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# Successful Management of a Recurrent Primary Malignant Fibrous Histiocytoma of the Lung: Report of a Case

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We describe herein an extremely rare case of a recurrent primary pulmonary malignant fibrous histiocytoma 3 months after operation that occurred in a 55-year-old man who was treated with chemotherapy and radiotherapy successfully. Until now, 36 months later, the patient has shown no evidence of tumor recurrence. The clinical, radiographic, and pathologic features are reported here together with a brief review of the literature.

Key words: 1. Lung neoplasms
2. Surgery
3. Chemotherapy
4. Histiocytoma

## CASE REPORT

### 1) Case summary

A 55-year-old male presented with a two months history of cough and chest pain. Six months ago, there were no abnormal findings in chest X-ray at that time presented with mild chest pain. At the time of cough with chest pain presentation, chest X-ray showed a about 5 cm sized round mass in left hilar area and computed tomography (CT) scan of chest showed a about 5×4 cm lobulated mass arising superior segmental bronchus in lower lobe with interlobar pulmonary artery invasion (Fig. 1A, B). Positron emission tomography (PET)-CT was revealed only increasing uptake of hilar mass without anywhere active lesion. Brain magnetic resonance (MR) showed no evidence of metastasis. Bronchoscopic finding was a complete obstruction of superior segmental bronchus. Bronchoscopic and CT-guided needle biopsy of the lesion results a histological diagnosis of suggestive sarcoma. The hilar lesion could be considered as the primary malignant tumor, based on clinical findings including CT scan of chest, PET-CT, and brain MR. The patient underwent a left pneumonectomy with a dissection of the regional lymph nodes.

### 2) Pathology, postoperative recurrence, and management

The resected tumor was encapsulated, firm, and areas of focal hemorrhage and necrosis with pulmonary arterial invasion by tumor.

Microscopically, the tumor was consisted of a pleomorphic and spindle cell proliferation in a storiform pattern with high grade differentiation (Fig. 1C). By an immunohistochemical staining, the tumor was positive for CD68, but negative for S-100 protein and desmin. The histological final diagnosis was typical of malignant fibrous histiocytoma (MFH) by the features of microscopic finding and special staining. There was no evidence of regional lymph node metastasis and bronchial and arterial resection margin were negative.

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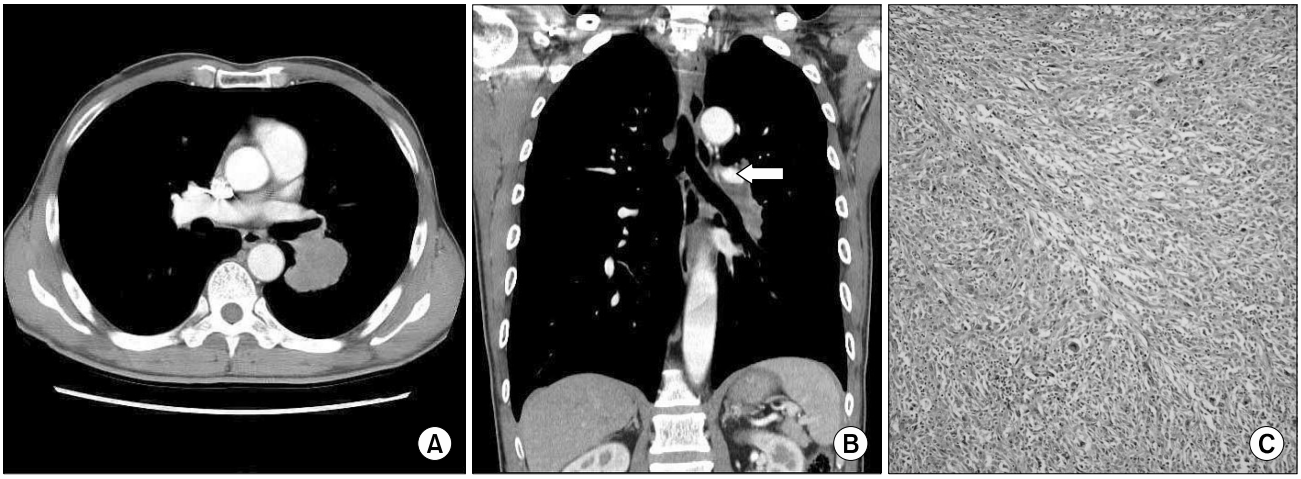


**Fig. 1.** Computed tomography scan of chest shows (A) about 5×4 cm lobulated mass arising superior segmental bronchus in lower lobe with (B) interlobar pulmonary artery invasion (arrow). (C) The tumor was composed of pleomorphic, spindle-shaped cells arranged in a storiform pattern (H&E, ×40).

