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Nonsurgical management of mesenchymal hamartomas of the chest wall

Mila Kolar[a,*], Mercedes Pilkington[a], Mariana Silva[b], Donald Soboleski[c]

[a] Department of Surgery, Queen's University, Kingston, Ontario, Canada
[b] Department of Pediatrics, Queen's University, Kingston, Ontario, Canada
[c] Department of Radiology, Queen's University, Kingston, Ontario, Canada

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ABSTRACT

Background: Mesenchymal hamartomas of the chest wall (MHCW) are extremely rare extrapleural benign tumors. There is increasing utilization of nonoperative management given increased understanding of its benign biological behavior.

Methods: A review of cases utilizing nonoperative management was completed. We present a case of complete spontaneous resolution of a MHCW managed nonoperatively with radiologic surveillance for five years.

Results: Most reported cases were single case reports, with a total of 12 cases including our patient. From the reported cases on nonsurgical management eight were unilateral (67%), and four bilateral (33%). Two patients with unilateral involvement had multifocal lesions (29%). The majority of unilateral lesions were on the right side (85%). Three patients (27%) underwent subsequent surgical management after an initial period of conservative management. Two of these had bilateral involvement and the lesion was resected on only one side, while the third patient had unilateral multifocal involvement and underwent surgery for the predominant lesion only.

Conclusion: Mesenchymal hamartomas of the chest wall are extremely rare extrapleural benign tumors. There is increasing utilization of nonoperative management given increased understanding of its benign biological behavior. We present a case of complete spontaneous resolution of a MHCW managed nonoperatively with radiologic surveillance for five years.

1. Introduction

Mesenchymal hamartoma of the chest wall (MHCW) is a rare benign extrapleural tumor, which originates from the mesenchymal chondro-osseous tissue of the ribs. The lesion arises in the antenatal period and presents in the neonatal period or early infancy. It can present with varying degrees of respiratory distress, as an asymptomatic chest wall mass, a rib deformity, or may be an incidental finding on imaging [1–3]. With just 104 reported cases, it has an incidence of 0.03% among all primary bone tumors [1,2].

Due to its clinical, radiographic, and histologic features this lesion can be difficult to differentiate from more worrisome aggressive malignancies. The diagnosis can be challenging due to its large tumor size [4], initial period of rapid tumor growth [3], aggressive appearance on imaging (cortical rib erosions, rib destruction and deformities) [5], focal stromal hypercellularity, proliferative appearance on histology [6,7], or multifocal tumor origin [8]. While MHCW can be managed nonoperatively, it must be distinguished from the more common malignant chest wall tumors in children (mostly sarcomas) that require aggressive multimodality management [9–12].

Historically en bloc surgical resection was the most commonly reported treatment regardless of presentation [13–18]. Given that there have been no reported cases of malignant transformation or metastatic disease, it has been increasingly recognized that nonoperative management is safe. There have been several case reports describing nonoperative management but the follow-up information has been somewhat limited and follow-up to complete resolution has rarely been reported [1,3,17,19].

Following an initial rapid growth during the first year of life [3,7] MHCW has a self-limited growth pattern. In our case, complete regression was seen after a period of 5 years of radiographic follow-up. This should help reassure those choosing non-operative management of this lesion.

To our knowledge this is the second reported case with complete spontaneous resolution of the lesion with nonsurgical management.

* Corresponding author.
E-mail address: mila.kolar@queensu.ca (M. Kolar).

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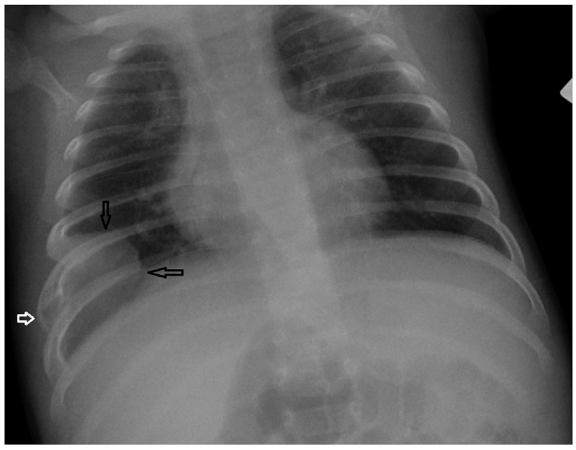

Fig. 1. CXR - Soft tissue density mass projecting over right lower hemithorax (black arrows). Bony inhomogeneity of anterior right 7th rib with lytic appearing regions and adjacent subtle pleural thickening (white arrow).

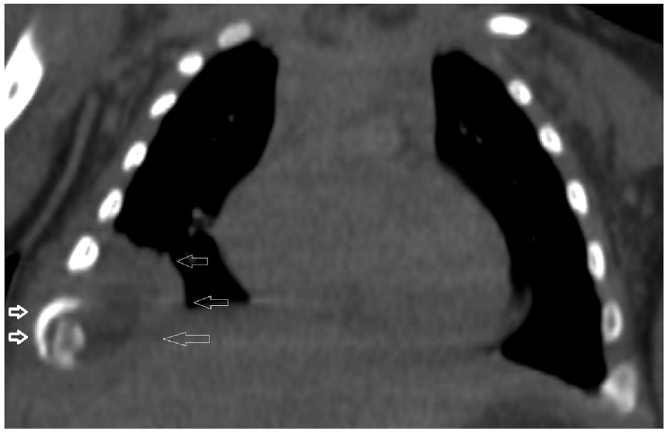

Fig. 2. Coronal CT image demonstrates cortical expansion and interruption of rib (thick arrows). Adjacent inhomogeneous mass with more central hypoattenuating component (thin arrows).

