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Clinical Observations

IMAGING FINDINGS OF MALIGNANT FIBROUS HISTOCYTOMA (MFH) IN THE LUNG

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Object: To show the characteristic findings of primary malignant fibrous histocytoma (PMFH) in the lung. Methods: Fifty-one cases of pulmonary PMFH were reviewed, 7 from our hospital and 44 from Chinese literature, altogether 51 cases with chest radiograms, 9 cases with CT scans and 1 case with MRI. Results: PMFH comprises 0.01% of all pulmonary malignant tumors seen in the same period. The mean age was 55 years. The most common complaints were cough, hemoptysis and chest pain. In 40 followed cases, recurrence/metastasis developed in 15 cases (42.9%), 80% arising within a year. The sites of the lesion were: 34 in the right lobe and 17 in the left lobe. It was peripheral in 49 cases (96.1%) and central in 2 (3.9%). The image findings were: large solitary mass (>5 cm, 68.6%) with regular or irregular margin. Most of the tumors were well-circumscribed (56.9%) or lobulated (43.1%) with homogeneous attenuation (64.7%). A few of the tumors had cystic change or cavitation. On CT, the lesion showed low attenuation with necrosis. It might involve the adjacent structures. Conclusions: PMFH of the lung shows no characteristic findings on the images, but in a large mass, with regular, well-defined, less lobulated, shaggy border or thicken wall, which could be differentiated from carcinomas or benign lesions in the lung. CT and MRI are able to provide useful information in demonstrating the nature and extent of invasion by primary fibrous histocytoma in the lung.

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Key words: Fibrous histocytoma, Radiography, Lung neoplasm.

Malignant fibrous histocytoma (MFH), a soft tissue sarcoma in the adults and elderly, commonly occurs in the extremities, trunk and retroperitoneum and secondarily in the craniofacial area but as a primary lesion, it is rarely seen in the lung. Only a few cases have been reported in China and abroad. In this paper, we presented 51 cases of primary pulmonary MFH, 7 from our hospital and 44 from the Chinese literature. Characteristic clinical and imaging findings were elucidated.

MATERIALS AND METHODS

Materials of Our Hospital

Six patients with MFH and one patient with carcinosarcoma (MFH+squamous cell carcinoma) as proved by pathology were seen from November, 1985 to November, 1995, including 6 males and 1 female. The ages ranged from 49 to 68 years with a median of 57 years. Clinical symptoms: cough in 4 cases, hemoptysis in 4 cases, chest pain in 2 cases and shortness of breath or symptomlessness in 1 case respectively. Five cases were diagnosed as lung cancer and one each as sarcoma or bronchocele

preoperatively. The lesion location: 3 cases in the left upper lobe and one case each in the left lower lobe, right upper lobe, right lower lobe and azygos lobe. All tumors were peripheral.

Materials from the Chinese Literature

Forty-four cases of pulmonary PMFH with imaging information as reported in 15 different journals in China during the past decade were analyzed, with 37 males and 7 females. The ages ranged from 10 to 70 years with a median of 54 years (32 cases were ≥40 years). Clinical symptoms: cough 30, hemoptysis 29, chest pain 12, low fever 8, weight loss and fatigue 4, symptomlessness 3 cases. Before operation, 25 cases had been misdiagnosed as lung cancer and 7 as benign lesion (TB 2, echinococcosis 1, cyst 1). Tumor location: 20 cases in the right upper lobe, 3 in the middle lobe, 8 in the lower lobe; 10 in the left upper lobe, 4 in the lower lobe (one patient had two lesions in each of the lungs. As the tumor in the left lobe was not resected, it is not included). Two lesions were located centrally, 1 in the left main bronchus, and the another in the right upper bronchus. Forty-two lesions were peripheral.

