severe respiratory distress, depending on the location and size of the tumor [2]. Cough, fever, and scoliosis may be present [2].

Chest X-ray shows a mass with well-defined sclerotic margins and popcorn-like speckled calcifications that involved and expanded the ribs [6,7]. Ultrasound shows a heterogeneous mass with solid, cystic, and focal calcification components [1]. Chest CT scans and magnetic resonance imaging (MRI) can be used to determine the lesion origins and the relationships with adjacent structures [8]. On CT scans, MCHW can be observed to originate from the thoracic wall and present with large, heterogeneous masses with cystic and solid regions, expanding into the ribs and containing calcifications [9]. On MRI, the lesions display heterogeneous signal intensity on T1-weighted and T2-weighted images, with both solid and cystic components [9]. The solid component displays mild to moderate enhancement. Sometimes, fluid-fluid levels can be detected, suggesting a secondary aneurysmal bone cyst [1]. Some differential diagnoses on imaging include other malignant tumors such as a primary Ewing sarcoma of chest wall, metastases from neuroblastoma, leukemia, and lymphoma, or benign lesion as fibrous dysplasia, hemangioma, and Langerhans cell histiocytosis [1]. MCHW may be misdiagnosed with primary bone aneurysm cyst [9].

Histologically, the tumor consists of cartilage, immature mesenchymal elements, aneurysmal bone cyst areas, and new bone formations [3]. MCHW can be managed with conservative or surgical; and complete surgical resection is the primary therapy option, especially in symptomatic patients [2]. Recurrences have been reported in cases in which complete resection was not possible [9]. The rate of malignant transformation is extremely rare [9]. Generally, MCHW has a good prognosis, especially in cases of complete resection [7].

The clinical findings of our patient were consistent with the imaging features. The tumor was large and compressed the right bronchi and pulmonary parenchyma. The chest radiograph suggested that the tumor originated from the chest wall due to the involvement of the ribs. The well-circumscribed margins indicated a benign lesion. The chest CT findings for this tumor were also typical of MHCW. The histological results and complete resection of the tumor indicate a good prognosis for this patient.

Conclusion

In conclusion, MHCWs are extremely rare and typically affect children. Although MHCW is benign, large tumors can cause serve respiratory distress. CT and MRI can provide useful findings for the diagnosis and support when planning treatment.

Author contributions

Nguyen NT and Nguyen MD contributed equally to this article as co-first authors. All authors have read the manuscript and agree to the contents.

Ethical Statement

Appropriate written informed consent was obtained for the publication of this case report and accompanying images.

Patient Consent

Informed consent for patient information to be published in this article was obtained.

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