

Two cases of hamartoangiomyomatosis with characteristic scintigraphic findings

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Hamartoangiomyomatosis is a rather rare pulmonary disease which occurs in young to middle-aged women. The chief complaint of this disease is dyspnea. The chest X-ray findings of this disease are such complicated features as reticular, reticulogranular, miliary and honeycomb-like shadow. A pulmonary perfusion scintigram was prepared with a scinticamera after intravenous injection of 10 mCi of Tc-99m MAA. The anterior image showed a remarkably reduced bilateral pulmonary blood flow, especially in the middle and lower areas of the lungs. In the upper portions, the blood flow remained relatively sufficient. The posterior and lateral images also gave similar findings. In two cases of hamartoangiomyomatosis, pulmonary perfusion scintigraphy was successful in identifying the characteristic findings of the disease.

Key words: Hamartoangiomyomatosis, Pulmonary perfusion scintigram

INTRODUCTION

HAMARTOANGIOMYOMATOSIS is a rather rare pulmonary disease which occurs in young to middle-aged women. The chief complaint of this disease is dyspnea.

Chest X-ray findings of the disease include diffuse microgranular, reticular, reticulonodular and miliary or honeycomb-like pulmonary lesions, occasionally accompanied by emphysematous radiologic findings. A majority of the patients with this disease die within 10 years due to respiratory insufficiency.

In this paper, we report two cases of hamartoangiomyomatosis that showed characteristic findings on the pulmonary scintigram.

CASE REPORT

Case 1

A 33-year-old woman was admitted to our hospital because of dyspnea. For 6 years she had been experiencing dyspnea whenever she climbed stairs, but she had ignored it because it had usually disappeared

after she took a rest. The condition became worse, however, and she could not walk more than 10 meters even on level ground without difficulty in breathing. Increased coughing during sleep made her worry about pulmonary tuberculosis, and she visited a hospital. The initial examination showed no abnormalities. She ceased smoking afterwards and this decreased the coughing but the dyspnea and palpitation on exertion persisted. She visited the hospital again, and chest radiography and other examinations were conducted, but no abnormalities were found. Because the signs thereafter increased and worsened, and her body weight decreased, she visited the Department of Respiratory Medicine in our hospital.

At the time of our initial examinations, her dyspnea was so severe that even walking 4 to 5 meters on level ground caused wheezing. The exertional dyspnea was so progressive that even a prolonged conversation provoked to panting, and she experienced increased coughing and sputum. She was therefore hospitalized.

The chest X-rays prepared on admission (Fig. 1) showed diffuse reticular and microgranular shadows throughout the lungs, and there was an apparent finding of super-inflation. A pulmonary perfusion scintigram was prepared with a scinticamera after intravenous injection of 10 mCi of Tc-99m MAA

Received April 23, 1987; revision accepted June 8, 1988.

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(Fig. 2). The anterior image showed a remarkably reduced bilateral pulmonary blood flow, especially in the middle and lower areas of the lungs. In the

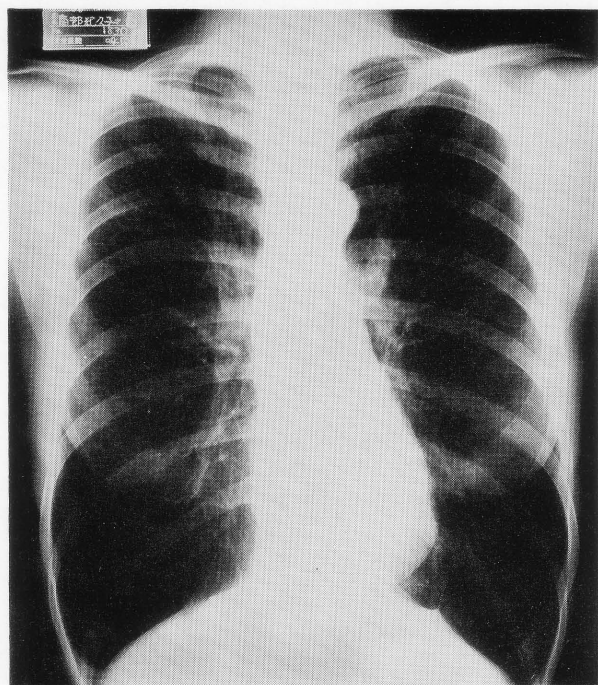


Fig. 1 Chest X-ray film of case 1 shows diffuse reticular and microgranular shadows.

upper portions, the blood flow remained relatively sufficient. The posterior and lateral images also gave similar findings. The patient was given a diagnosis of hamartoangiomyomatosis by transbronchial lung biopsy that showed increased proliferation of hamartomatous smooth muscles in the perivascular and peribronchial tissues and the alveolar walls.