Follow-up

Forty-four cases were followed and analyzed, 5 from our hospital and 35 from the literature. Among the 40 cases, the causes of death were unknown in 5 and 11 cases who were followed for less than 2 years were excluded from the calculation of recurrence/ metastasis or survival rates. Among the 40 cases, recurrence/metastasis occurred in 15, including 10 patients died. The rate of recurrence/ metastasis was 42.9% (15/35). The time of recurrence/metastasis was 1–17 months with a median of 6 months, 80% (12/15) of which occurred in 1 year. The ≥2 years survival rate was 31% (9/29 cases); ≥3 years survival 17.2% (5/29 cases); ≥5 years survival 6.9% (2/29 cases). One patient of our hospital had the longest survival of 10 years and 6 months, and is still enjoying a healthy life.

RESULTS

Cases of Our Hospital

Seven cases from our hospital with chest films in 7, CT scan in 5 and MRI in 1. The size of the mass: 3.5×4 cm–13×15 cm, <5 cm in 1 case, ≥5 cm in 3, and ≥10 cm in 3.

Findings of Chest Films

Of all the 7 cases, 2 were irregular in shape, 5 were lobulated, 4 had indistinct margin, 2 had spiculation and 3 had clear margin. The density was homogeneous in 2 cases and heterogeneous in 1. Cystic changes was seen in 1 case. Four cases had matastatic lymph nodes in the pulmonary hilum and mediastinum (Figures 1, 2).

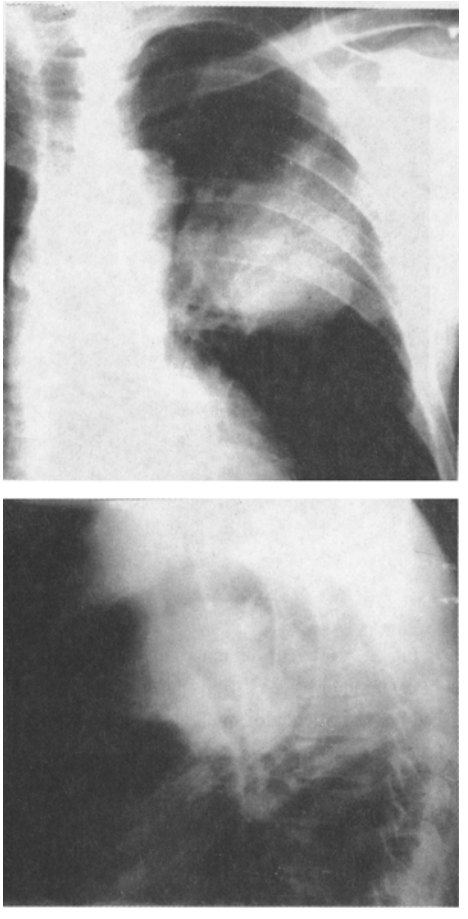


Fig. 1. A 49-year-old man with cough, hemoptysis and chest pain for 7 months.

1a: Posteroanterior chest film: A rounded mass in the left upper lung field is about 9×10 cm, with ill-defined border and heterogeneous density.

1b: Lateral chest film: A mass in apicoposterior segment with lobulated configuration. Part of the margin is indistinct.

