Mesenchymal Hamartomas of the Chest Wall in Infancy: Radiologic and Pathologic Correlation

Ji-Young Kim¹, Woo-Hee Jung¹, Choon-Sik Yoon², Myung-Joon Kim², Hae-Kyoon Kim³, Kil-Dong Kim³, and Sang-Ho Cho¹

Abstract

Mesenchymal hamartoma of the chest wall is a rare tumor with about 53 reported cases in the English literature. We reviewed six chest wall mesenchymal hamartomas in four patients, including two cases with multiple lesions, with specific focus on the radiologic and pathologic correlation. All cases occurred in neonates or infants with ages ranging from seven hours to seven months. They were diagnosed with plain chest radiographs (n=6), ultrasonography (n=2), chest CT scan (n=6), whole body bone scan (n=2) and MRI (n=3). All cases except a small one without cystic change showed the typical features of mesenchymal hamartoma radiographically and pathologically. Radiologically they were well-circumscribed masses with solid and cystic components with multiple fluid-fluid levels in association with single or multiple rib destruction or change. The CT scan showed the typical findings of chest wall hamartoma, and the MR showed heterogeneous signal intensities of the mass on T1- and T2-weighted images. The MR also revealed more concisely a secondary aneurysmal bone cyst formation with multiple fluid-fluid levels on the T2-weighted image. Microscopically, they showed alternating areas of cartilaginous islands and primitive appearing mesenchymal proliferation, which corresponded well with the solid component on the radiologic findings. The areas of bone formation and blood-filled cystic spaces matched the calcified or ossified densities and the cystic components, respectively. A small case without cystic change showed peculiar radiological and pathological findings resembling an osteochondroma. In conclusion, mesenchymal hamartoma of the chest wall in infancy is quite rare and sometimes can be misdiagnosed as malignancy due to the bone-destroying radiographic appearance and the highly cellular and mitotically active microscopic features, unless the radiologists and pathologists are aware of the characteristic clinical, radiological, and pathological findings. Imaging studies can usually make a correct diagnosis with good correlation to the pathologic findings.

Key Words: Mesenchymal hamartoma, cartilaginous hamartoma, rib, chest wall, infant, CT, MRI

INTRODUCTION

Mesenchymal hamartoma of the chest wall is a rare tumor with about 53 cases having been reported in the English literature. It typically presents as a solitary chest wall mass in neonate or in early infancy. Multiple lesions are extremely rare with only five reported cases so far. There have been only a few reports of CT or MR findings in chest wall hamartoma.²⁵ We reviewed the CT and MR findings of six mesenchymal hamartomas of the rib and correlated them with pathologic findings.

MATERIALS AND METHODS

From 1990 to 1999, we experienced six mesenchymal hamartomas of the chest wall in four infant patients, including the one case previously reported by us.¹ Three were males and one was female. The age range was from seven hours to seven months (mean age: four months). The clinical presentations were cyanosis and respiratory distress in the seven-hour-old newborn, and nonspecific (cough and fever) in three patients who had incidentally presented on a plain chest radiograph. Plain chest radiographs (n=6), chest CT scan (n=6), whole body bone scan (n=4), ultrasonography (n=2), and MRI (n=3) were performed on all six chest wall mesenchymal hamartomas. Gross and microscopic pathologic findings

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Departments of ¹Pathology, ²Radiology and ³Chest Surgery, Yonsei University College of Medicine, Seoul, Korea.
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Address reprint requests to Dr. W. H. Jung, Department of Pathology, Yonsei University College of Medicine, C.P.O Box 8044, Seoul 120-752, Korea. Tel: 82-2-3497-3541, 3540, Fax: 82-2-3463-2103, E-mail: jungwh96@yumc.yonsei.ac.kr
were also reviewed and examined in relation with the radiologic features.

RESULTS

We have summarized the cases of chest wall hamartoma in Table 1.

