Case Report

J Korean Soc Radiol 2023;84(1):304-310
https://doi.org/10.3348/jksr.2022.0071
eISSN 2951-0805

JOURNAL of THE KOREAN SOCIETY of RADIOLOGY

Extrapleural Malignant Solitary Fibrous Tumor Presenting as Invasive Bilateral Paravertebral Tumors: A Case Report

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Received May 11, 2022
Revised June 3, 2022
Accepted June 18, 2022

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Solitary fibrous tumors (SFTs) commonly arise from the pleura and are mostly benign. However, they may develop anywhere in the body, and 10%–30% are malignant. Classically, SFTs appear as solitary enhancing masses, and bilateral presentation is extremely rare. In this case, an 88-year-old male presented with back pain and a history of chronic tuberculous empyema. Imaging studies revealed bilateral paravertebral masses with aggressive radiologic features, which were speculatively presumed as thoracic malignancies in association with chronic empyema. Herein, we report a unique case of bilateral paravertebral malignant SFTs that were accurately diagnosed with a CT-guided coaxial needle biopsy.

Index terms Solitary Fibrous Tumor; Spinal Neoplasm; Multidetector Computed Tomography; Magnetic Resonance Imaging

INTRODUCTION

Solitary fibrous tumors (SFTs) are well-known tumors of pleural origin, and the majority of tumors are benign; however, they can be found anywhere in the body (1, 2). Typical findings of SFTs on CT and MRI are well-circumscribed enhancing masses adjacent to the broad base of the pleura (3). However, SFTs can rarely appear as bilateral paravertebral tumors with radiologic findings simulating other thoracic malignancies, it is difficult to make an accurate diagnosis on the basis of imaging findings. Herein, we describe a case of malignant SFT presenting as bilateral paravertebral masses and provide a review of previous reports.

CASE REPORT

An 88-year-old male patient visited our hospital due to back pain with the symptom onset of three months ago. He had a history of left pneumonectomy due to pulmonary tuberculosis 30 years ago. He showed no neurologic deficit on physical examination and no abnormal laboratory findings.

The patient underwent contrast-enhanced chest CT. A left paravertebral lobulated mass measuring approximately 8 cm was noted at the T6 level with homogenous enhancement, compromising the left neural foramen and central spinal canal with spinal cord compression (Fig. 1A). Another smaller mass similar to the previous one was noted in the right paravertebral space at the same level, compromising the right neural foramen and central spinal canal. The continuity between the two paravertebral masses was unclear. Associated bony erosions were suspected at the T6 vertebra and left 6th costovertebral junction. A large amount of loculated fluid collection was noted in left hemithorax with partially calcified uneven pleural thickening, suggesting chronic empyema. Enlarged lymph nodes were seen in the both paratracheal and subcarinal areas, and the left para-aortic space of the upper abdomen.

On spinal MRI, bilateral paravertebral masses revealed diffuse T1 low and T2 heterogeneous high signal intensity (SI) with direct invasion of the epidural space at the T6 level (Fig. 1B). Continuity between the two paravertebral masses was not identified. There was bone marrow SI change in the vertebral body of T6 and the lower endplate of T5 with no evidence of bony cortical destruction or widening of the neural foramen. Bone marrow SI change was also noted in the posterior arc of the left 6th rib.

In consideration of unusual CT and MRI findings, advanced age of the patient, and history of chronic empyema, there was a high probability of malignancy, and our primary radiologic diagnosis was malignant lymphoma. Malignant mesothelioma and sarcoma, which are known to have association with chronic empyema, were included in the radiologic differential diagnosis. Multiple lymphadenopathies and suspected metastatic bone lesions supported the probability of malignancy. As the tumors extended through the neural foramen into the spinal canal, a possibility of malignant neurogenic tumors such as malignant schwannoma can be taken into account; however, there was no significant neural foraminal widening on imaging findings.

On subsequent fluorine-18-fluorodeoxyglucose (^{18}F-FDG) PET/CT, the bilateral paravertebral masses revealed intense FDG uptake (maximum standardized uptake value = 14.3, Fig. 1C). There were hypermetabolic bone lesions in T5-7 vertebrae, bilateral 6th and left 7th ribs, and enlarged mediastinal and upper abdominal lymph nodes also revealed high FDG uptake, suggesting metastasis.

