Primary pulmonary solitary fibrous tumour with brain metastases

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Abstract

Solitary fibrous tumour (SFT) is a mesenchymal neoplasm of subendothelial origin that can be found in all anatomical locations, but rarely in the lungs. A 71-year old female was referred to our hospital because of the increase in size of a solitary pulmonary mass. Chest contrast-enhanced dynamic computed tomography showed a well-circumscribed lobulated mass measuring 3.1 × 1.6 cm in the posterior segment of the right upper lobe of the lung. Positron emission tomography with 18F-fluorodeoxyglucose (FDG) demonstrated that the mass had high FDG uptake. A right upper lobectomy of the lung and mediastinal lymphadenectomy were performed. The tumour was pathologically diagnosed as an SFT. Seven months later, the patient was found to have brain metastases of the tumour, which led to dizziness. A craniotomy and successive radiosurgery with a gamma knife were performed for the metastatic tumours. She is still alive without evidence of disease 12 months after the treatment of the metastases. Pulmonary SFT seldom behaves aggressively, and only two previous cases of primary pulmonary SFT with brain metastases have been reported. Local therapy including surgery and radiotherapy against metastases from SFT could help improve the survival of such patients.

Keywords: Solitary fibrous tumour • Haemangiopericytoma • Lung • Brain metastases

INTRODUCTION

Solitary fibrous tumour (SFT) is a mesenchymal neoplasm of subendothelial origin that was thought to occur primarily in the pleura at one time, but has since been reported to develop in all anatomical locations. Primary pulmonary SFT is rare, and usually behaves like a benign to low-grade malignant tumour. We report here the case of a patient with a resected primary pulmonary SFT, who developed brain metastases 7 months after the surgery.

CASE REPORT

A 71-year old female was referred to our hospital with a solitary pulmonary mass that had increased in size from the first detection on an annual chest roentgenogram 2 years prior. Chest contrast-enhanced dynamic computed tomography (CT) showed a well-circumscribed lobulated mass measuring 3.1 × 1.6 cm that had a higher attenuation value in the delayed phase than the early phase, with a homogeneous pattern in the posterior segment of the right upper lobe of the lung (Fig. 1A–C). It had measured 2.4 × 0.9 cm in the CT scan 2 years earlier. Positron emission tomography (PET) with 18F-fluorodeoxyglucose (FDG) demonstrated a high FDG uptake (maximum standardized uptake value, 19.5) in the mass. A transbronchial lung biopsy specimen did not reveal a definitive diagnosis.

The intraoperative frozen section of the tumour suggested a malignant neoplasm. A right upper lobectomy and mediastinal lymphadenectomy were therefore performed, resulting in complete resection of the tumour. The surgical specimen showed a yellow-white, well-circumscribed, multinodular tumour, which was measured at 2.7 × 1.7 × 1.5 cm (Fig. 2A). A pathological examination showed that the tumour was composed of closely packed high-grade dysplastic polygonal cells with round nuclei and eosinophilic cytoplasm in a branching stromal vascular ‘haemangiopericytomatosus’ pattern. There were more than 20 mitoses per 10 high-power fields (HPFs), scattered areas of necrosis and sparse desmoplastic reactions (Fig. 2B). No evidence of pleural, vascular or lymphatic invasion, or lymph node metastasis, was found. Immunohistochemically, the tumour cells were positive for CD34 and alpha-smooth muscle actin and negative for bcl-2, S-100, thyroid transcription factor-1 and c-kit. Based on these histologic findings, the tumour was diagnosed as an SFT.

Seven months later, the patient complained of dizziness and had a brain CT and magnetic resonance imaging, which detected two nodules in the cerebellum. One nodule measuring 3.0 cm was located in the right cerebellar hemisphere, and the other measuring 0.9 cm was located in the vermis. A craniotomy was performed for the larger nodule, and the pathological findings of the resected specimen were compatible with metastasis from the pulmonary SFT. One month after craniotomy, radiosurgery with a gamma knife was performed for the remaining nodule. The patient is currently alive without evidence of disease 12 months after the local therapy for the brain metastases.

DISCUSSION

SFT is a rare spindle cell neoplasm arising from the pleura, first described by Klemperer and Rabin in 1931. In 1942, Stout and...
Murray suggested the definition of haemangiopericytoma (HPC) as a neoplasm composed of capillary blood vessels with one or more layers of rounded cells arranged around them. SFT has various histological features, ranging from a heterogeneous, multinodular, partially sclerotic microscopic appearance (fibrous variant of SFT) to a monotonous, highly cellular microscopic appearance (cellular variant of SFT) as was shown in our case. Because the histological characteristics of HPC resemble those of cellular variants of SFT, and there have been no clear criteria to distinguish them, what was called HPC prior to 1990 now tends to be called SFT [1].

Because of the low-frequency occurrence of primary pulmonary SFT, its clinical behaviour has not been well characterized. According to a review of 144 cases of primary pulmonary SFTs by Essola et al. [2], 11 patients (8%) had simultaneous distant metastases at the time of diagnosis. The major sites of those metastases were the lungs, pleura, abdominal organs, retroperitoneum, subcutaneous tissue and bone. To our knowledge, only two previous cases of primary pulmonary SFT with brain metastases have been reported.

The first case reported in 1962 was a 43-year old female who complained of chest pain and haemoptysis [3]. Some months after the lung resection, she suffered from repeated episodes of loss of consciousness, and a brain metastatic tumour in the right cerebellar hemisphere was found. A craniotomy was performed, but she died 30 h later without recovering consciousness. The second case reported in 2003 was a 33-year old patient with a cough and chest pain [2]. Extended pneumonectomy for a 20-cm mass was performed, but a local relapse developed 3 months later. Fifteen months after thoracotomy, three cerebral metastases were found with symptoms of left hemiplegia, and the patient died 7 months after radiation therapy for the brain tumours.

Chick et al. [4] investigated the radiological findings of pulmonary SFTs, and described SFTs as well-circumscribed masses with variable enhancement following intravenous contrast administration on CT, leading to a differential diagnosis including hamartomas and carcinoid tumours. In our case, contrast-enhanced dynamic CT showed a well-circumscribed lobulated mass with homogeneous enhancement. Most SFTs present low FDG uptake on PET, however, the tumour in our case revealed a high uptake of FDG. This is interpreted as being suggestive of malignancy.

In a clinicopathological and immunohistochemical study of 24 cases of pulmonary SFTs, Rao et al. [5] reported that the lesions were divided into low-grade, intermediate-grade and high-grade histologies on the basis of the degree of cytological atypia, cellularity, mitotic activity and areas of necrosis. As in our case, more than 10 mitoses per 10 HPFs, high cellularity, areas of tumour necrosis and nuclear pleomorphism were shown in two high-grade
cases. They also reported that positivity of the tumour cells for CD34 was observed in the majority of cases with usefulness in diagnosis of SFT, and it was compatible in our case.

Pulmonary SFTs sometimes behave like malignant tumours and can metastasize to distant organs. Local therapy comprising surgery and radiotherapy against metastases of SFT may improve the survival of such patients.

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REFERENCES