

Malignant Fibrous Histiocytoma of Chest Wall

Jin-Kyung An and Ki Keun Oh

Department of Diagnostic Radiology, Yonsei University, College of Medicine, Research Institute of Radiological Science, Yonsei University, Seoul, Korea.

Primary malignant fibrous histiocytoma (MFH) of the chest wall is rare. We report a case of primary MFH arising from the chest wall, which was thought to be a metastasis or myeloma. The imaging study revealed a single mass of the chest wall involving a rib. Resection and chest wall reconstruction was done. The histologic diagnosis was storiform-pleomorphic primary MFH. Although MFH of the chest wall is an uncommon pathology, it should be considered in the differentiation of a single bony destructive lesion involving the rib with a soft tissue component.

Key Words: Thorax, neoplasm, malignant fibrous histiocytoma, diagnostic imaging.

INTRODUCTION

The most common chest wall mass with rib destruction is metastasis, followed by multiple myeloma.¹ In adults, the ribs are among those bones with remaining red marrow, making them susceptible to hematogenous metastases from cancers of the breast, lung, kidney and thyroid.² MFH is the most common soft tissue sarcoma in adults and generally occurs in the extremities, particularly in the thigh, whereas chest wall involvement is uncommon. Belal et al. retrospectively reviewed 109 cases of MFH and reported that 47% of these tumors arose in the lower limbs, 18% in the upper limbs, 16% in the head and neck, 9% in the trunk, 5% in the pelvis and 5% in other parts of the body.³ One case report reviewed 37

cases of MFH in the chest wall, which had previously been reported in the Japanese and English literature.⁴

This report describes and illustrates the clinical, radiologic and pathologic features of a case of primary MFH of the chest wall affecting a 25-year-old woman. We also discuss the differential diagnosis of chest wall mass with rib destruction.

CASE REPORT

A 25-year-old woman developed intermittent left chest wall pain over a 4 month period. She suffered speaking induced dyspnea. She had no prior history of smoking or asbestos exposure. She had no known underlying systemic disease.

Radiologic studies included a posteroanterior and lateral chest radiograph, ultrasonography, computerized tomography (CT) and whole body bone scan (WBBS). The posteroanterior and lateral chest radiograph revealed a round mass lesion in the left anterior chest (Fig. 1). Sonography showed a well defined, oval shaped, hypoechoic mass involving a rib, which measured 4.2×1.9×4.5 cm (Fig. 2). CT demonstrated the presence of a relatively well defined, ovoid shaped mass at the anterior chest wall. Bony infiltration was suggestive of the tumor having a malignant nature. The mass showed heterogeneous weak enhancement (Fig. 3). The interface between the mass and the lung was smooth, and compression of the lung was observed. There was no mediastinal lymph node enlargement. Neither lung showed evidence of hematogenous or lymphangitic metastasis. The WBBS revealed markedly increased bony uptake with a central photon defect of the left 3rd ante-

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Reprint address: requests to Dr. Ki Keun Oh, Department of Diagnostic Radiology, Research Institute of Radiological Science, Yonsei University College of Medicine, 134 Shinchon-dong, Seodaemun-gu, Seoul 120-752, Korea. Tel: 82-2-3497-3511, Fax: 82-2-3462-5472, E-mail: kbrdoh@yumc.yonsei.ac.kr

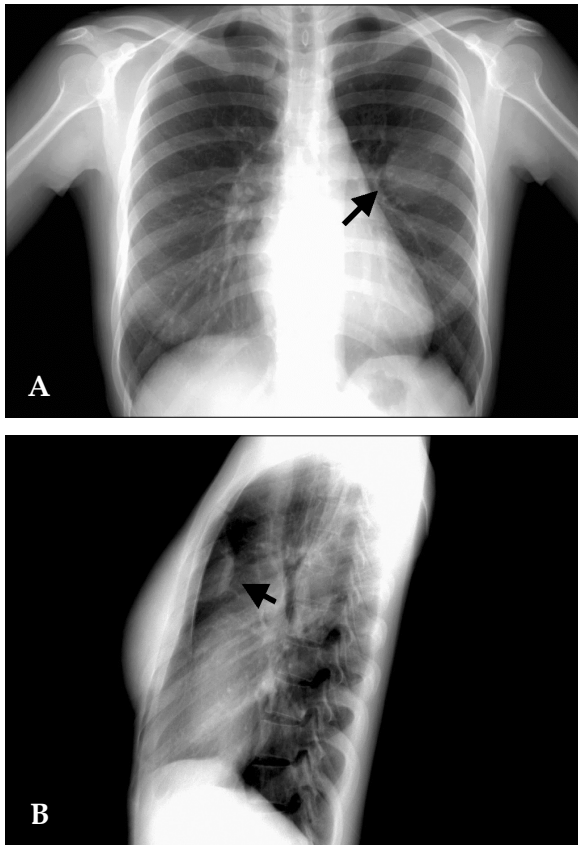


Fig. 1. The posteroanterior (A) and lateral (B) chest radiograph revealed a round mass lesion (arrows) in the left anterior chest.

