Giant Solitary Fibrous Tumor of the Pleura Causing Respiratory Insufficiency: Report of 3 Cases

Masaru Abe, MD,1 Hiroaki Nomori, MD,1 Motoji Fukazawa, MD,1 Hiroshi Sugimura, MD,1 Makoto Narita, DDS,2 and Akihiko Takeshi, MD1

We present 3 cases of solitary fibrous tumors (SFTs) occupying entire hemithorax and resulting in respiratory insufficiency. All patients were treated by complete resection, resulting in immediate re-expansion of the lungs and recovery from respiratory insufficiency. Although, two patients remain alive without recurrence, one patient had pleural recurrences three times over a 20-year period, all of which were treated by surgical resection. All of the primary tumors exhibited areas of hypercellularity, hemorrhage, or necrosis. All of the recurrent tumors in the recurrent case displayed large areas of hypercellularity, similar to the part of primary tumor. Although, the MIB-1 index in primary tumors was less than 5%, the index of the recurrent tumors increased up to 11% with repeated recurrence. Giant SFTs usually display hypercellularity, hemorrhage, or necrosis. Tumors with hypercellularity could recur. MIB-1 index could display malignant characteristics of recurrent tumors. Long-term follow-up for more than 10 years after surgery is necessary, particularly for tumors with areas of hypercellularity.

Keywords: solitary fibrous tumor, localized mesothelioma, pleura, recurrence

Introduction

A solitary fibrous tumor (SFT) of the pleura is a rare neoplasm, and giant SFTs occupying the entire hemithorax are even more unusual. Although, some SFTs recur even after complete resection, definitive malignant criteria have not been clarified, making their clinical behavior unpredictable. In this study, we present 3 cases of giant SFTs occupying nearly the entire hemithorax and causing respiratory insufficiency to examine their pathological characteristics. We also reviewed 14 cases of recurrent SFTs to discuss the mechanism of recurrence.

Case Report

Patient 1

A 48-year-old homemaker woman with a mass occupying the entire left thorax who complained of dyspnea and requiring oxygen support was referred to our hospital. She had not undergone a routine medical examination for more than 20 years. The operative findings revealed that the tumor arose from the visceral pleura of the left lower lobe without any invasion or adhesion into the surrounding tissues, which was completely excised. Although, the histological findings revealed spindle tumor cells arranged with varying amounts of collagen, focal myxoid changes, and hyalinization of fibrous tissue, findings that were compatible with SFTs, some areas of hypercellularity and necrosis were observed (Fig. 1A). Mitosis was rarely present. Immunostaining with CD34

1Department of Thoracic Surgery, Kameda Medical Center, Kamogawa, Chiba, Japan
2Department of Clinical Pathology, Kameda Medical Center, Kamogawa, Chiba, Japan

Received: June 27, 2013; Accepted: July 8, 2013

Corresponding author: Hiroaki Nomori, MD. Department of General Thoracic Surgery, Kameda Medical Center, 929 Higashi-cho, Kamogawa, Chiba 296-8602, Japan

Email: hnomori@qk9.so-net.ne.jp

©2014 The Editorial Committee of Annals of Thoracic and Cardiovascular Surgery. All rights reserved.
A tumor 1.5 cm in size was found in the parietal pleura of the left lower thorax, which was near the primary site. The tumor was completely excised, and it histologically displayed large areas of hypercellularity, as observed for the part of primary tumor (Fig. 1B). The mitotic index was 4/10 high power fields (HPFs) in the highest area. Eighteen years after the initial operation, a second recurrence with three tumors up to 3.1 cm in size was found in both the parietal and visceral pleura of the left lower thorax, which were near the primary site. The tumors were completely excised, and they histologically displayed large areas of hypercellularity but no mitoses. Twenty years after the initial operation, the third recurrence occurred, with a 3-cm tumor, along with some other small surrounding tumors, being found at the parietal pleura of the left lower thorax, near the primary site. These tumors were completely excised, and they histologically displayed large areas of hypercellularity. The mitotic index was less than 1/10 HPFs. The MIB-1 index, which was measured according to the method of Martin, et al., was 0.2%, 1.7%, 6.6%, and 11% for the primary tumor, first recurrence, second recurrence, and third recurrence, respectively, indicating that the value increased with repeated recurrence. Twenty-four years after the initial operation, although multiple recurrences occurred in the left thorax (Fig. 1C), the patient is now alive at the age of 71 years without any symptoms, and therefore, she is being followed up without any treatment.

