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A Case of Juvenile Primary Pulmonary Malignant Fibrous Histiocytoma

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An 18-year-old female presented with an abnormal shadow of a 22-mm nodule in the left anterior segment of the upper lung lobe (S^3) on chest X-ray and computed tomography (CT). Thoracoscopic partial lobectomy was performed, when a frozen section suggested spindle cell sarcoma of extrapulmonary origin, including malignant fibrous histiocytoma (MFH). A histologic diagnosis of the MFH was rendered postoperatively on permanent sections. As no other primary focus was detected in spite of intensive investigation, including ^{18}F-fluorodeoxyglucose positron emission tomography - CT scans, a definitive diagnosis was made of primary pulmonary MFH. Primary pulmonary MFH is usually treated by total resection, however, in the current case we performed a partial lobectomy with clear margins and a watchful follow-up without any additional aggressive treatment. No relapse has been observed for 3 years after the surgery.

Key words: malignant fibrous histiocytoma (MFH), juvenile, lung, video-assisted thoracoscopic surgery (VATS), one-port method

INTRODUCTION

Malignant fibrous histiocytoma (MFH) is a soft tissue sarcoma that often occurs in the lower limbs and hip region, occasionally in the retroperitoneum but less commonly in the trunk, and is most common in patients over age 40. So, MFH rarely occurs in the lung and is very rare in patients under age 20. We surgically treated a case of primary pulmonary MFH in an 18-year-old female who has survived without relapse for 3 years since the surgery.

CASE REPORT

The patient was an 18-year-old female. A shadow of a 22-mm nodule was detected on the chest X-ray in the left upper lung field (Fig. 1) on a routine checkup when she entered university. An enhanced chest computed tomography (CT) performed at a nearby hospital demonstrated a similar tumor with distinct border and smooth surface in the left anterior segment of the upper lung lobe (S^3), which was evenly imaged (Fig. 2). She was referred to our hospital for diagnosis and treatment by surgery, without undergoing bronchoscopy. She did not experience any loss of weight, fever, or other symptoms. Hematological and biochemical examinations revealed no abnormalities, including the CEA level. A benign tumor such as sclerosing hemangioma was suspected on radiological images. Compared with the past chest X-ray taken a few years earlier, we evaluated this tumor as rapidly enlarging, and the differential diagnosis included malignant neoplasms and tuberculoma.

We performed a partial lobectomy of the left upper lung lobe with use of video assisted thoracoscopic one-port method surgery (Fig. 3), since it is less invasive, low-risk and provides a cosmetic advantage compared to open surgery. The attending pathologist informed the surgeons of a frozen section diagnosis of nonepithelial malignancy, most likely a lung metastasis of a malignant soft tissue tumor including MFH. As a result, a complete lobectomy was not performed.

Histopathological investigation on permanent sections revealed spindle cell sarcoma, arranged in a storiform pattern and intermingled occasionally with multinucleated giant cells with sharp external borders (Fig. 4a, b). Mitotic figures are rare with MIB-1 index less than 5%. Immunohistochemically vimentin is positive, S100 is focally positive only in the giant cells (Fig. 5), whereas SMA and desmin is negative. All these findings are quite consistent with MFH.

Postoperative investigation including the systemic ^{18}F-fluorodeoxyglucose positron emission tomography demonstrated no extrapulmonary primary focus. Therefore we finally diagnosed this case as primary pulmonary MFH. Since the patient did not want additional intervention, we have been conducting watchful follow-up observations, and there have been no relapses for 3 years since the surgery.

DISCUSSION

Malignant fibrous histiocytoma (MFH) is histopathologically characterized by an admixture of fibroblast-like and histiocytic cells, arranged in a storiform pattern [1]. Histogenesis still remains controversial, including a theory of histiocytic origin [2, 3] and of undifferentiated mesenchymal cell origin [4]. MFH

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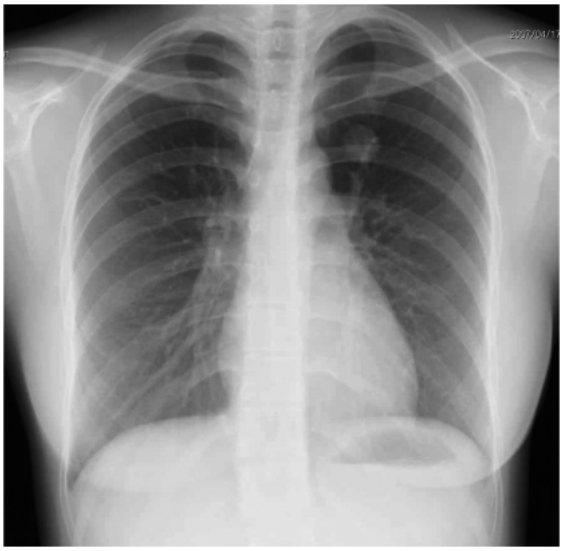

Fig. 1 A plain chest X-ray image at the first visit to our hospital. A nodule with a distinct border was observed in the left upper lung.

