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CASE REPORT

Primary Pulmonary Malignant Fibrous Histiocytoma

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Abstract

Malignant fibrous histiocytoma (MFH) cases are classified within the group of nonclassified sarcomas. The etiopathogenesis is unclear; however, MFH commonly develops in scar tissue and in areas exposed to radiation. MFH is the most common soft tissue sarcoma in adults and may be borne in the lungs, chest wall, mediastinum, or other tissues. Primary MFH of the lung constitutes less than 0.2% of all pulmonary neoplasms; thus, an optimal treatment strategy has not yet been elucidated. We aimed to report a case of MFH of the lung with subsequent treatment administration.

KEYWORDS: Malignant fibrous histiocytoma, pulmonary neoplasm, lung

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INTRODUCTION

Primary malignant fibrous histiocytoma (MFH) of lung is a rare kind of tumor; so that an optimal management of cases with primary malignant fibrous histiocytoma (MFH) has not yet been decribed. We aimed to share our experience on primary MFH of the lung.

CASE PRESENTATION

A 50-years-old man was admitted to our clinic with dyspnea and chronic cough. He had a history of smoking approximately 30 packs of cigarettes per year. A clinical examination including auscultation revealed decreased sounds of the left hemithorax. A full blood count showed a normal hemoglobin level, and renal and liver function test results were also normal. Chest X-ray showed opacity in the left lung, and a chest computed tomography (CT) scan revealed a 75×68×96 mm mass in the left lung (Figure 1). Additionally, there were a suspicion of chest wall invasion, particularly at the second rib. A preoperative bronchoscopic examination did not yield a definitive diagnosis, and transthoracic, fine-needle aspiration biopsy of the lesion was nonrepresentative (necrotic tissue). Positron emission tomography (PET) scanning showed high-intensity fludeoxyglucose (FDG) uptake in the 79×75×100 mm mass in the upper left lobe (SUVmax (maximum standard uptake value) of 9.7), but there was no suspicious uptake of metastatisis, in other areas (Figure 2). The patient's skeletal system and intracranial structures showed no FDG uptake. The patient's respiration function test results were unremarkable.

We decided to perform mediastinoscopy due to a high suspicion of a malignancy because of the high FDG uptake in the tumor. Mediastinal 4R, 4L, and 7 levels of lymph nodes were sampled, which revealed anthracosis on frozen section analysis. In the same operation, the left hemiclamshell approach was used due to its ability to provide a wide exposure that allowed the safe and complete removal of the lung cancer involving the mediastinum and apical thoracic dome. We prefer the hemiclamshell incision to provide optimal exposure of the hilar and mediastinal vascular structures. Upon exploration, an 8×10 cm lesion in the upper left lobe was detected. No chest wall invasion was found. Wedge resection from the preoperative, undiagnosed mass was performed, and frozen section analysis revealed a malignant tumor. We subsequently performed lobectomy of the upper left lung lobe. Levels 5, 6, and 11 were dissected after lobectomy. The patient's postoperative course was uneventful. Drains were taken out on postoperative day four, and the patient was discharged on the fifth postoperative day.

Cellular neoplasms that consist of malignant fusiform tumor cells that create short bundlers and storiform pattern are detected on histopathological analysis. Bizarre malignant cells and necrosis attend to these cells. Tumor cells showed vimentin (+), CD 68 focal (+), S100 (-), SMA (-), PanCK (-), and Ki67 80% (+) immunoreactivity on immunohistochemical evaluation, which strongly sug-

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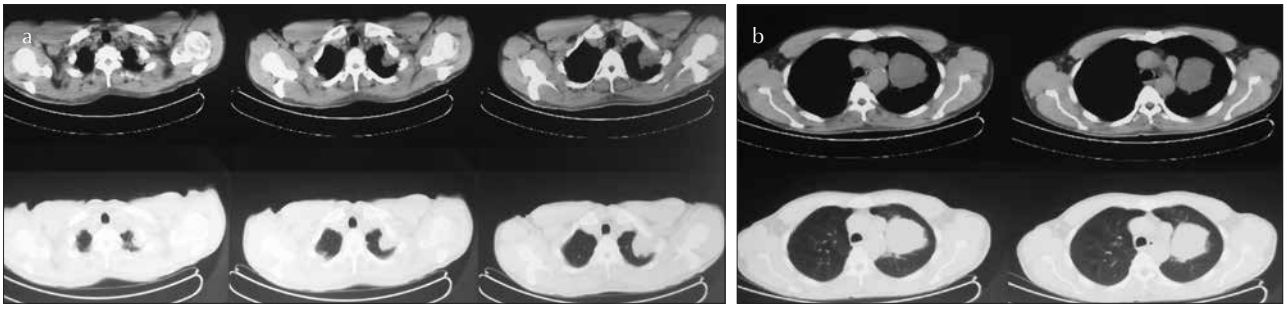

Figure 1. a, b. A chest computed tomography (CT) scan revealed a 75×68×96 mm mass in the left lung. (a) Suspicion of tumor invasion to the first rib. (b) Adjacency of the tumor to mediastinal vascular structures

