

Radiological Findings of Giant Pulmonary Chondromatous Hamartoma: Case Report¹

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Among lung tumors, multiple hamartomas are uncommon and multiple chondromatous hamartomas are extremely rare. In this report, we describe a patient with an interval growth of a giant chondromatous hamartoma with accompanying multiple satellite nodules in the adjacent lung and the parietal pleura. We also present the follow up chest radiograph, computed tomograph, magnetic resonance images and photographs of the operating field of the tumor with pathologic correlation.

Index words : Lung tumor
Multiple tumors
Hamartomas
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Hamartomas, which are classified as either chondromatous or leiomyomatous, are the most common benign tumors of the lung. However, multiple pulmonary chondromatous hamartomas are rare with less than 20 cases having been previously reported (1). These lesions have also been known to coexist with certain other diseases. The majority of such tumors are small and solitary (2). We present a case of a giant pulmonary chondromatous hamartoma with multiple satellite nodules in the adjacent lung and parietal pleura, which was followed for three years since it was originally discovered.

Case Report

A 40-year-old woman presented with progressive dys-

pnea for one month. Three years before a pleural-based mass in the patient's left lung was discovered incidentally, but the patient ignored the recommendation for further evaluation of the lesion (Fig. 1A). Follow-up chest radiographs showed interval growth of the mass that occupied the left hemi-thorax from the diaphragm to the aortic arch and a newly developed pleural effusion (Fig. 1B).

Contrast-enhanced CT (5-mm collimation) revealed a huge mass with peripherally thick linear calcifications, soft tissue densities, and ill-defined fat densities (HU: -40 - -50) (Fig. 1C). A small amount of pleural effusion and an adjacent atelectasis of the left lower lobe were combined.

MR imaging was performed to evaluate the resectability of the mass. A pre-enhanced axial T1-weighted image (TR/TE 158/6.2) showed well-defined heterogeneous intermediate signal intensity (higher than that of skeletal muscle but lower than that of fat) of the mass without typical signal intensity representing the fat and the calcification (Fig. 1D). However, the calcification showed various signal intensities on MR images and a partial averaging effect could not be excluded. A contrast-en-

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hanced sagittal T1-weighted image (TR/TE 110/6.2) showed strong enhancement of the periphery and septa of the mass (Fig. 1E). A T2 (haste tra fast)-weighted coronal image (TR/TE 1030/64) showed heterogeneous intermediate to high signal intensity (Fig. 1F). The periphery and septa of the mass showed high signal intensity on a T1-weighted image and low signal intensity on a T2-

weighted image.

A tumor resection was performed and the resected specimen revealed pulmonary chondromatous hamartoma, reactive pleuritis with cholesterol granulomas of the visceral pleura, and a hemorrhagic infarction of the adjacent lung.