A CT-guided coaxial needle biopsy of the left paravertebral mass was successfully performed (Fig. 1D). On microscopic examination of the biopsy specimen, hypercellular spindle cell tumor was observed with moderate to marked nuclear pleomorphism and increased mitotic activity (> 5/10 on high power fields), arranged in a storiform pattern and intratumoral staghorn vascular structure. And those findings were suggestive of malignant SFT (Fig. 1E), Immunohistochemistry revealed positive staining for CD99, and negative staining for CD34 (Fig. 1F). An excisional biopsy with surgical management was planned for the next step but

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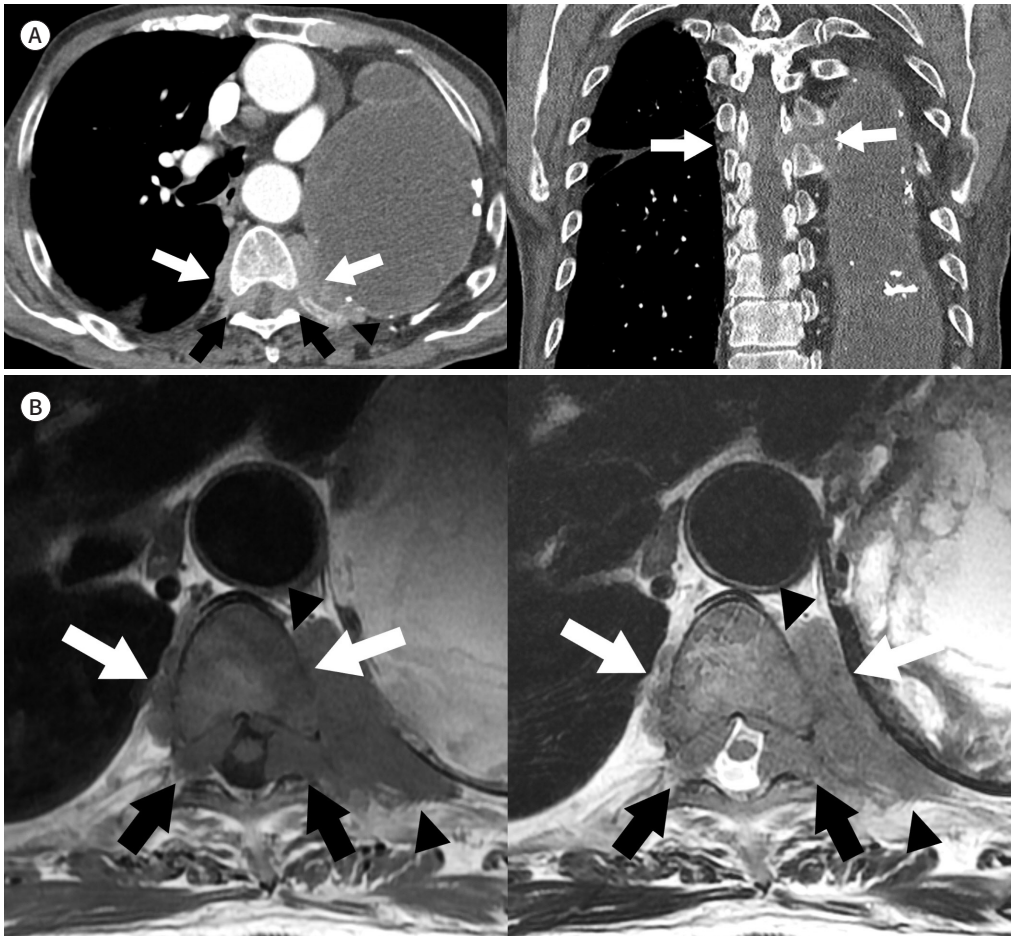
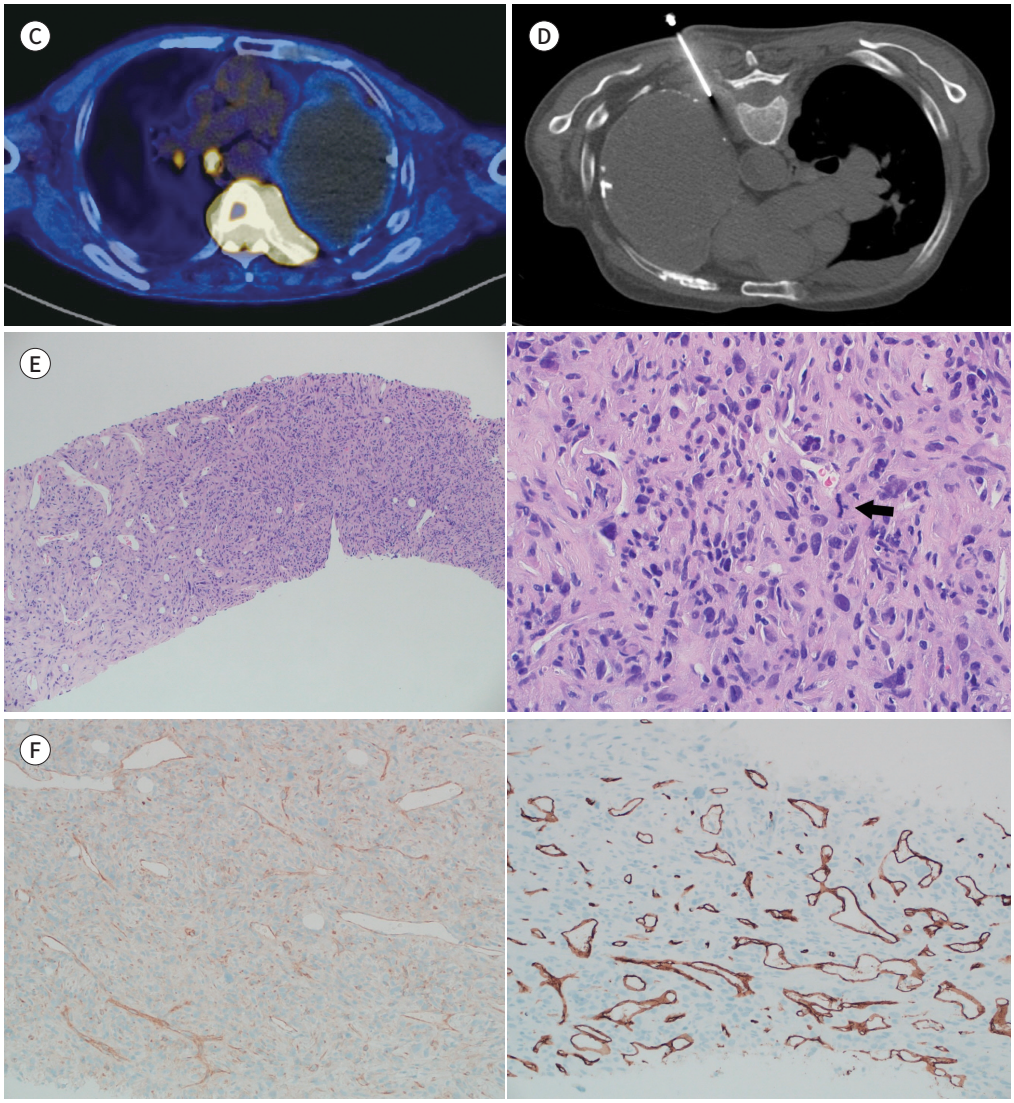
Fig. 1. Extrapleural malignant solitary fibrous tumor of the pleura in a 88-year-old male patient who presented with back pain and had a history of pneumonectomy due to pulmonary tuberculosis.
A. Axial (left) and coronal (right) contrast-enhancing chest CT scans demonstrate infiltrative bilateral paravertebral enhancing masses at T6-7 levels (white arrows) with extension into the central spinal canal. However, there is no neural foraminal widening (black arrows). Bony erosion is seen at the costovertebral junction of the left 6th rib (arrowhead). Left pneumonectomy status is noted with a large expansile fluid collection in the left hemithorax and partially calcified uneven pleural thickening.
B. Axial T1-weighted MR image (left) and T2-weighted MR image (right) display diffuse T1 low and T2 heterogeneous high signal intensity space occupying lesions in both paravertebral aspects at the T6 level (white arrows) with direct invasion into the epidural space of the spinal canal. There is no definite evidence of bony cortical destruction or widening of the neural foramen at the T6 level (black arrows). Combined bone marrow signal intensity changes of the T6 vertebral body and the left posterior arc of the 6th rib are noted (arrowheads).