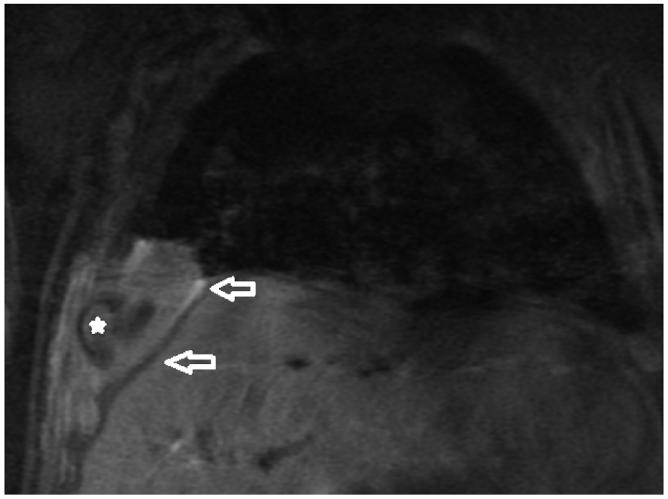

Fig. 3. Coronal T1 post-Gd demonstrates an inhomogeneous mass involving anterior right 7th rib (arrows). Rib is expanded with abnormal marrow signal and cortical interruption medially (star).

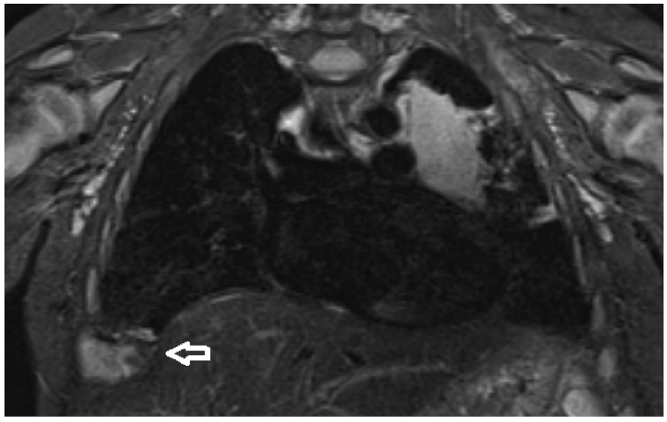

Fig. 4. Follow-up coronal post-Gd MR performed 1 year later demonstrates marked decrease in size and inhomogeneity of the chest wall mass (arrow) bulging into hemithorax and resolution of high signal/enhancement in adjacent musculature.

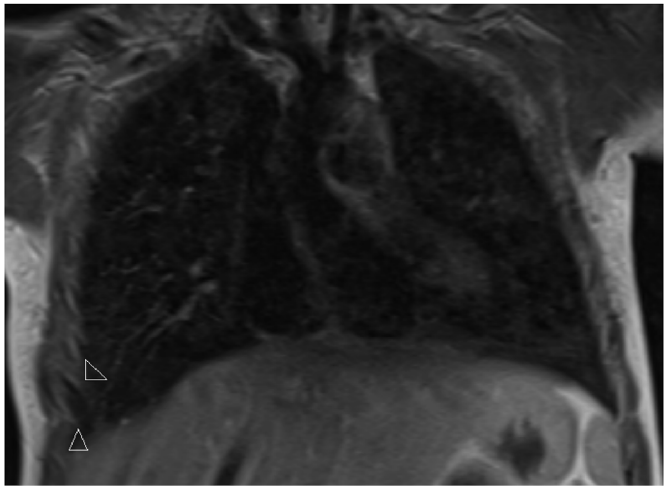

Fig. 5. Coronal T1 post Gd image obtained 5 years after initial presentation demonstrates resolution of the previous chest wall/pleura mass. Residual cortical thickening is evident at anterolateral right seventh rib (arrows).

2. Case report

The patient first presented to pediatric surgical attention at the age of two months after arriving in the emergency department following an episode of respiratory distress due to viral upper respiratory tract infection (URTI). He was an ex-premature fraternal twin born at 32 weeks and had been discharged from hospital following an uneventful neonatal intensive care unit stay.

In addition to evidence of an URTI, he was noted to have a mild deformity of the right lower anterolateral chest wall, with no other abnormal clinical findings, aside from nasal congestion. Laboratory investigations were remarkable for anemia of prematurity (Hgb 79 g/L) and decreased WBC (2.6×10^9/L). Echocardiogram showed a small PDA with no other cardiac abnormalities. Septic work up was negative.

A chest radiograph was obtained, revealing a rounded opacity of the right lower anterolateral chest wall involving the 7th and 8th ribs (Fig. 1). This prompted further radiologic CT evaluation that showed mixed cystic and solid mass (3.4cm × 4cm × 2.8 cm) adjacent to the costochondral junction of the right 7th rib (Fig. 2). The lung and mediastinum were normal in appearance. CT-guided biopsy of the lesion was performed, with histopathologic findings consistent with a mesenchymal hamartoma of the chest wall.

Following resolution of the viral upper respiratory tract infection