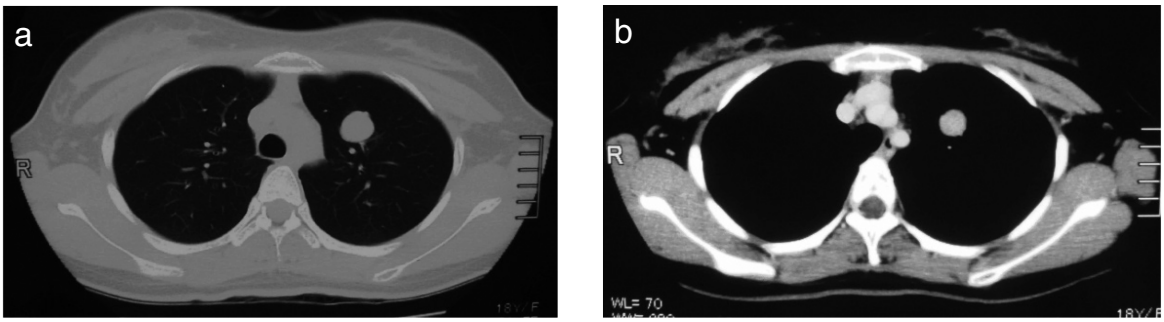

Fig. 2 Enhanced chest CT images, showing a nodule in the left lung S^3.

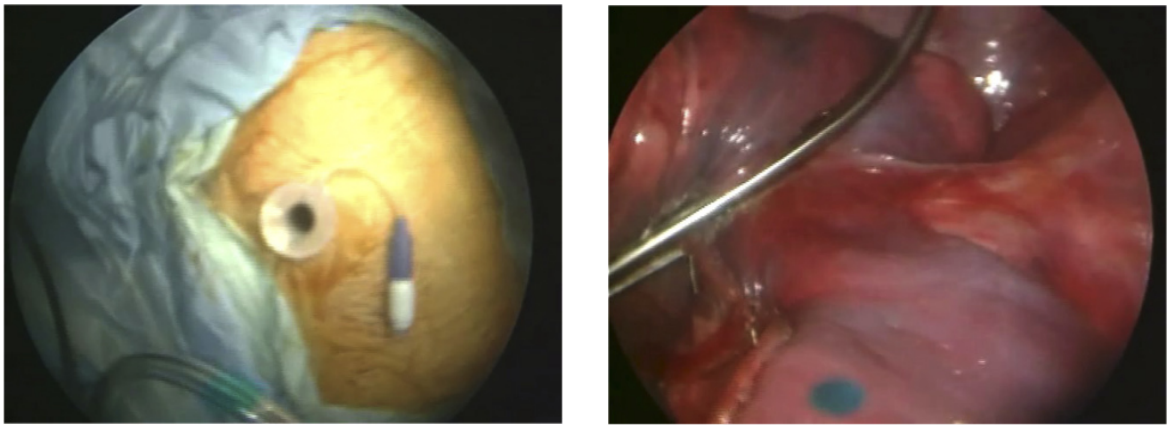

Fig. 3 Surgical findings. One-port method (left). Partial lobectomy of the left upper lung lobe using thoracoscopy (right).

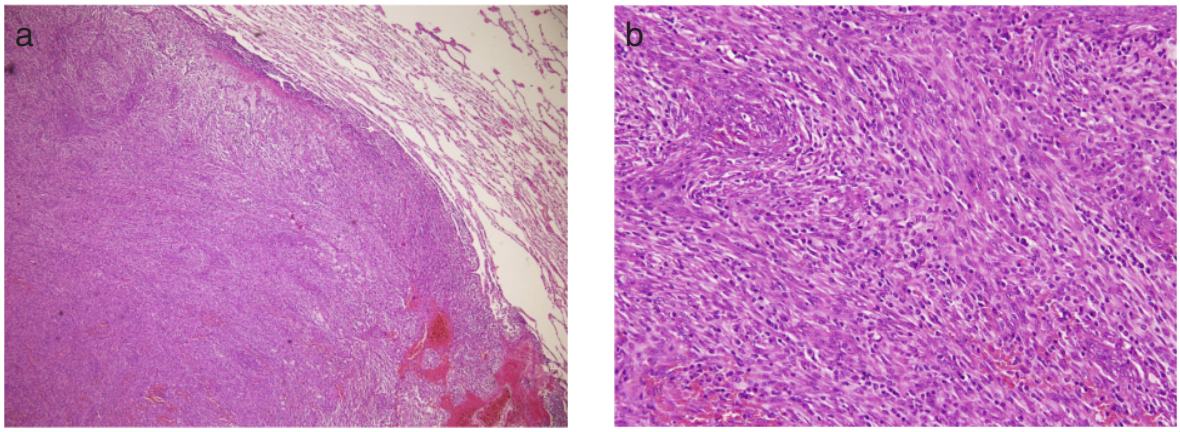

Fig. 4 The tumor with a distinct border (4-a) (× 20), consisting of spindle tumor cells in a storiform pattern, intermingled with giant cells (4-b) (× 100).

