**Signature:** © Pol J Radiol, 2015; 80: 283-285
**DOI:** 10.12659/PJR.894124

Polish Journal of Radiology
www.PolRadiol.com
CASE REPORT

**Received:** 2015.03.16
**Accepted:** 2015.04.06
**Published:** 2015.05.28

**Authors' Contribution:**
A Study Design
B Data Collection
C Statistical Analysis
D Data Interpretation
E Manuscript Preparation
F Literature Search
G Funds Collection

# Bilateral Multifocal Hamartoma of the Chest Wall in an Infant

**Erdem Yilmaz[1 ABDEF], Oguz Bulent Erol[2 AB], Melih Pekcan[2 BC], Gokcen Gundogdu[3 BDE], Bilge Bilgic[3 BD], Feryal Gun[4 BC], Ensar Yekeler[5 ABD]**

[1] Department of Radiology, Trakya University, Edirne, Turkey
[2] Department of Radiology, Istanbul Faculty of Medicine, Istanbul, Turkey
[3] Department of Pathology, Istanbul Faculty of Medicine, Istanbul, Turkey
[4] Department of Pediatric Surgery, Istanbul Faculty of Medicine, Istanbul, Turkey
[5] Department of Pediatric Radiology, Istanbul Faculty of Medicine, Istanbul, Turkey

**Author's address:** Erdem Yilmaz, Department of Radiology, Radiology, Trakya University, Edirne, Turkey, e-mail: yilmazerdem79@yahoo.com.tr

## Summary

**Background:** Hamartoma of the thoracic wall is a rare benign tumor that occurs in infancy and can be mistaken for a malignancy due to its clinical and imaging features. Hamartomas are extrapleural soft tissue lesions that cause rib expansion and destruction and appear on imaging as cystic areas with fluid levels and calcification. They can cause scoliosis, pressure on the neighboring lung parenchyma and mediastinal displacement. While conservative treatment is recommended in asymptomatic cases, growing lesions require surgical excision.

**Case Report:** In this report, we present the imaging findings in a 3-month-old infant that presented with a firm swelling in the chest wall and was histopathologically confirmed to have a bilateral multifocal hamartoma.

**Conclusions:** Radiological imaging methods are important for accurate diagnosis of this very rare condition that can be confused with a malignancy.

**MeSH Keywords:** Diagnostic Imaging • Hamartoma • Thoracic Wall

**PDF file:** http://www.polradiol.com/abstract/index/idArt/894124

## Background

Hamartoma of the chest wall is a very rare benign tumor generally seen in early infancy and childhood [1]. Hamartomas originate in the ribs and are histologically characterized by the presence of cartilage tissue and aneurysmal cystic structures [2]. They vary in size and can grow large enough to cause respiratory distress. The clinical features and aggressive imaging appearance of these lesions can lead to a misdiagnosis of malignancy [3]. Hamartomas are generally solitary, although rare cases of bilateral lesions have been reported. In this report, we present the radiography and computed tomography (CT) imaging findings in a case of bilateral multifocal chest wall hamartoma.

## Case Report

A 3-month-old male patient was admitted to the pediatric surgery department with complaints of a palpable, firm swelling in the right chest wall near the scapula. There were no respiratory problems or history of trauma. The parents were first-degree relatives. According to patient history, the lump had not been detected at birth but was noticed several weeks later, and had slowly been growing since.

The patient was in a good general condition. Physical examination revealed an immobile, painless, firm palpable mass measuring roughly 3×2 cm on the right midaxillary line, protruding over the intercostal space. Cardiac, respiratory and abdominal exams were normal. Chest radiograph showed multifocal opacities in the mid-upper and