Table 1. The Summary of Cases of Chest Wall Hamartoma in Infancy

<table>
<thead>
<tr>
<th>Case No.</th>
<th>1-1</th>
<th>1-2</th>
<th>2</th>
<th>3</th>
<th>4-1</th>
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<td>m/4 mo</td>
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<td>single</td>
<td>two</td>
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<td>Sex/Age</td>
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<td>incidental</td>
<td>left, 3rd &amp; 4th ribs</td>
<td>incidental</td>
<td>right, 4th &amp; 5th ribs</td>
<td>left, 10th rib</td>
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<td></td>
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<tr>
<td>Location</td>
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</tr>
<tr>
<td>Size (cm)</td>
<td>4×5×7</td>
<td>2×3×3</td>
<td>5×6</td>
<td>5×6</td>
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<td>+</td>
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<td>+</td>
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<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
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<td>+</td>
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<td>+</td>
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<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Invasion</td>
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<tr>
<td>MRI</td>
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<td></td>
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<tr>
<td>Solid</td>
<td>T1 iso, T2 iso</td>
<td>Gd-T1 (+)</td>
<td>T1 iso, T2 iso, Gd-T1FS (+)</td>
<td>T1 iso, T2 iso</td>
<td>Gd-T1FS (+)</td>
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<tr>
<td>Cystic</td>
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<td>Gd-T1 (-)</td>
<td>T1 high, T2 high</td>
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<td>uptake †</td>
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<td>+</td>
<td>+</td>
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<td>+</td>
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<td>Ossification</td>
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<tr>
<td>Woven bone</td>
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<td>+</td>
<td>+</td>
<td>+</td>
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<td>+</td>
</tr>
<tr>
<td>Enchondral</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
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</tr>
<tr>
<td>ABC formation</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
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</tbody>
</table>

Radiologic findings

Three chest wall hamartomas among the six lesions showed characteristic findings in plain chest radiographs. These findings were considerably large intrathoracic soft tissue masses with multiple calcification and ossification, and single or adjacent multiple rib changes with destruction, splaying, thinning or erosion (Fig. 1). One smaller lesion (case 1-2), located

T1, T1-weighted image; T2, T2-weighted image; Gd-T1, Gadolinium-enhanced T1-weighted image; Gd-T1FS, Gadolinium-enhanced T1-weighted fat saturation image; Pri. msnch prol., primitive mesenchymal proliferation; Woven bone, woven bone formation; Enchondral, enchondral ossification.
in the left retrocardiac area, could not be detected due to cardiac shadow on the plain radiograph.

Whole body bone scans were performed in two patients (cases 1 and 4) and showed increased radioisotope uptake. Ultrasonography in two lesions (cases 3 and 4-①) showed heterogeneous mixed echoic masses with solid and cystic portions, as well as highly echogenic areas with posterior shadowing suggesting calcification or ossification (Fig. 2). Characteristically, the chest CT scan in five of six lesions showed intrathoracic masses with solid and cystic components, with single or multiple rib involvement and with multiple ossification or calcification (Fig. 3 and 4). In one small lesion involving a posterior rib (case 4-②), ossified bony septae and rims were noted with small multiple low attenuation areas on CT scan.

MRI in two of three lesions showed typical findings of solid and cystic components with multiple fluid-fluid levels (Fig. 5). The cystic components presented high signal intensities on the T1-weighted image and intermediate signal intensities with multiple fluid-fluid levels on the T2-weighted images. Solid components alternating with the cystic areas demonstrated intermediate and low signal intensities. In the one small lesion (case 4-②), the T1- and T2-weighted MR images showed homogeneous low
signal intensity without cystic components (Fig. 6).

Pathologic findings and radiologic-pathologic correlation

The pathologic findings of the tumors were similar to one another. They were large, expansile smooth surfaced, slightly lobulated pinkish masses arising from one or a few ribs partly destroying the adjacent ribs. Cut sections revealed scattered large nodules of whitish glistening cartilaginous tissue alternating with soft tan granular or firm tissue. Cysts filled with dark red brown blood were also present. The calcified foci were gritty (Fig. 7). Microscopic examination showed
the typical features of mesenchymal hamartomas of rib in infancy. All lesions except the smallest one (case 4-2) had sheets of primitive appearing mesenchymal stroma with hypercellularity and frequent mitotic figures regionally (Fig. 8). Frank anaplasia or atypical mitoses was not seen.