Patient 2

A 64-year-old man who complained of dyspnea for a few days was referred to our hospital under mechanical ventilation because of severe respiratory failure. He had not undergone a routine medical examination for more than 30 years, because he had no regular occupation. CT revealed a mass 24 × 16 cm in size occupying the left thorax and displacing the mediastinum (Fig. 2). Percutaneous needle biopsy findings suggested SFT. Because the tumor extensively adhered to the left upper lobe, it was completely excised together with the left upper lobe. Although, the patient required ventilator support for 10 days after surgery, he was discharged from the hospital 21 days after surgery with sufficient lung re-expansion. Although, the histological findings were compatible with an SFT without invasion to the lungs, areas of hypercellularity and hemorrhage were scattered throughout the tumor. Mitosis was rarely detected, and the MIB-1 index was 4.5%. Currently, 2 years after surgery, the patient is alive without recurrence.
Giant Solitary Fibrous Tumor of Pleura

Patient 3

A 65-year-old man who complained of worsening dyspnea for 10 days was referred to our hospital with oxygen support. He had not undergone a routine medical examination for more than 20 years, because he had no regular occupation. CT revealed a mass 17 × 14 cm in size occupying the entire left thorax and displacing the mediastinum (Fig. 3). Percutaneous needle biopsy suggested SFT. An operation was performed via lower door open thoracotomy, i.e., posterolateral thoracotomy extending along the anterior costal arch. The tumor arose from the visceral pleura of the left lower lobe and adhered to the diaphragm, and it was completely excised. Although, the histological findings were compatible with SFT, the tumor displayed an area of hypercellularity with abounding vascularity and scattered necrosis. Mitosis was rarely present, and the MIB-1 index was 1.9%. After surgery, the lungs re-expanded, allowing the patient to recover from pulmonary insufficiency. He is currently alive without recurrence for 8 months after surgery.

Discussion

Although, it has been reported that SFTs are usually diagnosed in the sixth to seventh decade of life, it is conceivable that tumorigenesis begins much earlier in life because of the slow growing characteristic of this tumor. The three patients described here had giant SFTs occupying nearly the entire hemithorax, and they had not undergone routine medical examinations for more than 20 years; this was likely associated with their tumors being diagnosed in such large sizes. Because surgical resection could permit sufficient re-expansion of the lungs and result in a good prognosis, the differential diagnosis for tumors occupying the entire hemithorax should include SFTs of the pleura to provide an opportunity for surgical resection.

Although, there are no definitive criteria of malignancy for SFT, several reports describe potential malignant criteria to include a large tumor size (more than 5 or 10 cm), a sessile lesion, infiltrative margins, hypercellularity, nuclear pleomorphism, an area of tissue necrosis or hemorrhage, and an increased mitotic index (more than 4 mitoses in 10 HPFs). Although, all tumors in the present 3 cases had no nuclear pleomorphism and exhibited mitotic indexes of less than 4/10 HPFs, they had areas of hypercellularity, hemorrhage, or necrosis, findings that were coincident with the previously reported potential criteria. However, all of the recurrent tumors in Patient 1 exhibited large areas of hypercellularity as observed for the primary tumor, they did not have other malignant findings. We therefore, consider that hypercellularity is the most significant histological malignant finding in SFTs. SFTs exhibiting large areas of hypercellularity could have a risk of recurrence even after complete resection. Although, yearly follow-up is recommended after the resection of malignant SFTs, our experience with Patient 1 indicates that follow-up should continue for more than
Recurrence of SFTs after surgery is known to usually occur in the ipsilateral thorax. We reviewed 14 cases of recurrent SFTs that reported the tumor origin and histological findings. Of the 14 tumors, the primary tumors and the secondary recurrent tumors were ipsilateral in 12 patients and contralateral in 2 patients. Of the 12 patients with ipsilateral recurrences, the sites of the primary and secondary tumors varied (visceral pleura to visceral pleura in 3 cases, parietal pleura to parietal pleura in 3, visceral pleura to parietal pleura in 3, parietal pleura to visceral pleura in 1, and miscellaneous in 4), but in all cases, the sites of recurrences were near the sites of the primary tumor. Patient 1 described in this article also had several recurrences near the primary site. On the basis of its benign pathological characteristics, recurrent SFTs might develop via a mechanism other than dissemination as seen for general malignant tumors. We previously reported a case of SFT consisting of a pedunculated primary tumor originating from the diaphragm and a metastasis located in the parietal pleura, which were in contact with each other. We consider that SFT would have no ability to disseminate to the pleura and pleural metastases of SFTs might occur via contact with a metastasis rather than via dissemination.

While there have been no reports describing the possibility of metachronous independent SFTs, some of the second SFT might occurred from the multicentric origins, because it has usually a slow growing characteristic like bronchioloalveolar carcinomas, which have multicentric origins sometimes. For the cases with second SFTs suspected metachronous independent SFTs rather than metastasis, a genetic analysis for kaptotype would be necessary.

Because a giant tumor occupying the entire hemithorax can possibly be an SFT, an opportunity to perform curative surgical treatment should not be missed, even for patients with severe respiratory insufficiency. During the operation, the intrathoracic cavity should be thoroughly observed, particularly the area near the tumor, to ensure that a neighboring metastasis is not missed. After resection, long-term follow-up for more than 10 years is recommended, especially for tumors displaying hypercellularity.

**Disclosure Statement**

All authors have no conflicts of interest including any financial interests or connections, direct or indirect, or other situations that might raise the question of bias in the work reported or the conclusions, implications, or opinions stated - including pertinent commercial or other sources of funding for the authors or for the associated department or organization, personal relationships, or direct academic competition.

**References**