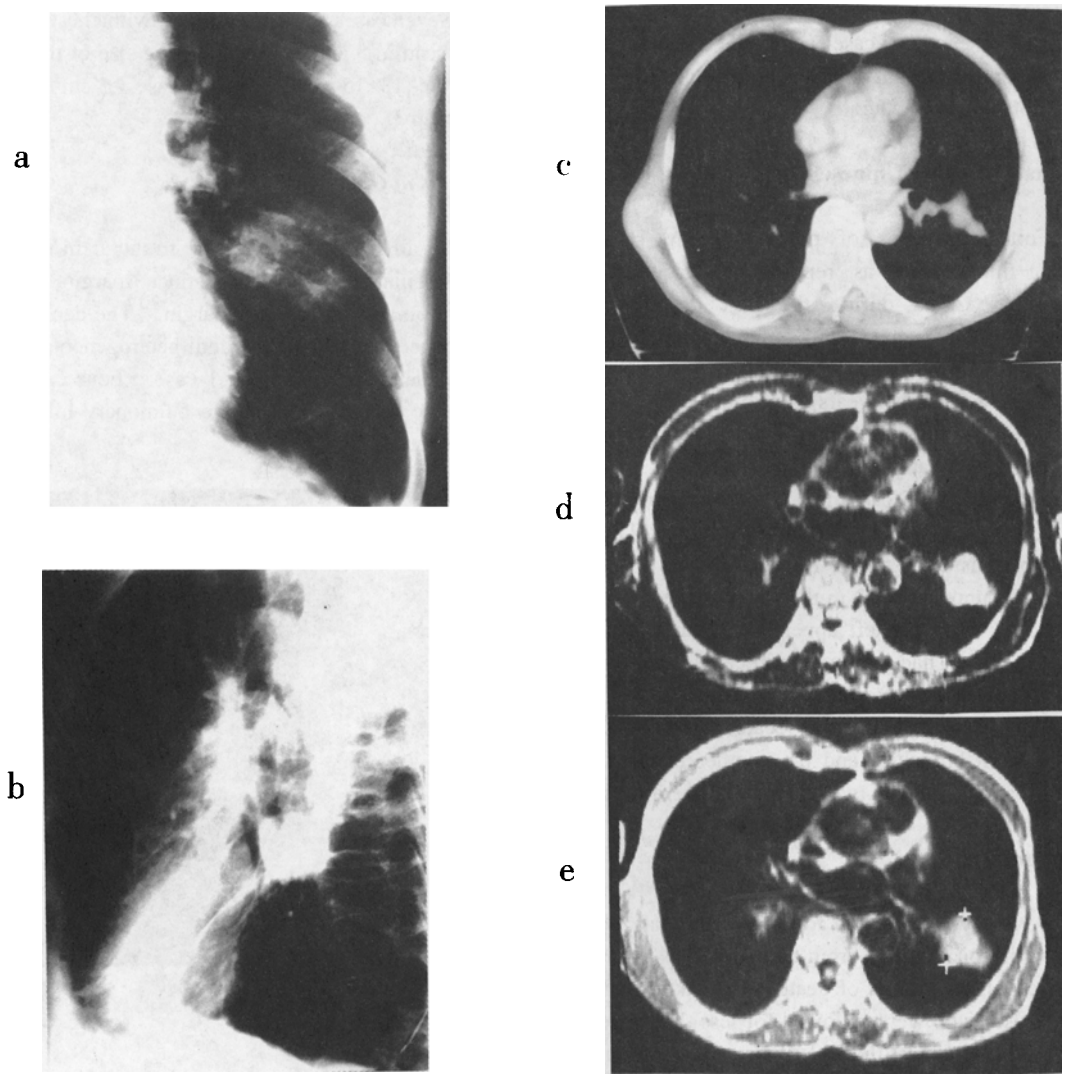


Fig. 2. A 60-year-old man with cough and hemoptysis for one week.

2a: Posteroanterior chest film: An irregular mass about 3.5×4 cm is observed in the left middle lung field, with ill-defined border, spiculation and heterogenous density, contacting with pleura.

2b: Lateral chest film: A mass in the root of the lower lobe contacting with the pleura; part of the margin is clear.

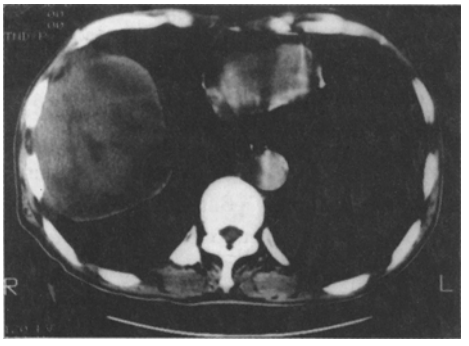
2c: CT scan: A mass in the lower lobe with lobulated configuration and clear margin.

2d: T1WI of MRI: A mass with clear margin. The signal intensity is moderate and homogeneous.

2e: T2WI of MRI: The mass is obviously highly signaled and heterogeneous, with 2 lower signal areas on the margin.

Findings of CT Scan

Among the 5 cases with CT scan, the shape of the mass was irregular and lobulated in 2 and regular in 1. The margin was clear in 3 cases and indistinct in 2 (one had spiculation). The density of tumor was low and not very homogeneous with CT numbers of 5.6–37 Hu. Cystic change was observed in 2 cases, one in each invading the chest wall, pericardium or pulmonary vein. 1 case had matastatic lymph nodes in the mediastinum (Figures 2, 3).