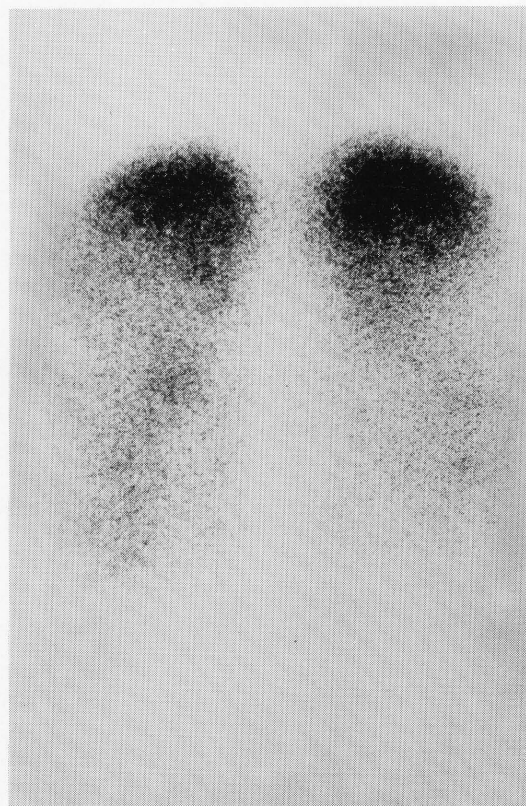
Case 2

A 35-year-old woman suffering from dyspnea for 4 years was admitted to our hospital. She was previously healthy, and had no experience of breathlessness. The onset of breathlessness appeared at the time of her marriage 4 years previously, and was especially severe when she climbed stairs. The symptoms, however, were transiently relieved after the birth of her first child. The breathlessness recurred and worsened three years later and the birth of a second child in February of 1986 was not followed by any improvement in the symptoms.

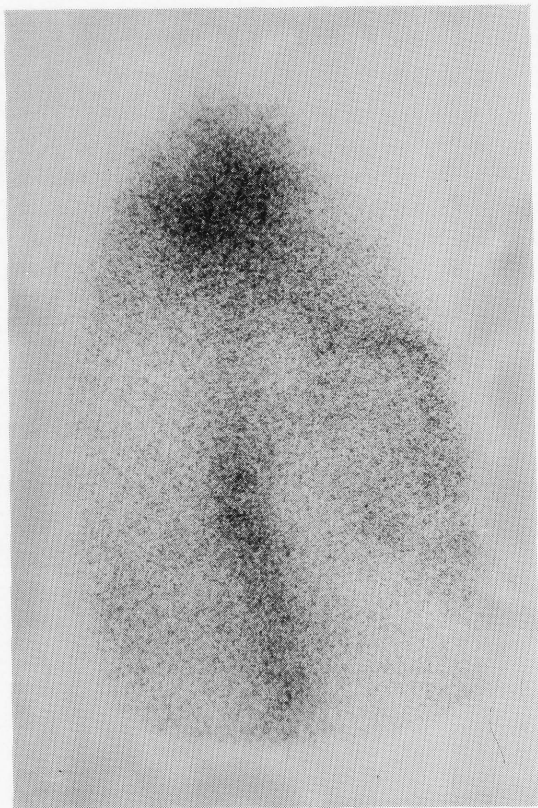
The patient therefore visited a hospital in April, 1986 for closer examination. She was returned to our hospital and hospitalized. Chest X-rays showed diffusive reticular shadows throughout both lungs, coupled with super-inflation (Fig. 3). X-ray tomography and X-ray CT examinations gave a clearer image of the reticular shadow (Figs. 4 and 5). The pulmonary perfusion scintigrams (Fig. 6) suggested