During the operation, approximately ten satellite

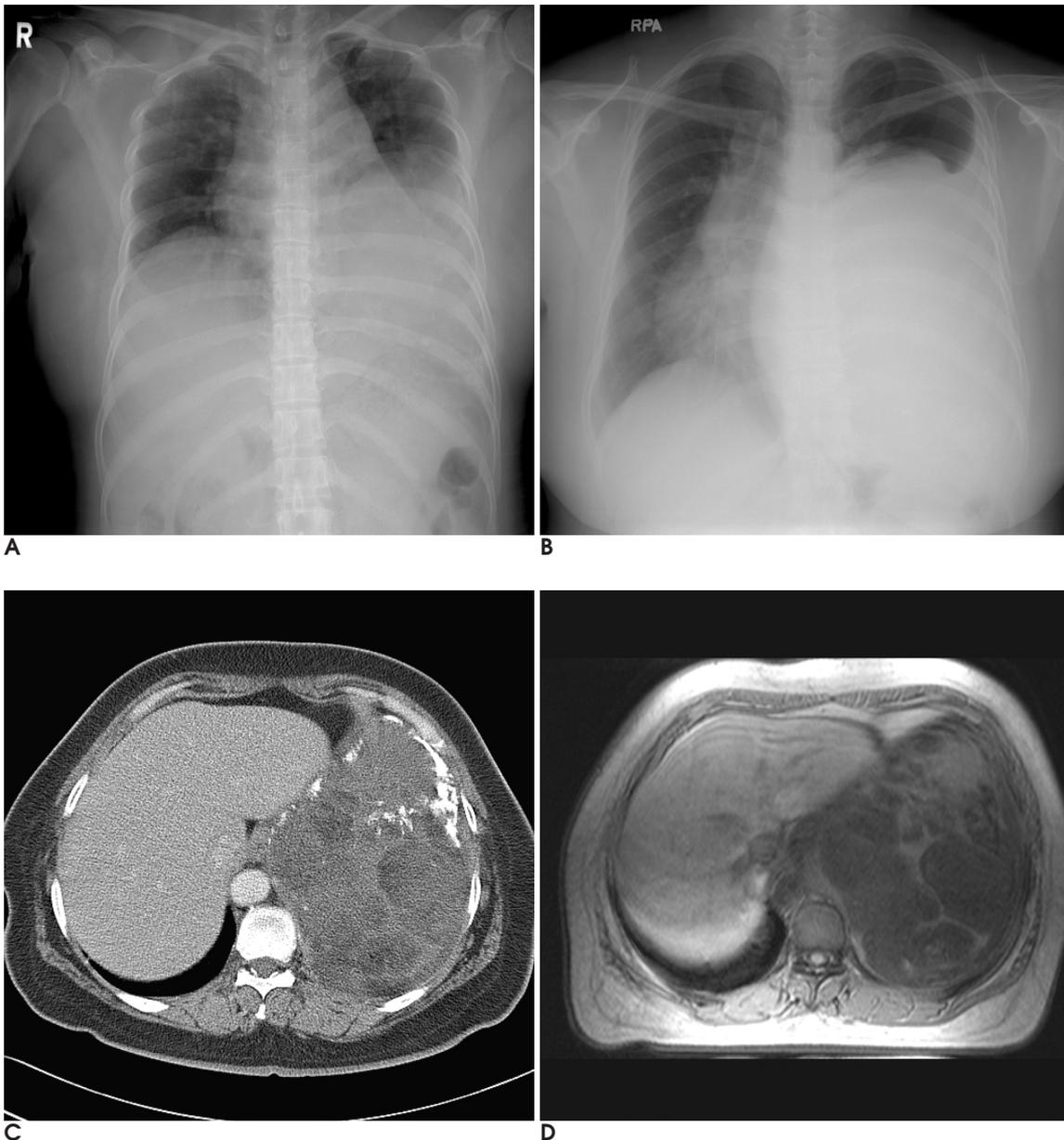


Fig. 1. A 40-year-old woman with progressive dyspnea for 1 month.

A. AP view of the chest radiograph that was performed three years before shows an ill-defined pleural-based mass with some calcifications.

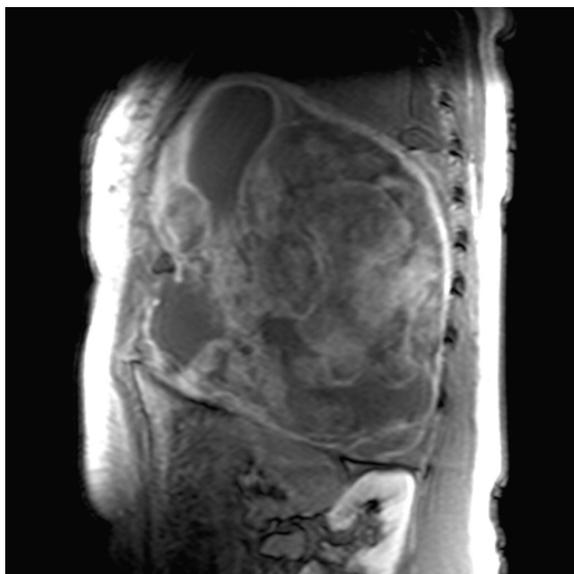
B. Posteroanterior chest radiograph reveals an interval increase in the size of the mass that occupies the left hemi-thorax from the diaphragm to the aortic arch. Note the left pleural effusion.

