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Respiratory insufficiency in a newborn with mesenchymal hamartoma of the chest wall occupying the thoracic cavity

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Abstract The authors describe a newborn patient with mesenchymal hamartoma of the chest wall associated with pulmonary hypoplasia. A massive thoracic tumor was diagnosed by prenatal ultrasonography and magnetic resonance imaging at the 28th week of gestation. She was delivered through cesarean delivery at the 36th gestational week. Respiratory distress because of pulmonary hypoplasia necessitated neonatal intensive care. The tumor extensively involved the left hemithorax including all 12 ribs and the first 10 thoracic vertebrae, resulting in marked deformity of the thorax. At 5 days of age, she underwent the incisional biopsy through a left thoracotomy. Histopathology of biopsy specimens showed multiple components of mesenchymal origin including premature cartilage, bone, and cystic lesions resembling aneurysmal bone cyst. The tumor then showed a rapid overgrowth, but subsequently exhibited a self-limited growth for months, in which her respiratory condition gradually improved to spontaneous breathing without oxygenation support. The present case advocates perinatal preparations for associated pulmonary hypoplasia and conservative management for the neoplasm in fetuses prenatally diagnosed as having this unique pathological entity.

Mesenchymal hamartoma of the chest wall is an extremely rare disease, which usually presents at birth or early in life [1]. The tumor often involves plural ribs and vertebrae, resulting in marked deformity of the thorax [2]. These lesions can be easily misinterpreted as chondrosarcoma, osteosarcoma, or malignant mesenchymoma owing to varied mesenchymal tissue components exhibiting marked immaturity or morphologic irregularity [3]. In this disease, most patients underwent surgical resection in infancy irrespective of their symptoms, and only limited information is available on the natural history of the disease [4,5].

In this report, we describe a neonate with a huge mesenchymal hamartoma of the chest wall associated with pulmonary hypoplasia who was treated conservatively with aggressive respiratory support for this rare neoplasm.

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1. Case report

A fetal patient was referred to our institution for closer examination of an intrathoracic mass at the 28th gestational week. Prenatal ultrasonography showed polyhydramnios and a progressively growing lesion in the left thoracic cavity. Intrauterine magnetic resonance imaging (MRI) at 32 weeks' gestation showed the left hemithorax occupied with a tumor consisting of heterogenous solid and cystic components (Fig. 1). The mediastinum was deviated to the right side, and the right lung was markedly compressed. No left lung was identified. She was born weighing 2410 g through cesarean delivery at 36 weeks' gestation. The tumor extensively involved the left hemithorax including all 12 ribs and the first 10 thoracic vertebrae, resulting in a marked deformity of the thorax (Fig. 2A). The respiratory movement was severely retracted and she appeared markedly cyanotic. Arterial blood gas analysis then indicated a pH of 6.752, PaO_2 of 71.7 mm Hg, $PaCO_2$ of 139.8 mm Hg, HCO_3^- of 18.4, and base excess of −24.1 mmol/L. An urgent intratracheal intubation was followed by mechanical ventilation with 100% oxygen. Intensive respiratory care using high-frequency oscillation (HFO) was required to treat persistent pulmonary hypertension because of pulmonary hypoplasia for the following 4 days.

At 5 days of age, she underwent an incisional biopsy of chest wall and intrathoracic lesions through the left thoracotomy. The tumor contained multiple solid and cystic lesions of heterogenous appearance, some of which were easily hemorrhagic (Fig. 2B). Histopathology of biopsy specimens proved multiple heterogeneous components of mesenchymal origin including premature cartilage, bone, and cystic lesions resembling aneurysmal bone cyst (Fig. 3). We performed conservative treatments for the lesion also because it involved the thorax too extensively to be properly reconstructed using prosthetic materials. The tumor exhibited a rapid growth of heterogeneous components for the next 3 months, but subsequently showed a self-limiting behavior within the first year of life. Her respiratory condition gradually improved; persistent pulmonary hypertension seen in the initial 4 days subsided, the endotracheal tube was removed at 4 months, and oxygenation support with mask was discontinued at 10 months of age. She is doing well with no respiratory

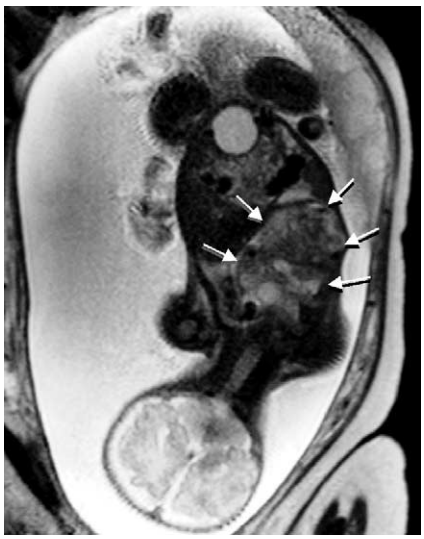

Fig. 1 Intrauterine MRI of a fetal patient with mesenchymal hamartoma of the chest wall at the 32nd week of gestation. The left hemithorax is occupied by the tumorous lesion consisting of multiple heterogenous solid and cystic components (arrows). The mediastinum is deviated to the right side, and the right lung is markedly compressed.

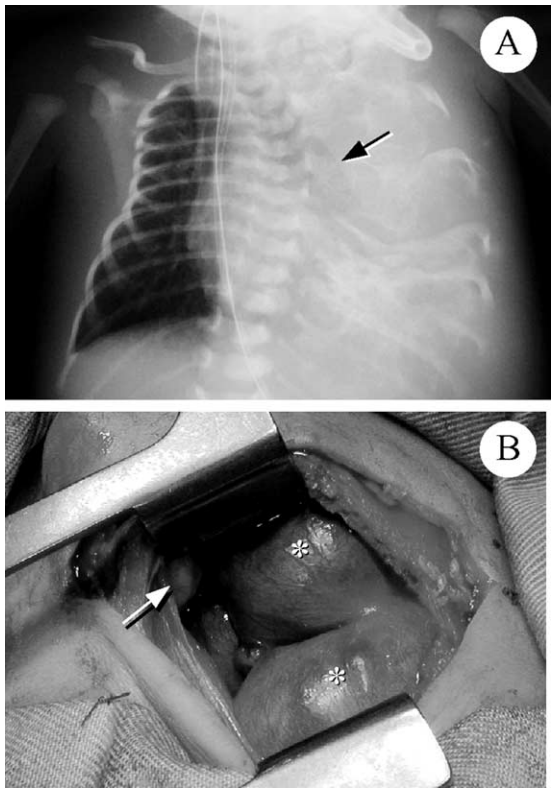

Fig. 2 A, A plain x-ray film of a newborn patient with mesenchymal hamartoma of the chest wall. The tumor extensively involves the left hemithorax including all 12 ribs and thoracic vertebrae (T1 through T10), resulting in marked deformity of the thorax. The left thoracic cavity is occupied by the tumor and the left lung appears severely hypoplastic (arrow). B, Intraoperative findings of the chest wall tumor at the age of 5 days. The tumor contains multiple solid and cystic lesions of heterogenous origin, some of which were easily hemorrhagic (asterisks).