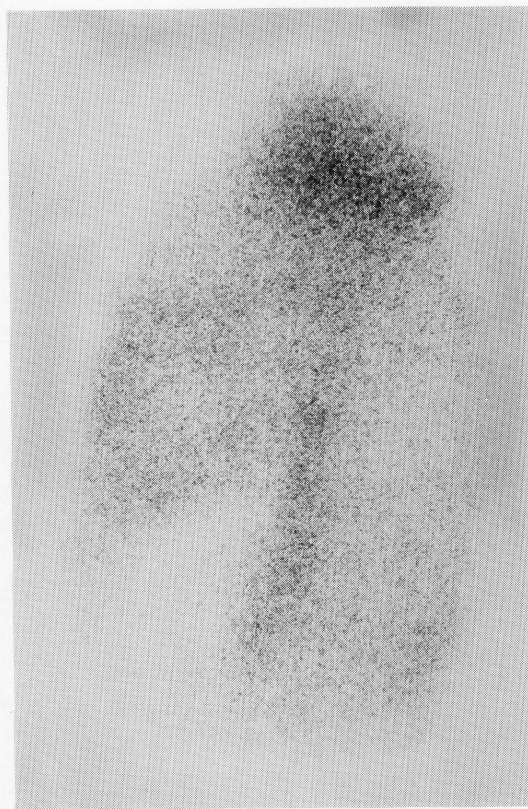
(a)



(b)



(c)



(d)

Fig. 2 Pulmonary perfusion scintigrams of case 1 show blood flow reduction in middle and lower areas of the lungs. a) Anterior, b) Posterior, c) R-Lat, d) L-Lat.

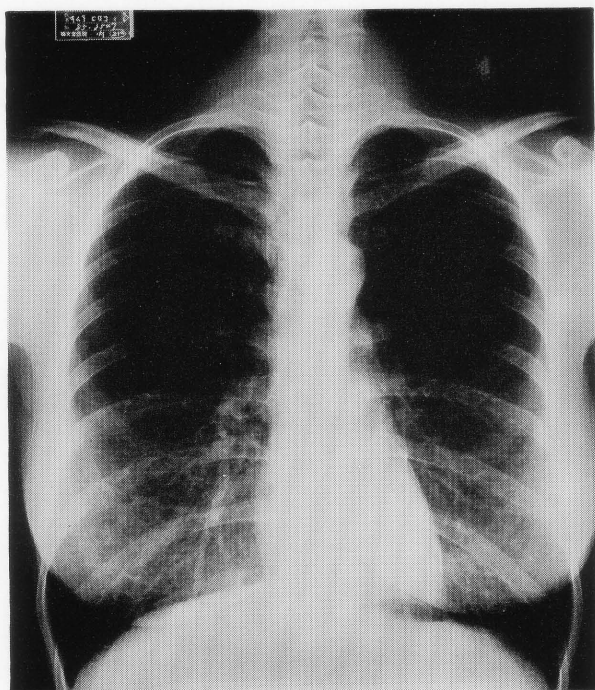


Fig. 3 Chest X-ray film of case 2 shows diffuse reticular shadows throughout both lung areas.



Fig. 4 X-ray tomography of case 2 gives clear images of the reticular shadows.

a more preferential distribution of blood flow in the upper lung areas than in the middle and lower areas in each of the anterior, posterior and lateral images.

DISCUSSION

The chest X-ray findings of pulmonary hamarto-angiomyomatosis are such complicated features as reticular, reticulogranular, miliary and honeycomb-like shadows. The clinical symptoms of the disease are exertional breathlessness, pulmonary dysfunction and hypoxemia. A complication of spontaneous