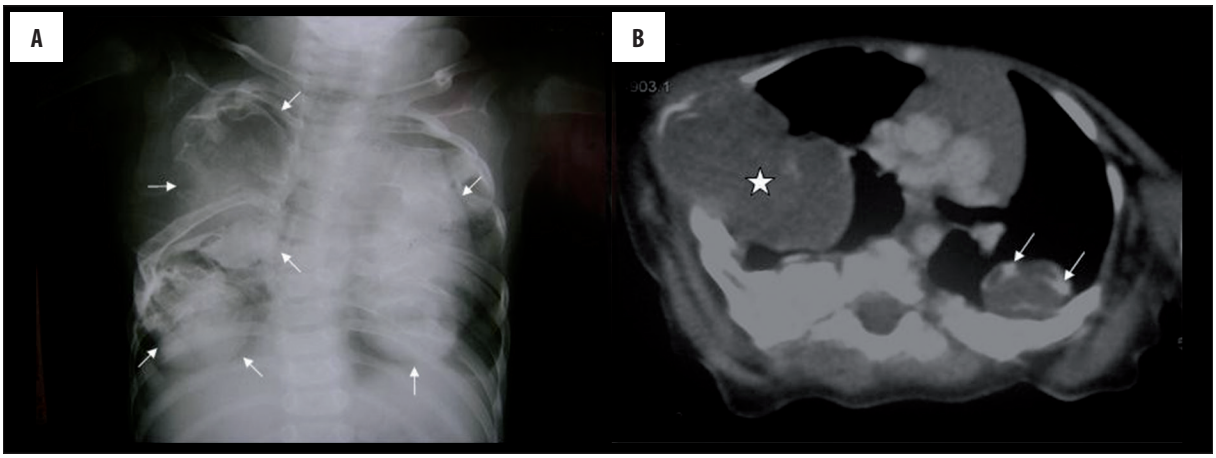


**Figure 1.** On chest radiograph, masses (arrows) in the chest wall (more prominent on the right) causing extensive destruction, deformation and splaying of the ribs, and mild levoscoliosis (**A**). On contrast-enhanced thoracic CT, intrathoracic extension of masses originating from the right 3rd–4th ribs and the left 3rd–4th ribs, more prominent on the right (asterisk). Calcifications can be seen inside the hamartoma of the left thoracic wall (arrows) (**B**).

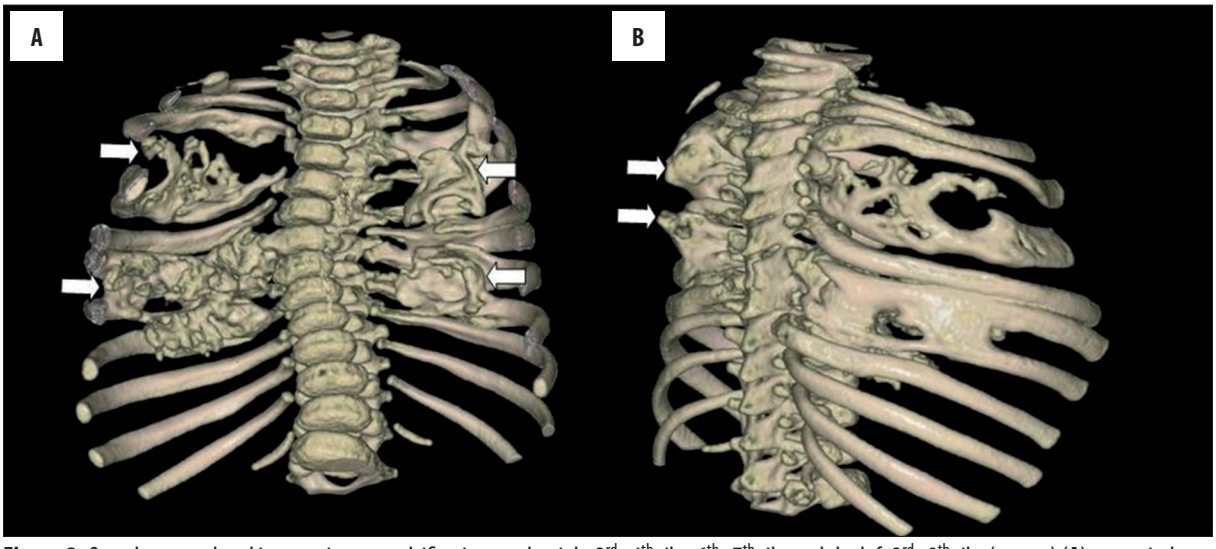


**Figure 2.** On volume rendered images, intense calcifications at the right 3rd–4th ribs, 6th–7th ribs and the left 3rd–8th ribs (arrows) (**A**), posteriorly expanded and deformed left 3rd, 4th and 6th ribs (arrows) (**B**).

lower zone of the right hemithorax and extending from top to bottom of the left hemithorax. Extensive bilateral rib destruction, splaying and deformation were observed in addition to levoscoliosis (Figure 1A).