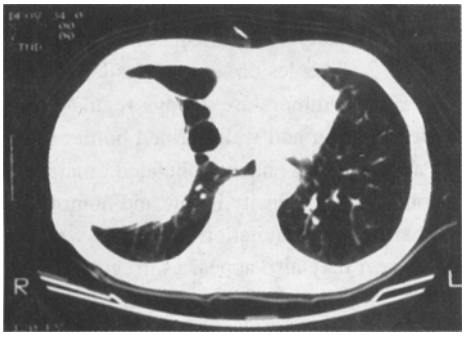


Fig. 3. A 68-year-old man with cough for 1 month. 3a: CT scan: A large mass in the right lower lobe is about 11×8 cm, with clear margin, heterogeneous density and low density areas within. It is contiguous to the lateral chest wall.
3b: CT scan: The margin of the mass is clear. There are a few bubbles in the peripheral lung.

Findings of MRI

One case was lobulated with clear margin, on T1WI, the signal intensity of the tumor was moderate and homogeneous, but the signal was enhanced obviously and heterogeneous on T2 WI. There were only 2 small low intensity signal areas in it (Figure 3).

Cases from the Chinese Literature

Forty-four cases from the literature had chest films in 44 and CT scan in 4. The size of the mass: 2×2 cm–20×16 cm, <5 cm in 15 cases, ≥5 cm in 21, ≥10 cm in 6, ≥20 cm in 2.

Findings of Chest films

The shapes of the mass were rounded or oval in 27 cases and irregular in 1. Seventeen cases were lobulated; clear margin was seen in 26 (4 were capsulated), the margin was not clear in 12 and 2 cases had spiculation. The density was homogeneous in 31 cases and heterogeneous in 4. One case showed high density. The cavitation was thick-walled and irregular at the inner wall in 1 case.

Findings of CT Scan

The shape of mass was regular and not lobulated in 4 cases. The margin was clear in 3 cases and indistinct in 1. The density was high and homogeneous in 3 cases but 1 case showed heterogenisity. The density of the margin was high and necrosis was seen in the center, which is called cystic change with thick wall.

DISCUSSION

As a separate disease entity, MFH was first reported in 1964 by O'Brien and Stout.[1] Since the tumor cells are pleomorphic, MFH is differentiated from the other soft tissue tumors with much difficulty. Enough pathologic material, stringent criteria of histomorphology and immuohistochemical stain are required for correct diagnosis. MFH, one of the most common soft tissue sarcomas in the adults and elderly, occurs in the extremities, trunk as well as retroperitoneum.[1,2] However, 3–35% of this disease is located in the head and neck.[3,4] Although about 80% of MFH found in the lung are metastastic lesions, primary PMFH is very rare. Sarcoma of the lung constitutes 1/500 of pulmonary malignant tumors. In primary sarcoma of the lung, leiomyosarcoma and fibrosarcoma are most common. MFH stands the third.[5,6] The 6 cases of pulmonary PMFH and 1 case of pulmonary primary carcinosarcoma (MFH+ squamous cell carcinoma) in our hospital represent 0.01% of all lung malignancies during this period signifying that MFH is the most common disease of this kind. Pulmonary PMFH may occur at any age, but more often in the adult and elderly with a mean age of 58.6 years. In this series, the median age is 55 years with 74.5% (38/51) more than 40 years old and 58.8% (30/51) more than 50 years old. The sex ratio was 5:1 (male: female). The common symptoms were cough (34 cases), hemoptysis (33 cases), chest

pain (14 cases), low fever, weight loss, fatigue and etc., and 4 without any symptoms. The clinical characteristics such as age and symptoms in this series are similar to that reported abroad.[5,7] Because of the low incidence and nonspecificity in image findings, the correct diagnosis of MFH is difficult before surgical exploration.