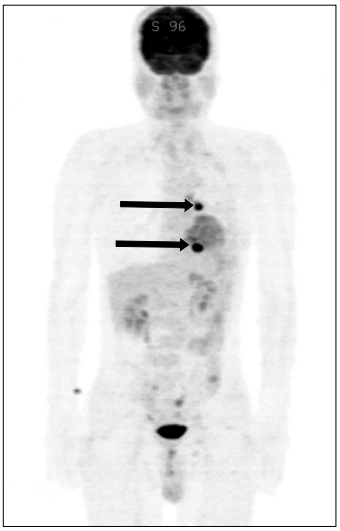

**Fig. 2.** Positron emission tomography-computed tomography images showing a local relapse of a tumor in postpneumonectomy space with an increased uptake (arrows).

Postoperative recurrence was developed on pleura in pneumonectomy space during first regular follow-up three months after the operation. The CT scan of chest and PET-CT showed a positive finding of recurrence (Fig. 2). There were no findings of extrathoracic metastasis.

The patient was received chemotherapy including doxorubicin, followed by radiotherapy for salvage management. The patient is still alive without any evidence of cancer recurrence in the chest and another site for 36 months after operation.

## DISCUSSION

MFH is the most common soft tissue sarcoma in adult, arising frequently in the extremity, retroperitoneum and trunk [1]. Metastasis to the lung as secondary MFH is not uncommon. But primary MFH of the lung is extremely rare, accounting only for less than 0.2% of lung neoplasm [1]. To diagnose primary MFH, there must be no evidence of tumorous lesion except for primary site by careful physical examination and radiological study. In our case, there was only pulmonary lesion in CT scan of chest and PET-CT without extrathoracic lesion. Up to date, Maeda et al. [2] reviewed the clinical features of only 93 cases of MFH, searching English medical literature in the world.

The median age of these patients was 55 years, range from 10 to 80 years [3]. In our case was compatible with the literature. Pediatric cases have been reported as age range [4]. Male was predominance, but some literature reported female frequently occurred [3]. The common symptoms of patients were coughing, chest pain, and dyspnea. Weight loss, hemoptysis, and fatigue were also observed. But about 32% patients were symptom free [2,3].

In our case, just six months after first symptom of scanty

chest, the tumor has appeared in CT scan of the chest and PET-CT. There were relatively fewer symptoms at the early stage and the tumor thought to be having a tendency of rapid growing. Maeda et al. [2] reported that a preoperative diagnosis is difficult and only 4% of their reported cases were histologically diagnosed prior to resection. Maybe, this is because of rarity of MFH and insufficient biopsy specimens by rapid growing. Our case was similar to reported cases. Therefore, symptom is never missed to detect this disorder.

The microscopic features of MFH are similar to other soft tissue sarcoma. The differential diagnostic diseases are other sarcoma, malignant schwannoma, spindle-cell and anaplastic carcinoma, and malignant melanoma. So, immunohistochemical stain is required for the precise diagnosis [5]. Weiss and Enzinger [1] have proposed storiform, pleomorphic, myxoid, inflammatory, giant cell, and angiomatoid variants may be occurred. The cell feature of our case was a pleomorphic spindle-shaped cell arranged in a storiform pattern.

Maeda et al. [2] reported, cases without a resection who died of the disease, the median survival was 9 months, range from 2 months to 24 months, and the 5-year survival rate for the patients with resection was significantly better than those without resection. As for treatment of MFH, surgical resection is the most important therapy.

Weiss and Enzinger [1] reported that cases with lymph node metastasis account for 12% of all cases. Other report has similar result in 19%. The 5-year survival rate for the patients without lymph node metastasis was better than those with lymph node metastasis. And patients with lymph node metastasis at surgery were frequently recurred compared with without lymph node metastasis [2]. Our case had turned out not to be lymph node metastasis at surgery. But recurrence was detected at pleural space 3 months after operation. We are thinking of the tumor behavior being rapid growing and relative early recurrence in high grade.

Systemic metastases at presentation are not infrequently reported. A brain is a common site of metastasis. In our case, metastasis to the brain was not discovered in radiographic study.

It is likely to controversy over adjuvant therapy such as radiation and chemotherapy. Radiation therapy has not commonly been used in the treatment regimen [3]. Drugs used for chemotherapy are doxorubicin, dacarbazine, cyclophosphamide, and cisplatin. No series reported the response to chemotherapy [3]. But chemoradiation is likely to be having an effect on recurrent case of MFH the same as our case.

So, we report recurrent case of postoperative primary MFH was treated with chemotherapy and radiotherapy successfully, and we do suggest primary MFH should be considered for careful and closed follow-up to detect recurred case as early as possible and postoperative adjuvant chemotherapy need to a selected patient who has large tumor size and high grade differentiation.

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