Contrast-enhanced thoracic CT performed for differential diagnosis revealed solid mass lesions measuring 3×3 cm in the right hemithorax at the 3rd–4th ribs and 2×2 cm at the 6th–7th ribs, and 5×3 cm in the left hemithorax at the 3rd–8th ribs, with intense calcifications and marked intrathoracic extension (Figure 1B). Volume rendered images clearly demonstrated the destruction, expansion, and calcified components in the affected ribs as well as splaying of the bilateral 3rd–8th ribs (Figure 2A). The left 3rd, 4th and 6th ribs appeared posteriorly expanded and deformed (Figure 2B).

Open biopsy was performed on the palpable mass at the right 3rd rib. Histopathological analysis showed aneurysmal cystic walls, scattered osteoclast-like giant cells and reactive bone and cartilage tissue, leading to a diagnosis of thoracic wall hamartoma (Figure 3A, 3B). Gradual partial excision was planned due to extensive intrathoracic involvement. After thoracotomy, the two masses in the right chest wall and the mass in the left chest wall were partially excised. The postoperative condition of the patient was good and he was discharged without complications. No recurrences were seen during a 2-year follow-up period.

## Discussion

Hamartoma of the thoracic wall is a rare benign tumor that generally occurs early in life [1]. Accurate diagnosis of hamartomas is important as most masses detected in the chest wall in infancy and childhood are malignant [2]. Differential diagnosis of thoracic wall hamartoma includes numerous different types of benign and malignant lesions of the thoracic wall [4,5].

Over a hundred cases of chest wall hamartomas have been described in the literature [6], most of them unilateral and

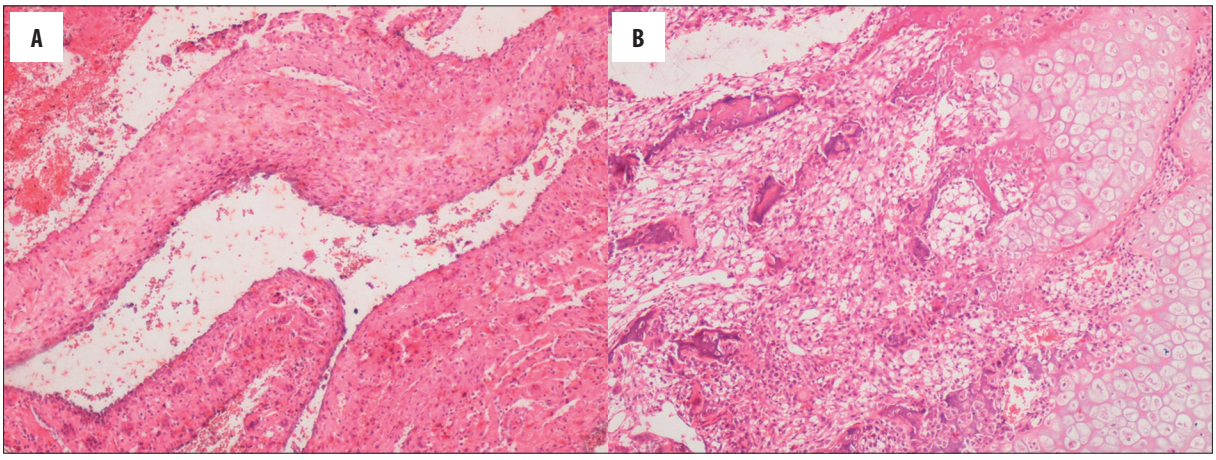


**Figure 3.** Histopathological findings of chest wall mass biopsy. Aneurysmal cystic walls and scattered osteoclast-like giant cells (**A**). Reactive bone and cartilage tissue (hematoxylin and eosin stain, original magnification ×40) (**B**).

solitary [2,7]. To the best of our knowledge, 7 bilateral and 4 multiple unilateral cases have been reported. A bilateral multifocal hamartoma, as in our case, is evidently a very rare occurrence [5,7].