Tumors located in the trunk and retroperitoneum appear as a large soft mass with irregular shape. The density is usually heterogeneous with necrosis areas on the imaging. The mass usually gives obvious enhancement on contrast-enhancement by CT scan.[4] The lesion usually invades the adjacent structures and organs. Miller[8] reported the mass showing homogeneous signal intensity with clear margin and low signal intervals on T1WI. The signal intensity is high and heterogeneous, accompanied with low signal intervals and indistinct border on T2WI on the MRI. 70% of the pulmonary PMFH occurs in the peripheral lung field without preference on the left or right. More than 50% of tumors invades the lower lobe appearing always as a single lesion. The mass is large showing low density and clear sharp margin. 50% involves the pericardium and pleura.[5-7] In this series, 34/51 of lesions were located in the right lung, 21 in the upper lobe, 3 in the middle lobe, 9 in the lower lobe and 1 in the azygos lobe. 17/51 tumors were located in the left lung, 13 in the upper lobe and 4 in the lower lobe. Only 3.9% (2/51) were central and 96.1% (49/51) were peripheral, which reflects some difference from the foreign report. Yet, similarity was noted as to the morphological characteristics and image findings of the lesion—a soft mass which was very big. About the size, ≥5 cm in 24 cases (47.1%); ≥10 cm in 9 (17.6%) and ≥20 cm in 2 (3.9%). Chest films showed that the mass was rounded, oval or irregular in shape. 22 cases were lobulated. In 29 cases the margin was clear (4 with capsule) and in 16 cases the margin was indistinct (3 with spicules). The density was homogeneous in 33 cases, heterogeneous in 5 and high in 4. Cystic change was observed in 1 case and the shape could change with deep inhalation. One case had thick wall cavitation and irregular inner wall. In 9 cases with CT scan, the shape of the mass was regular in 5, irregular and lobulated in 2, six cases had clear margin and 3 had indistinct margin. The density was homogeneous and high in 3 cases. Four cases of our hospital showed low density with CT number of 5.6–34.2 Hu. Two of the 4 cases showed typical cystic changes, each invading the chest wall, pericardium or pulmonary vein. There were 3 cases with metastases to the pulmonary hilum and midiastinum. Only one patient who had MRI scan showed that the mass was lobulated with clear margin. On T1WI, the tumor was moderate in intensity and homogeneous in signal. But on T2WI, the signal intensity was high and heterogeneous. CT and MRI are helpful for showing the shape, density and extent of invasion. They can also help arrive at a correct diagnosis preoperatively and planning of the treatment.

Based on this analysis, the chief differences of image findings of pulmonary PMFH as compared with those of lung cancer are: 1. Most lesions (96.1%) are peripheral; 2. The lesion is usually large (≥5 cm 68.6%); 3. The tumors are always regular in shape having clear margin and well defined border (56.9%), though a few may have lobulated margin and spiculation; 4. The density is low and homogeneous. Cystic change and cavitation are rarely observed. Yet lung cancer may also appear as irregular nodule or mass with indistinct and lobulated margin with spicules. However, there are always concurrent signs of infiltration, obstructive pulmonitis or atelectasis in the peripheral lung field if it is a lung cancer. Being different from benign pulmonary diseases, the margin of MFH mass is clear but not sharp, the mass is homogeneous but always has low signal area in it. Cystic changes could be found, but the wall is thick and irregular. The soft tissue part may be enhanced in contrast material enhancement on CT. On T2WI of MRI, the signal intensity of the mass is heterogeneous and obviously high, which is different from the moderately high intensity signal of lung cancer with necrosis in the tumor.

The treatment of MFH is primarily surgical, but combined treatment could be used though the outcome is not satisfactory. The local recurrence rate could be as high as 51% after complete resection.[9] For the completely resected cases the mean 5-year survival rate is about 50%.[10] In 40 followed cases in this series, the recurrence/metastasis rate was 42.9% and 80% of them developed this within one year. For these cases, the 5-year survival rate is only 6.8%. In this series, a patient with the longest survival has been alive for 10 years and 6 months. He was suspected as having recurrence half a year after the initial operation. Then, combined radiation therapy and chemotherapy was given and the patient has survived long. This may prove that MFH is a kind of tumor

with high malignancy and frequent recurrence, and may be poor in prognosis. If the lesion is found early enough and treated energetically, the patient could survive for a long time.

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