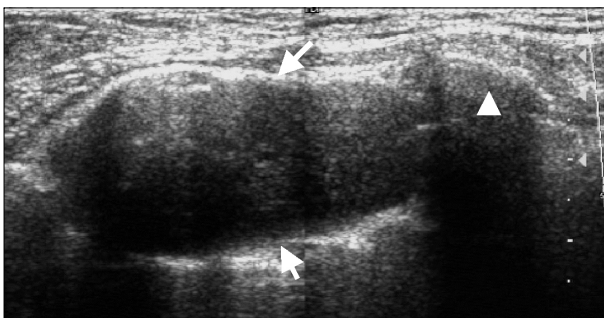


Fig. 2. Sonography showed a well defined, oval shaped, hypoechoic mass (arrows) involving a rib (arrowhead).

rior rib (Fig. 4). No other abnormal, significantly increased bony uptake was noted in the whole skeleton.

Resection of the left 3rd rib, partial resection of the left 2nd and 4th ribs, and chest wall reconstruction with Marlar mesh was done. The gross specimen showed a multilobulated pinkish-white

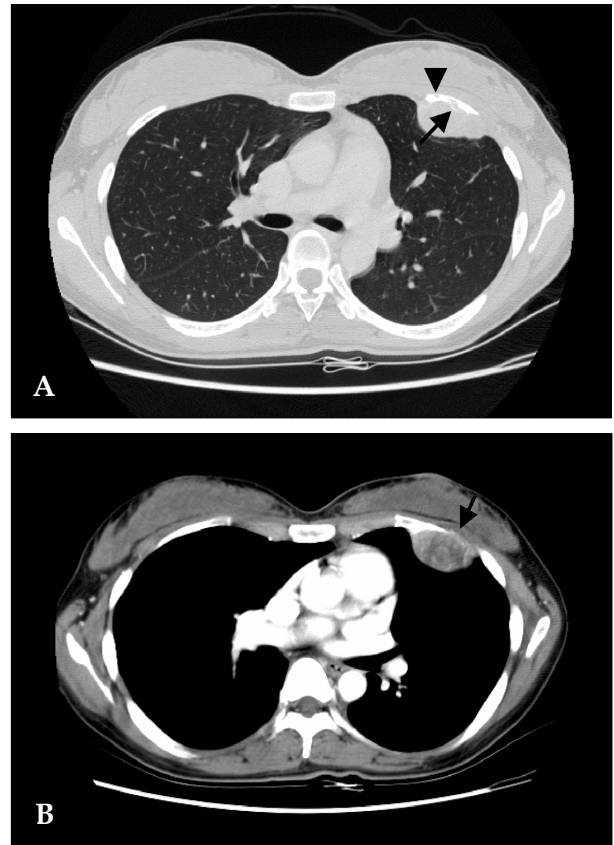


Fig. 3. CT demonstrated the presence of a relatively well defined, ovoid shaped mass at the anterior chest wall. A. Bony infiltration (arrow) and defect (arrowhead) was suggestive of the tumor having a malignant nature. The interface between the mass and the lung was smooth, and compression of the lung was observed. B. The mass showed heterogeneous weak enhancement (arrow). There was no mediastinal lymph node enlargement.

solid mass which adhered to and infiltrated the inner surface of the rib (Fig. 5). One segment of the mass situated within the bony defect (Fig. 6A). Histopathology showed a storiform arrangement of malignant cells, including a plump cytoplasm and a few mitotic figures, which was suggestive of storiform-pleomorphic malignant fibrous histiocytoma (Fig. 6B).

DISCUSSION

MFH is the most common soft tissue tumor in adults (20 - 30%), and is principally located in the extremities or the retroperitoneum.⁵ This tumor preferentially involves the deep fascia, skeletal



Fig. 4. The WBBS revealed markedly increased bony uptake (arrow) of the left 3rd anterior rib without other abnormal uptake on the whole skeletons.

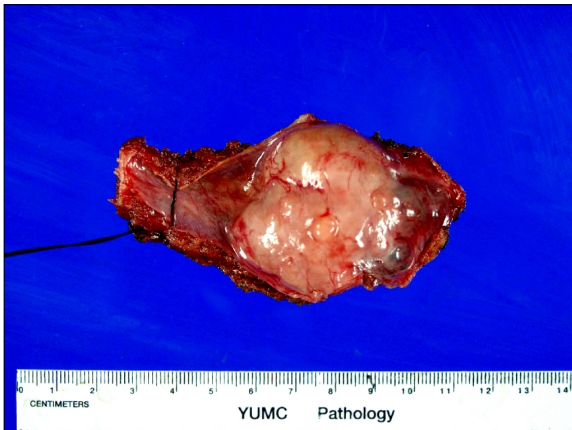


Fig. 5. Gross specimen showed a multilobulated pinkish-white solid mass.

muscle or superficial subcutis.⁶⁻⁸ The peak incidence is in the fifth decade of life. Several subtypes of MFH are described in the literature, namely storiform-pleomorphic, myxoid, giant cell, and inflammatory.