Cartilaginous islands were present in and among the primitive stroma and appeared to be derived from it (Fig. 9). The spindle cells surrounding the cartilaginous islands had more plump eosinophilic cytoplasm and more round nuclei resembling the chondroblasts. Some of the cartilage islands reveal foci of enchondral ossification (Fig. 10). In addition to the cartilaginous islands, there were areas of woven bone production (Fig. 11A) that seemed to be derived from some of the proliferating primitive mesenchymal stroma. Abundant thick hyalinized eosinophilic osteoid-like material appeared to be surrounded by the stromal cells and merged to the more organized bony trabeculae (Fig. 11B). In the vicinity of the stromal cells, there were many osteoclast-like giant cells that were especially frequent around the blood-filled cystic spaces resembling the aneurysmal bone cysts. This was a common feature in all four patients to varying
degrees (Fig. 12).

In the two cases with multiple lesions, case 1 and case 1-2 showed similar microscopic features. In case 4, the smaller mass (case 4-2) was quite different from the larger one (case 4-1). It almost looked like an osteochondroma since it was a protruding bony mass, continuous with the normal marrow cavity and having a cartilaginous cap and a core consisting of normal-looking bony marrow (Fig. 13). However, there was a small concentration of primitive appearing mesenchymal cell proliferation at the initial section, and with multiple deeper sections, a small cartilaginous island finally appeared in the center of the primitive mesenchymal cell area (Fig. 14). Woven bone areas, blood-filled cystic spaces, or osteoclast-like giant cells was not noted.

Fig. 12. The osteoclast-like giant cells are especially frequent around the blood-filled cystic spaces (H & E, ×100).

Fig. 13. The smaller mass (case 4-2) is very similar to an osteochondroma with a cartilaginous cap and a core that consists of normal-looking bony marrow.

Fig. 14. The smaller mass (case 4-2) reveals a small immature cartilaginous island in the center of the primitive appearing mesenchymal cell area. The surrounding bony marrow is essentially normal (H & E, ×40).

In case 3, a substantial amount of brown fat tissue was present at the interface of the tumor and the non-tumorous soft tissue adjacent to the destruction of bony cortex.

These typical features of mesenchymal hamartoma correlated well with the radiologic findings. The solid components with intermediate or low signal intensities on MR corresponded to the areas of primitive mesenchymal cell proliferation and chondroid islands. The calcified or ossified areas were more readily manifested on the CT or plain chest radiographs. The cystic components manifested as multiple fluid-fluid levels on MR also corresponded to the blood-filled cystic spaces resembling aneurysmal bone cysts (Fig. 7). The smaller one without a cystic component (case 4-2) presented only as homogeneous low signal intensities on the MR scan, and the pathologic examination revealed an osteochondroma-like solid protruding bony mass. The focus of the primitive mesenchymal cell proliferation and cartilaginous island was only microscopic in dimension (Fig. 6, 13, and 14).

DISCUSSION

Mesenchymal hamartoma of the chest wall in infancy, also called cartilaginous hamartoma or mesenchymoma, is quite rare. Although it is still unclear whether it is a true neoplasm or not, the idea that it is a local disordered overgrowth of normal
skeletal component is more widely accepted at present. Since Blumenthal et al. described two typical cases in 1972 under the name of intrathoracic mesenchymoma, about 53 cases have been reported so far in the English literature.\textsuperscript{1,7} Mesenchymal hamartoma typically presents as a solitary well-circumscribed mass arising from the rib. Usually it is composed of a mixture of immature spindle cells, islands of cartilage, and areas of woven bone formation. Osteoclastic type giant cells and areas of enchondral ossification are often present. As areas of immature mesenchymal cells are often highly cellular and sometimes show frequent mitoses, it was often misdiagnosed as malignant.\textsuperscript{5,7} Sometimes it has prominent cystically dilated vascular channels resembling an aneurysmal bone cyst.\textsuperscript{8,9} Multiple lesions are extremely rare with only five reported cases so far.\textsuperscript{1-5} Four of these lesions were ipsilateral,\textsuperscript{1,4} and the one reported under the name of osteochondroma was bilateral.\textsuperscript{5} The two presenting cases with multiple lesions were also ipsilateral. In case 1, the two masses were quite similar to each other histologically, but in case 4, the smaller mass was quite different from the larger one. It was a protruding bony mass with a cartilaginous cap and the core composed mostly of normal-looking bone marrow with hematopoietic tissue. An important clue for the diagnosis was a small concentration of primitive appearing mesenchymal cell proliferation in the midst of otherwise normal bony marrow, and multiple deeper sections revealed a small island of immature cartilage in the center of the mesenchymal cell area. In addition, the cartilaginous tissue in the cap was slightly lobulated and looked immature, differing from the regular, organized, and epiphyseal plate-looking cartilaginous cap of an osteochondroma.