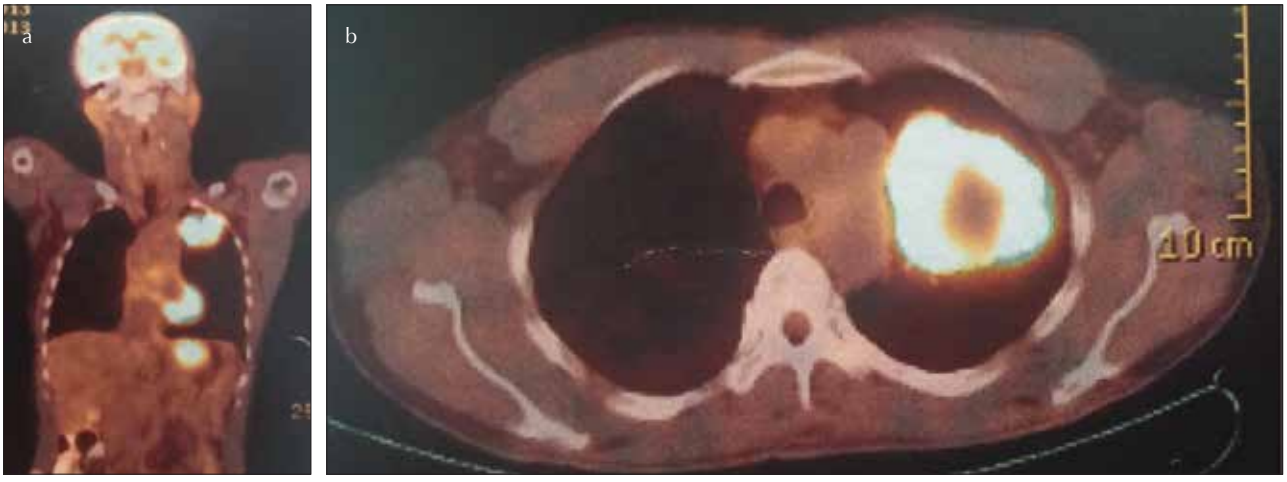

Figure 2. a, b. Positron emission tomography (PET) scanning showed high-intensity fludeoxyglucose (FDG) uptake in the mass in the upper left lobe (SUVmax of 9.7)