C. Contrast-enhanced CT scan shows a large well-defined mass that fills the entire left hemi-thorax at the liver. Indistinct fat area (HU - 30 - 70) and mild high-density septa exist within the mass. Multiple calcifications are seen along the outer portion of the mass.

D. Axial T1-weighted image (TR/TE 158/6.2) shows heterogeneous low to intermediate signal intensity of the mass.

chondromatous nodules that were about 1cm in diameter were identified on the lung surface and the parietal pleura (Fig. 1G).

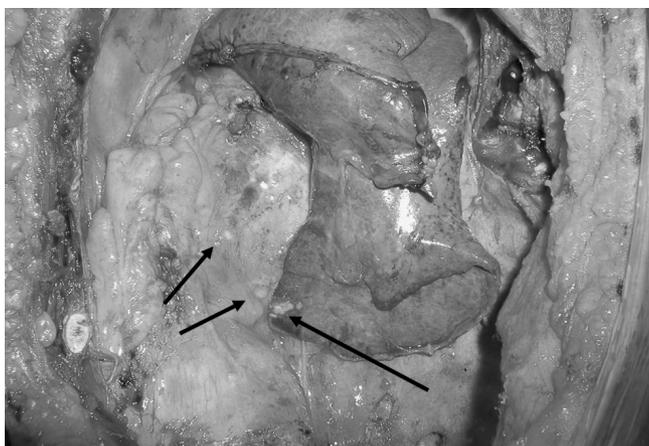
On pathologic examination, the mass was 25 × 20 × 15 cm and weighted 3.5 Kg. The mass was not encapsulated. A cut section of the huge mass showed a partly solid



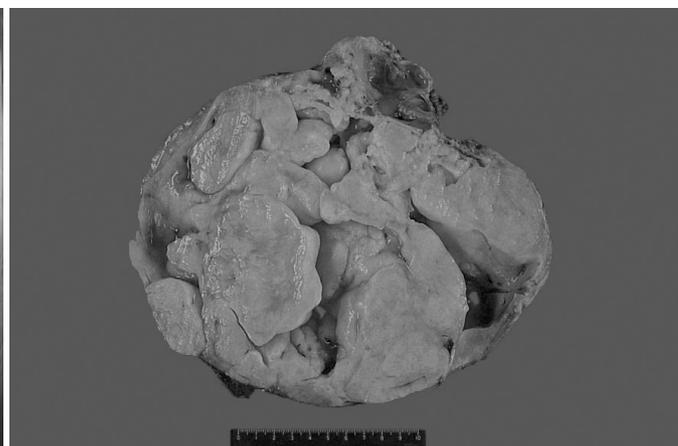
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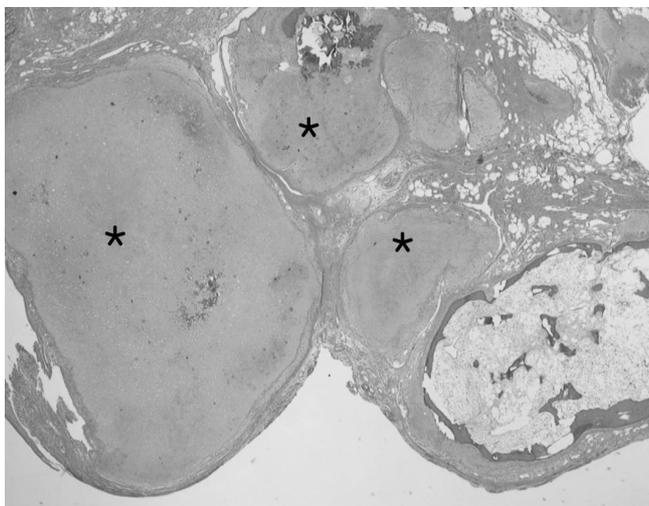
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Fig. 1. E. Contrast-enhanced sagittal T1-weighted image (TR/TE 110/6.2) shows strong enhancement of the periphery and internal septa of the mass.

F. Coronal T2 (haste tra fast)-weighted image (TR/TE 1030/64) shows heterogeneous intermediate to high signal intensity of the mass.