Microscopically, all cases had more or less characteristic findings of mesenchymal hamartoma. Primitive appearing mesenchymal stroma may be mitotically active regionally, which may cause a misdiagnosis as malignancy. Case 2 had very frequent mitoses with about 6 to 7 mitoses/10 HPF. However, atypical mitoses or frank anaplasia were not present. The other cases (cases 1, 3, and 4) had low mitotic counts.

In case 3, a substantial amount of brown fat was present at the interface of the tumor and non-tumorous soft tissue adjacent to the destruction of the bony cortex. The tumorous part and the brown fat were continuous with a thick band of fibrosis that was replacing the destroyed bony cortex. Two previous cases with mature adipose tissue have been reported,\textsuperscript{12} in which mature adipose tissue was intimately mixed with primitive spindle cell areas, and these cases were initially diagnosed as infantile fibrous hamartomas. The brown fat in case 3 may be the integral component of the tumor, but the significance is questioned by its location at the interface between the tumor and surrounding soft tissue. Since brown fat may normally be found around the ribs in infants, it is more likely that the brown fat was entrapped during tumor growth.

There have been only a few reports about CT and MR findings of chest wall hamartoma.\textsuperscript{15,18} Brand et al. stated that the CT scans revealed a solid mass in two infants and a solid and cystic mass in another patient without presenting CT images.\textsuperscript{15} Schlesinger et al. reported that the CT demonstrated cortical bone surrounding the mass, and suggested an osseous origin of the lesion. They also determined two distinct components on the T1-weighted image of the MR findings: one with a high signal intensity (blood-filled cysts) and the other with a low signal intensity (chondroid elements), which provided information concerning the internal matrix of the lesion.\textsuperscript{18} The demonstration of two distinct components with different signal intensities on the MR can help differentiate this lesion from an aneurysmal bone cyst, and suggests the diagnosis of a mesenchymal hamartoma of the chest wall.\textsuperscript{18}

As in our cases, the CT scan can showed cortical bone partially surrounding the mass with a portion of fluid-fluid level and the destruction of single or multiple ribs, which are characteristic of chest wall hamartoma. The MR can easily show a portion of multiple fluid-fluid levels and blood-filled cystic spaces, but it cannot be demonstrated correctly in a small lesion without aneurysmal bone cyst formation.

Although there have been cases showing regrowth after limited surgery,\textsuperscript{12,13} including the one finally diagnosed as a sarcoma,\textsuperscript{12} mesenchymal hamartoma in infancy is believed to be essentially benign and the cure generally consists of wide en bloc resection. All four of the cases presented were well without recurrence or metastasis after a follow-up period of one to ten years. Scoliosis is one of the complications after extensive rib resection,\textsuperscript{13-15} but none of the four presenting cases developed scoliosis.

Mesenchymal hamartoma of the chest wall in
infancy is quite rare and sometimes can be misdiagnosed as malignancy due to the bone-destroying radiographic appearance and the highly cellular and mitotically active microscopic features, unless the radiologists and pathologists are aware of the characteristic clinical, radiological, and pathological findings. Imaging studies can usually make a correct diagnosis with good correlation to the pathologic findings.

REFERENCES