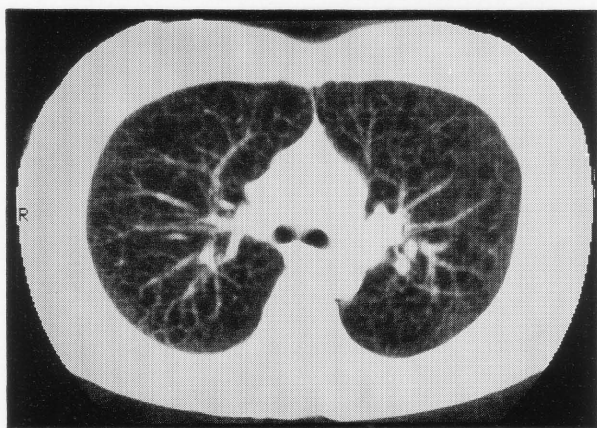


Fig. 5 X-ray CT examination of case 2 gives clear images of the reticular shadows.

pneumothorax is frequent and complications of chylopleura¹ and chyloperitoneum are said to occur in rare cases.

It is reported that alveolar lesions may commonly cause symptoms of restriction disorders in cases with reticular shadows in the X-ray findings,² and that bronchiolar lesions may commonly cause obstructive disorders in cases in which inflation is shown in the X-ray findings.

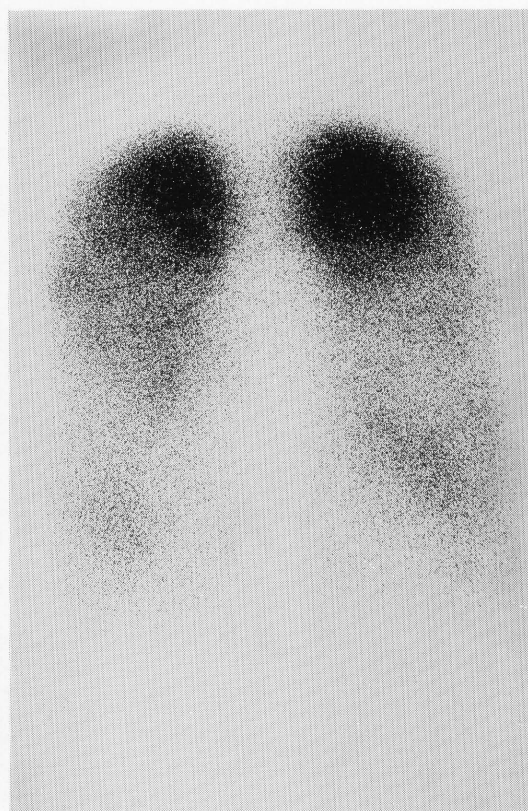
The disease is sometimes accompanied by lymphoma or tubular cerebro sclerosis in addition to the pulmonary changes. This disease had been reported as Angiomyomatose der Lunge, intrathoracic angiomyomatous hyperplasia or pulmonary lymphangiomyomatosis^{1,3,4} until Yamanaka named it hamartoangiomyomatosis from the morphological viewpoint, stressing the congenital factor and its hamartomatous characteristics in 1970.

Morphologically, smooth muscles are distributed in circles or spirals from the larger airways, terminal bronchial walls, respiratory bronchioles and alveolar tubes to the sacs at the alveolar entrances. The smooth muscles also cover the interlobular framework of the lower layers of the pleura pulmonalis in a reticular form, and play an important role in the respiratory motion.

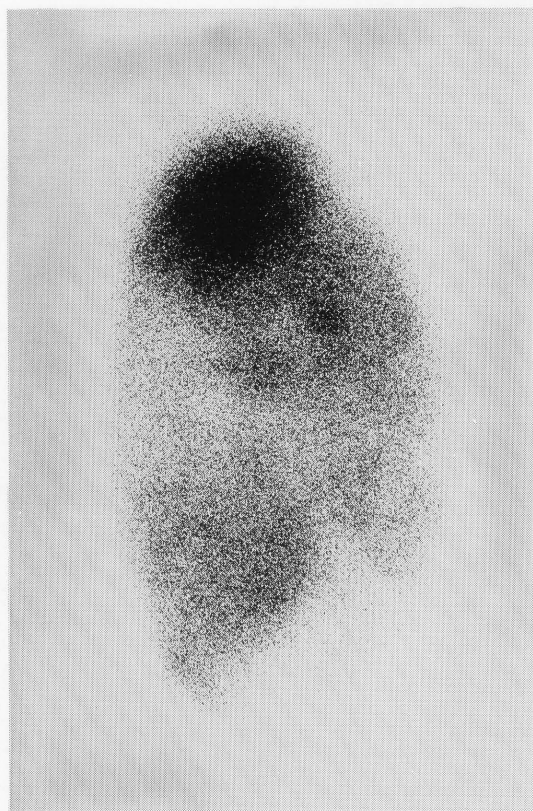
The disease is believed to occur due to hyperplasia of the smooth muscles in the lower layers of the