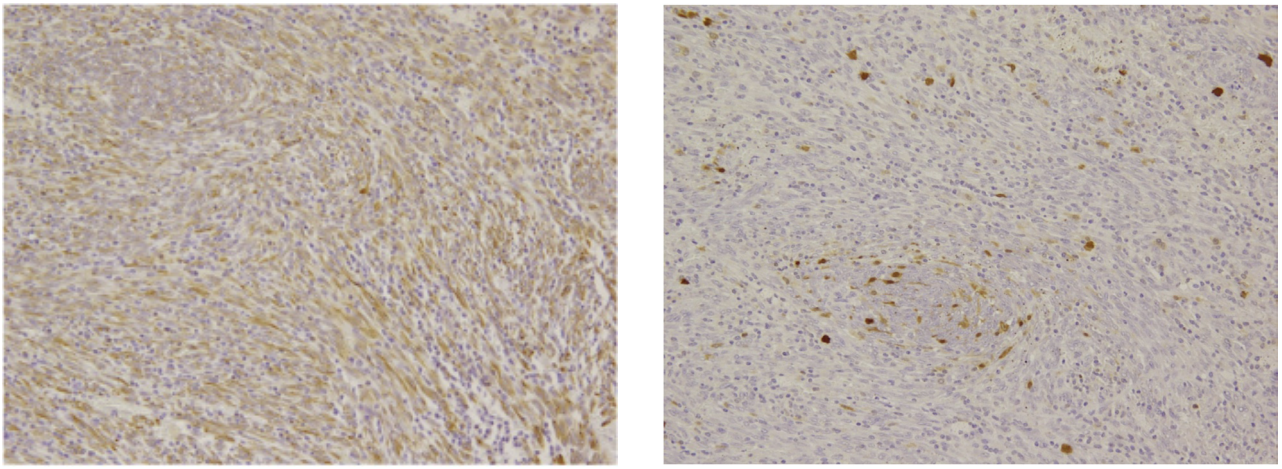

Fig. 5 Immunostaining (× 40). Vimentin positive (left), and S100 positive only for the giant cells (right).

occurs in the limbs and retroperitoneum, 68% and 16% of all cases, respectively [1]. MFH most commonly occurs in adults (peak incidence 61–70 years of age) [1] and juvenile cases are rare [1, 5–7]. The mean age of the patients is 54 years [6].

The occurrence of MFH in the lung is extremely rare, constituting 0.04% of all cases of lung tumor. There are no differences between the sexes. The symptoms of primary pulmonary MFH are chest pain, dyspnea, a cough, hemoptysis, and body weight loss [5, 7]. As is in the current case, some cases are presented without symptoms. The only effective treatment is total resection, and there are only few reports on the effectiveness of chemotherapy or radiotherapy. The rate of local relapse and remote metastasis is high and the prognosis is poor [1, 5, 6, 8], and the 2-year survival rate is 37% [5]. As for primary pulmonary MFH, a lobectomy is accepted as an appropriate surgical procedure. In our case, a lobectomy should have been performed if the definitive diagnosis had been made before the surgery.

A bronchoscopy is the essential preoperative examination for patients with a lung tumor, but was not performed in our case for 3 reasons. First, since the bronchiole on the central side was located outside of the tumor, there was low possibility of good specimen being obtained. Second, since the image of the tumor was clear in the enhanced CT, the tumor was highly likely to be hypervascular, which would raise the risk of bleeding. Finally, the patient rejected the examination by the bronchoscopy. Video-assisted thoracoscopic surgery is the optimal option for patients with an unconfirmed lung tumor, such as this case. Thoracoscopic surgery is a low-invasive technique [9] that enables the removal of a solitary lung tumor together with surrounding normal lung tissue. The prognosis is not poor upon a complete resection of MFH only if they are localized, clearly bordered, noninvasive, and small [6]. The patient has been recurrence-free for 3 years without undergoing a lobectomy, although a careful prolonged follow-up is still required.

CONCLUSION

In a case with primary pulmonary MFH in a young female, a partial lobectomy of the lung was performed using video-assisted thoracoscopic surgery. In spite of the poor prognosis of this disease, no relapse has been observed for the last 3 years since the surgery.

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