The sonographic findings of chest wall MFH are not well known, and one case reported a well defined inhomogeneous low-echoic mass.⁴ CT and magnetic resonance (MR) imaging are useful for the radiological evaluation of the soft tissue component. CT can provide more accurate detection of cortical bone destruction, whereas MRI displays the infiltration of the bone marrow and extent of the mass with good resolution.⁹ The mass usually shows intense enhancement on CT with a clear margin separating it from the surrounding tis-

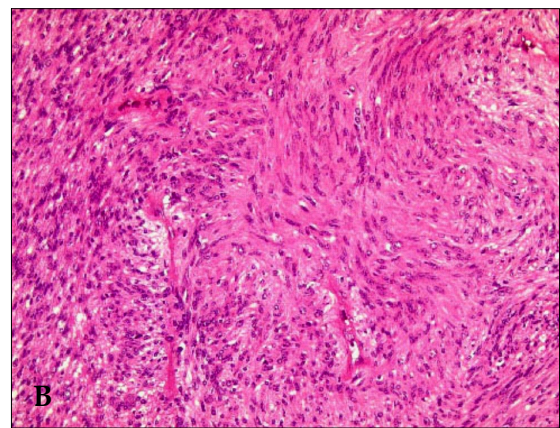
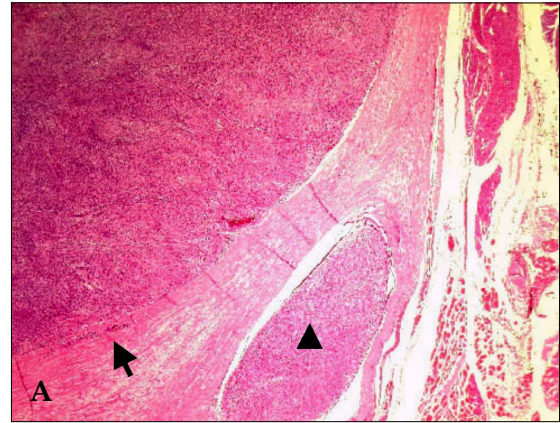


Fig. 6. Microscopic findings. A. The mass adhered to and infiltrated the inner surface of the rib (arrow). One segment of the mass (arrowhead) situated within the bony defect (H-E stain, $\times 40$). B. Storiform arrangement of malignant cells, including a plump cytoplasm and a few mitotic figures, was suggestive of storiform-pleomorphic malignant fibrous histiocytoma (H-E stain, $\times 100$).

sue.¹⁰ Furthermore, the mass often shows decreased central attenuation due to necrosis, hemorrhage and mucoid material.¹¹ On MR imaging, the mass shows an intermediate to low signal intensity on T1WI and high signal intensity on T2WI images, which frequently shows inhomogeneous central intensity. One study reported that 95% of chest wall MFH demonstrated the existence of intercostal muscle invasion on MR imaging.¹⁰

Our case was located in the inner portion of the rib, and was thought to have originated from the periosteum or the adjacent soft tissue component. The oval shaped small mass appeared to be benign, but a point of bony infiltration suggested that the tumor was of a malignant nature. The

mass showed a well defined, oval shaped, hypoechoic mass on the sonogram and heterogeneous weak enhancement on the CT image.

Our differential diagnoses were metastasis and myeloma. Metastasis is the most common chest wall mass involving the ribs and hematogenous metastasis is more common than direct invasion.¹² Multiple myeloma is also one of the most common chest wall masses. Hematogenous metastasis and multiple myeloma involves the red marrow, and therefore the mass is centrally located within the rib. Multiple myeloma is frequently multifocal and solitary rib involvement is uncommon. Our case was an eccentrically located single mass, increasing the probability of its being a primary chest wall mass. A diagnosis of benign neurogenic tumor originating from the intercostal nerve, which would result in rib erosion, notching and sclerosis, was excluded due to the infiltrative nature of the mass.

Wide resection is the first choice treatment and additional chemotherapy may play a role in treating osseous MFH. Good response to preoperative chemotherapy seems to be associated with an excellent prognosis.^{13,14} Postoperative radiation therapy is essential in cases of tumors with positive surgical margins following wide complete gross excision.³ The tumor grade, size and the presence of distant metastases at the initial presentation remain the most important prognostic factors for MFH.^{3,15}

In summary, we reported a case of primary MFH of the chest wall and reviewed the differential diagnosis and management of this uncommon type of tumor.

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