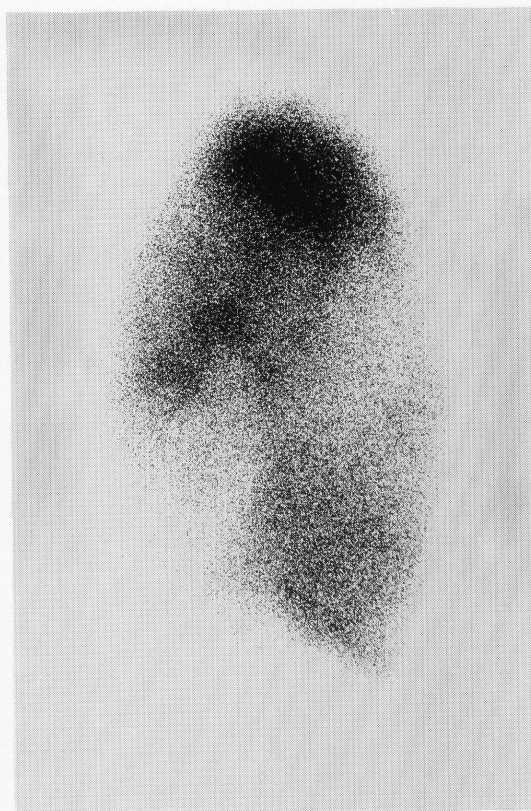
(a)



(b)



(c)



(d)

Fig. 6 Pulmonary perfusion scintigrams of case 2 suggest preferential distribution of the blood flow to the upper lung areas rather than to the middle and lower areas.

pleura pulmonalis, interlobular dissepiments and alveolar walls.⁵ The disease, however, usually features a lack of hyperplasia of collagenous fibers.

Differential diagnosis is required to distinguish this disease from such other diffuse pulmonary shadows as seen in histiocytosis X, sarcoidosis, diffuse interstitial pneumonia, pneumoconiosis and lymph-angiomatous tuberculosis.

When subjected to a pulmonary perfusion scintigraphic study using Tc-99m MAA, the two patients in this study showed increased blood flow in the bilateral upper lung areas and remarkably decreased the blood flow in the middle and lower areas. In cases of pulmonary emphysema, a Tc-99m MAA scan shows so-called "patchy-distribution," or various types of uneven distribution in the bilateral or lateral upper lung area, and uneven distribution in the bilateral middle and lower areas. The scintigraphic findings in the two cases of hamartoangiomyomatosis are characterized by a remarkable increase in the blood flow in the bilateral upper lung areas alone, decrease in the blood flow in the middle and lower areas and overturned cup-like images in the bilateral upper portions. These findings are not seen in the pulmonary perfusion scintigrams of

other diseases. However, there have been no reports which explain why hamartoangiomyomatosis shows a maintenance of pulmonary blood flow only in the upper areas. We think that the volume of pulmonary tissue is smaller in the apical region than in the lower area and thus the resistance to pulmonary pressure may be less in the apical region. However, this requires further investigation.

The two cases reported here showed characteristic pulmonary scintigrams showing greater blood flow in the upper lung areas than in the other cases and remarkable reduction in blood flow in the middle to lower areas. Such findings as characteristic of the present disease are not seen on the pulmonary perfusion scintigrams of other disease.

Thus, pulmonary perfusion scintigraphy coupled with various pulmonary function tests are useful for the differential diagnosis of hamartoangiomyomatosis.

CONCLUSION

In two cases of hamartoangiomyomatosis, pulmonary perfusion scintigraphy was successful in identifying the characteristic findings of the disease.

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