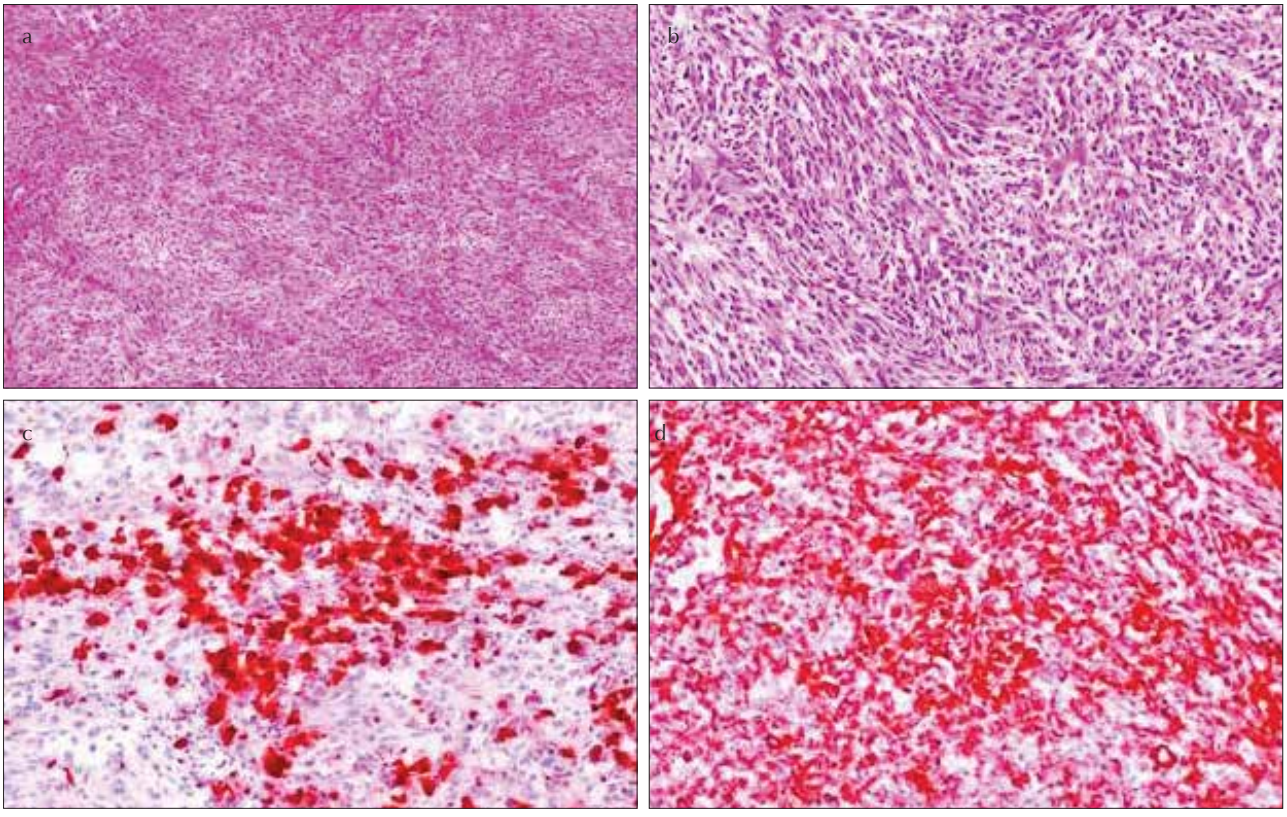

Figure 3. a-d. (a) Representative hematoxylin and eosin (HE) sections at original 4× magnification. Lesion created short bundles and consist of fusiform cells that show a storiform pattern (HE, 4×). (b) Bundles of malignant fusiform cells (HE,10×). (c) Tumor showed CD68 (+) immunoreactivity (HE,10×). (d) Tumor showed vimentin (+) immunoreactivity (HE, 10×)

gested malignant fibrous histiocytoma (MFH) (Figure 3). Resection margins were negative. Dissected lymph nodes were negative.

We took permission from patient to publish this case report at this stage. Recurrence was not detected in the three-year follow-up.

DISCUSSION

Reports on MFH were first described by Weiss and Enzinger [1]. MFH is one of the most common soft tissue tumors in adults. It can occur anywhere in the body, but it most commonly originates in the lower extremities and retroperitoneal region. MFH of the lung constitutes less than 0.2% of all pulmonary neoplasms. However, an optimal treatment strategy has not yet been elucidated [2].

Most MFH cases are asymptomatic (32%); however, the most common clinical symptoms are chest pain, dyspnea, cough, weight loss, fatigue, and hemoptysis [2-4]. Our patient's symptoms were cough and dyspnea. MFH can be seen between the ages of 10 and 80 year (average age, 55 years) [4]. There are different opinions on its frequency depending on gender [2].

Although most lesions are seen as solitary pulmonary masses or nodules, bilateral masses can also be seen. The frequency of occurrence in the right or left lobes is almost equal. Punctate calcifications on CT in pediatric patients have been reported [5]. Ipsilateral pleural effusion can occur in 20% of the patients [5], and endobronchial lesions have also been reported [6]. Without intending to make a generalized statement about FDG uptake in primary pulmonary MFH because it is a type of tumor that is rarely seen in the lungs, in our case, the SUVmax was 9.7. It is believed that PET-CT is useful to exclude other regions of tumors possibility [2].

Malignant fibrous histiocytoma histologically consists of varied spindle-shaped fibroblasts and histiocytes with atypical pleomorphic giant cells. Five distinct histologic subtypes (storiform-pleomorphic, myxoid, giant cell, inflammatory, and angiomatoid) have been described. The most common histologic subtype noted in primary pulmonary MFH is storiform-pleomorphic. Tumors are usually stained with desmin, actin, vimentin, keratin, and neurogenic tumors are often obtained with these stains [2]. In our case, a definitive pathology showed storiform patterns, bizarre cells, and vimetin- and CD68-positive cells. The cytological findings strongly suggested MFH.

Metastatic MFH lesions are more frequently reported in the lungs than primary pulmonary MFH lesions. Patients with MFH detected in the lungs should undergo an exhaustive diagnostic examination (including PET-CT and histopathological verification) to specify that the tumor is primary pulmonary MFH or a metastasis of extrapulmonary primary origin [7].

Because cases diagnosed with primary pulmonary MFH are quite rare, it is difficult to define a management protocol for these patients. In addition, patients with MFH usually have poor survival rates; it has been reported that the five-year survival rate for patients without lymph node metastasis is better than those with lymph node metastasis [8].

According to a review that analyzed previous case reports, surgical resections are preferred if the tumor seems resectable. These malignancies show a tendency for both local recurrence and distant metastasis, but despite their aggressive nature, there are several reports of patients with long-term survival. Better survival rates are mostly related to complete surgical resections with clear margins. In the same vein, survival rates are poor for patients with advanced-stage disease, incomplete resection, or tumor invasion to the mediastinum or chest wall, and the chances of recurrence and metastasis are higher [7]. Our patient had a three-year survival without recurrence.

In addition, adjuvant therapy can be performed; hence, radiotherapy is not preferred [9]. Although doxorubicin, dacarbazine, cyclophosphamide, and cisplatin might be used during chemotherapy, there are, in fact, no large-scale studies that report a favorable response to chemotherapy [7]. Close follow-up is advised due to the relatively high recurrence rates of MFH.

In conclusion, we presented a case of primary pulmonary MFH that is a very rare lung tumor. We also showed that although MFH is an aggressive tumor, long-term survival can be possible with surgical resection in early-stage tumors.

Informed Consent: Written informed consent was obtained from patients who participated in this study.

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