G. A photograph taken at the operation field after the removal of the huge mass. There are multiple chondromatous nodules in the parietal pleura (short arrows) and on the lung surface (long arrow).

H. Cut section of the huge mass revealed a heterogeneous appearance with many cleft-like spaces. Yellowish fatty component and whitish cartilaginous tissue are intermingled within the solid areas.

I. Multiple satellite chondromatous nodules (asterisk) are scattered in the parietal pleura (H and E, × 40).

and partly cystic appearance with large amounts of yellowish mucinous materials (Fig. 1H). Fat and cartilage were the predominant components, and irregular clefts and cystic spaces were noted. Microscopic findings of the huge mass and pleural nodules showed variable components of the mesenchymal tissues, including fibromyxoid tissue, adipose tissue, smooth muscle, cartilage islands with ossification. Among them, adipose tissue with myxoid stroma were predominant. Multiple satellite chondromatous nodules were scattered in the parietal pleura (Fig. 1I). Ed. Note: confirm figure. No atypical or malignant cells were found within the mass. There was no evidence of underlying disease or combined tumors in this case. The patient has been followed up for 2 years following the tumor resection, and her chest radiograph and CT showed no recurrence or metastasis.

Discussion

Pulmonary hamartomas are benign lesions with little or no risk of malignant transformation and minimal risk of recurrence (3). The majority are smaller than 4 cm and solitary. Most are located within the parenchyma, usually in a peripheral location (2, 3). Multiple pulmonary hamartomas are rare with an incidence of 2.8%. Only about 20 such cases have been reported previously in the literature (3).

Rare cases of giant pulmonary hamartomas (4) have been reported, but, to our knowledge, a case of a giant hamartoma with pleural droplets has never been described previously in a clinical setting. The explanation for the presence of pleural droplets is not clearly proved. Multicentric growth may explain the pleural droplets, but this is unlikely because multiple growths are decidedly rare (2).

In one case report, a 30 cm pulmonary hamartoma appeared to be composed of a multitude of small hamartomas (4). Unlike most expansive tumors, hamartomas lack a capsule and show no appreciable compression of the surrounding lung (2).

Although sarcomatous transformation in a long-standing, clinically silent hamartoma has been reported (5), most pathologists do not think a hamartoma can be transformed to a malignant neoplasm. However, considering the possibilities of associated synchronous or metachronous malignancies in hamartomas, including lung cancers (6), patients with such a hamartoma should be submitted for a complete evaluation and subjected to

regular follow ups. Ed. Note: confirm wording. In this case, we did not find any evidence of a malignant transformation nor an unexplained association with other diseases such as Cowden's disease (multiple hamartoma syndrome of autosomal dominant) or Carney's triad (co-existing pulmonary chondromatous lesions, gastric leiomyosarcoma, and functioning extra-adrenal paraganglioma) in the clinical review (1, 3).

The characteristic CT findings of normal hamartomas of the lung are of a smoothly contoured nodule 2.5 cm or less in diameter and focal collections of fat (CT numbers between -40 and -120 in at least eight voxels), or fat alternating with areas of calcification (CT numbers >175 HU) (7). Despite its huge size in our case, the internal architecture and attenuation of the mass corresponded to CT findings of a typical hamartoma.

On MR imaging, there is no evidence of fat and calcification. Hamartomas have intermediate signal intensity on T1-weighted images and high signal intensity on T2-weighted images. They frequently contain septa that have high signal intensity on T1-weighted images and low signal intensity on T2-weighted images (8). Similar findings were observed in this case. MR images correlated well with the surgically resected tumors.

To our knowledge, this case is unique because the pleural droplets appeared as peripheral calcifications of the mass on CT and unusually rapid interval growth for three years. Despite only the presence of benign cells in the mass and pleural droplets, growing hamartomas may be mistaken for carcinomas or sarcomas. In this situation, surgical excision is imperative to exclude malignancy. Pathologically, pulmonary hamartomas have typical benign features. However, a follow up is recommended in patients with multiple pulmonary chondromatous hamartomas for any tumor recurrence.

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