A 74-year-old asymptomatic female presented with an anterior mediastinal mass incidentally discovered on a routine chest X-ray. Systemic evaluation demonstrated no metastatic lesions. The patient underwent an extended thymectomy via median sternotomy on suspicion of a thymoma. The tumor had arisen from the left half of the thymus without a pedicle and had directly invaded into the left lung and pericardium. The tumor was resected with the entire thymic tissue, and the invaded lung and pericardium were resected en-bloc. The size of the tumor was 5.3×4.0 cm. A disseminated lesion on the mediastinal pleura was also resected. Histopathologically, the lesion mainly consisted of non-atypical spindle-shaped tumor cells in a so-called “patternless pattern” with various densities of collagenous background. Pleomorphism and mitoses were not significant. Immunohistochemical analysis revealed mesenchymal positive markers such as vimentin and CD34. Epithelial markers such as CAM 5.2 and AE1/AE3 were negative. S-100 protein and desmin were not stained. Solitary fibrous tumor of the thymus was diagnosed histologically. Postoperative adjuvant chemotherapy or radiotherapy was not undertaken because the benefits were uncertain. She is well without recurrence 3 months after the operation.

**Key words:** adult, neoplasm, solitary fibrous tumor, thymoma, mediastinum
The patient was diagnosed as having a transient ischemic attack and the symptoms disappeared within a day through conservative treatment. She was discharged on the 19th postoperative day without further disturbances.

SFT of the thymus was diagnosed from the histological examination of the specimen and the results of immunohistochemical study. The lesion mainly consisted of non-atypical spindle-shaped tumor cells in a so-called “patternless pattern” with various densities of collagenous background (Fig. 4). Pleomorphism and mitoses were not significant. There were no necrotic or hemorrhagic lesions in the tumor tissue, and lymphocytic infiltration was not obvious. Direct invasion into the lung was histologically confirmed. However, invasion into the pericardium was not demonstrated.

Immunohistochemical analysis revealed mesenchymal positive markers such as vimentin and CD34 (Fig. 5A). Epithelial markers such as CAM 5.2, AE1/AE3 were negative. Ki-67 (MIB-1) was positive in approximately 15% of the tumor cells (Fig. 5B). S-100 protein and desmin were not stained. Blood cell markers such as leukocyte common antigen (LCA; CD45), and Leu7 (CD57) were
Fig. 4. Histopathological examination (original magnification ×200, hematoxylin-eosin stain) reveals the characteristic “patternless pattern” with non-atypical spindle or round tumor cells in the collagenous background.

Fig. 5. Results of immunohistochemical staining (original magnification ×200).

A: CD34 is positive.
B: Ki-67 is expressed in approximately 15% of the tumor cells.
C: MIC2 protein is positive.
D: Bcl-2 oncoprotein is expressed. Adding to these results, with a lack of S-100 protein and keratin, solitary fibrous tumor was diagnosed.
negative. C-kit protein was not detected. Expression of both MIC2 protein (CD99) and bcl-2 oncoprotein was demonstrated (Fig. 5, C and D).

From these immunohistochemical results, SFT was diagnosed histologically. The resected disseminated lesion was also histopathologically and immunohistochemically confirmed to be the same SFT.

Postoperative adjuvant chemotherapy or radiotherapy was not undertaken because the benefit of this was uncertain. The patient is well without recurrence 3 months after operation.

**Discussion**

SFT was formerly called “localized (or solitary) fibrous mesothelioma,” or “benign (fibrous) mesothelioma,” and had been considered to be a subgroup of mesothelioma. After revision of pleural tumor classification by the World Health Organization in 1999, SFT was classified as an independent entity and was excluded from the mesothelioma subgroups.1)

SFT is an uncommon soft tissue neoplasm that mainly arises from the submesothelial layer of the pleura. In two-thirds of cases, SFT arises from the submesothelial connective tissue of the pleura, however, occurrences arising from various anatomical sites as well as the pleura have been recently reported, such as the meninges, orbit, upper and lower respiratory tract, salivary glands, thyroid, breast, heart, liver, kidney, retroperitoneum, soft tissue, and urogenital organs.

SFT originating in the mediastinum has been rarely reported.2–5) In our case, because the majority of the tumor was in the anterior mediastinum, traced to the thymus and encapsulated with macroscopically intact pleura, we diagnosed that the tumor had originated in the thymic mesenchymal tissue, by gross specimen and histological examination. SFT of pleural origin usually has a pedicle, however, in our case it was absent.4) Because the tumor was large and partially invading into the lung tissue, the exact origin could not be determined and the possibility of a pleural origin could not be excluded. Mediastinal SFT without actual connection to the pleura has been documented.6)

SFT is usually a slow-growing tumor with a favorable prognosis. The patient is usually asymptomatic until the tumor grows large enough to compress neighboring organs. Although the tumor is usually endocrinologically inactive, secretion of insulin-like growth factor II is documented, causing recurrent hypoglycemia.7,8) The clinical behavior is usually benign, however, aggressive cases with local extension and/or recurrence have been reported in the literature.2,9) Pleural dissemination has also been documented.10,11) Complete surgical removal is the best treatment for SFT.

Histopathologically, the “patternless pattern” with non-atypical spindle-shaped tumor cells in the collagenous background is the most characteristic, but not unique, finding in SFT.12) Other spindle cell tumors, such as mesothelioma, hemangiopericytoma, synovial sarcoma, fibrosarcoma, sarcomatoid carcinoma, and malignant peripheral nerve sheath tumor are considerations for differential diagnosis. In particular, hemangiopericytoma is known to have a histopathological and immunohistochemical overlap and it is questionable that it can be separated clearly from SFT.13) In cases with necrosis, hypercellularity, significant mitoses, atypia, and/or pleomorphism the SFT would be considered to have malignant features. However, in such cases, these histological findings do not always correlate with the clinical course.14) In our case, although there were no obvious malignant features revealed in the histopathological examination, the operative findings such as local invasion into the lung and pleural dissemination suggested that the tumor could have potential malignancy might have indicated a poorer prognosis, although the tumor has been resected completely.

Immunohistochemically, the tumor expresses CD34 in 80–100% of the cases and bcl-2 in more than 80%, though it is negative for keratin and S-100 protein.12,14,15) MIC2 (CD99) protein is also known to be expressed normally in the thymic cortex, but positive immunoreactivity for MIC2 in SFT has also been reported.16) Cases with malignant transformation were reported to show diminished CD34 expression and positive p53 immunoreactivity.17) In our case, Ki-67 was positive in approximately 15% of the cellular population. Ki-67 is considered to be an indicator of cell proliferation and was not significantly expressed in this case.

In our case, although malignant features were absent histopathologically and CD34 expression was not diminished, because of the local invasiveness and dissemination, malignant behavior of the tumor is predicted. Careful scrutiny will be needed for long-term postoperative follow-up.

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References