In diagnosing thoracic wall hamartoma, CT is helpful in addition to chest radiography as it clearly shows the origin of the tumor, its density, size, and relationship with neighboring tissues [7]. Hamartomas are large extrapleural soft tissue lesions that lead to rib expansion and destruction, and appear on imaging as cystic areas of calcification and fluid levels [2]. Secondary findings include scoliosis, pressure on the neighboring lung parenchyma, and mediastinal displacement [5]. While hamartomas of the chest wall have a characteristic radiological appearance, multiple lesions may be mistaken for multifocal metastatic diseases like lymphoma and leukemia [2], necessitating histopathological verification. In our case, direct radiography and CT imaging findings were verified with a biopsy.

Macroscopical appearance of a chest wall hamartoma is generally that of solid islands and cystic spaces. Microscopically there are cartilage islands, immature mesenchymal elements, aneurysmal bone cyst areas, and possibly also prominent new bone formations. Although these lesions lack nuclear atypia and hyperchromasia, they can be mistaken for malignancies when hypercellular [5,7].

Chest wall hamartomas are treated conservatively or surgically in case of pressure on respiratory passages or lungs. The en bloc excision technique is used to remove the involved ribs, pleura, muscles, and neurovascular structures. Although this method is curative, the large remaining thoracic defect often leads to scoliosis [1]. The minimally-invasive radiofrequency thermoablation technique can be used in case of recurrence after surgery, particularly when lesions are difficult to access [8]. Conservative treatment is preferred in asymptomatic and self-limiting cases [4,6]. Due to significant pressure of bilateral multifocal lesions on the lung parenchyma in our patient, partial resection of intrathoracic components was performed. We decided against total excision to avoid scoliosis complications as the lesions were spread over a large area.

## Conclusions

Bilateral multifocal hamartoma of the thoracic wall is a very rare condition that has only been reported a handful of times. Knowing the specific imaging features of hamartoma in infants is helpful in accurate diagnosis, appropriate management and planning of surgical treatment.

### Conflict of interest disclosure

The authors declare no conflict of interest

## References:

1. Dounies R, Chwals WJ, Lally KP et al: Hamartomas of the chest wall in infants. Ann Thorac Surg, 1994; 57: 868–75
2. Groom KR, Murphey MD, Howard LM et al: Mesenchymal hamartoma of the chest wall: radiologic manifestations with emphasis on cross-sectional imaging and histopathologic comparison. Radiology, 2002; 222: 205–9
3. Pawel BR, Crombleholme TM: Mesenchymal hamartoma of the chest wall. Pediatr Surg Int, 2006; 22: 398–400
4. Shimotake T, Fumino S, Aoi S: Respiratory insufficiency in a newborn with mesenchymal hamartoma of the chest wall occupying the thoracic cavity. J Pediatr Surg, 2005; 40: 13–16
5. Sodhi KS, Aiyappan SK, Menon P et al: Unilateral multifocal mesenchymal hamartoma of the chest wall: a case report and review of literature. J Pediatr Surg, 2009; 44: 464–67
6. Braatz B, Evans R, Kelman A, Cheng W: Perinatal evolution of mesenchymal hamartoma of the chest wall. J Pediatr Surg, 2010; 45: 37–40
7. Altaner S, Yoruk Y, Bilgi S et al: Multifocal mesenchymal hamartoma of the chest wall. Respirology, 2006; 11: 334–38
8. Bertocchini A, Falappa P, Accinni A et al: Radiofrequency Thermoablation in chest wall mesenchymal hamartoma of an infant. Ann Thorac Surg, 2007; 84: 2091–93