was not performed due to the deterioration of the patient's general condition. The patient received radiation therapy targeting the T6 vertebra for a month and was followed up at an outpatient clinic. Follow-up imaging studies after the radiation therapy revealed no significant interval change during the period of 4 months. However, unfortunately, the patient suddenly died due to acute duodenal perforation.

This study was approved by our Institutional Review Board, which waived the requirement for informed consent (IRB No. SCHUH 2022-04-004).

Fig. 1. Extrapleural malignant solitary fibrous tumor ofthe pleura in a 88-year-old male patient who presented with back pain and a history of pneumonectomy due to pulmonary tuberculosis (Continued).
C. The paravertebral masses show intense FDG uptake on fluorine-18-FDG PET/CT.
D. CT-guided coaxial needle biopsy was performed for the left paravertebral mass at the T6 level.
E. Microscopic findings show hypercellular proliferation of spindle cells with "staghorn" vascular spaces (left, H&E stain, × 100). Significant pleomorphism and atypical mitosis can be noted (arrow) (right, H&E stain, × 200).
F. Immunohistochemistry shows positive staining for CD99 (left, × 200) and negative for CD34 (right, × 200).
FDG = fluorine deoxyglucose, H&E = hematoxylin & eosin



DISCUSSION

SFTs are rare spindle cell neoplasms of mesenchymal origin. The pleura is a well-known site where SFTs occur frequently. However, previous reports have revealed that SFTs can occur anywhere in the body, from the brain to the bone and soft tissue. Among extrapleural locations, common sites are the extremities, deep soft tissues, and abdominopelvic cavity, however, SFT arising from or around the spine are extremely rare (2, 4-6).

Most of the patients with SFT are asymptomatic at the time of diagnosis, but a clinical manifestation depends on tumor size and location (3, 4). The imaging findings are often nonspecific. On contrast-enhanced CT, small SFTs present as well-defined hyperdense masses, whereas large tumors show more heterogeneous and variable enhancement patterns (4, 7, 8). On MRI, SFTs typically show low-to-intermediate SI on T1-weighted images and heterogeneously low SI on T2-weighted images, which is related to hypocellularity with prominent fibrous tissue (3). On PET/CT, the degree of FDG uptake is similar to that of the mediastinal blood pool; however, large tumors may show heterogeneous low FDG uptake, representing areas of necrosis, cystic degeneration, or calcification. High FDG uptake has not been proven to be a useful predictive parameter for malignant tumors (3, 7).

In one retrospective study of 11 cases of spinal SFTs, spinal SFTs were divided into 4 subtypes according to location: the vertebral (osseous), paravertebral, spinal canal, and mixed type. The vertebral type was the most common subtype, with 4 cases, and the paravertebral type was the least common subtype, with only one case. Due to its rarity and nonspecific imaging findings, the tumor can be easily misdiagnosed. Taking account of anatomical relationships, the dura, leptomeninges, nerve root, and spinal cord may be potential origins of paravertebral SFT (6).

In our case, the tumor appeared as infiltrative bilateral paravertebral masses extending to the central spinal canal. However, there was no bony destruction or neural foraminal widening. In addition, thoracoabdominal lymphadenopathy and metastatic bone lesions were suspected on imaging studies. Given the history and imaging findings of chronic tuberculous empyema in our patient, it is conceivable that the tumors can have potential relationship with chronic empyema. Our initial imaging differential diagnosis included malignant lymphoma, malignant mesothelioma, and sarcoma. The possibility of malignant neurogenic tumors was also considered due to the tumor location and a close approximation to the neural foramen (9). The most unique imaging feature in our case is that the tumors simultaneously presented as bilateral paravertebral masses with no definite continuity between the two masses, which has not been reported in previous literature so far. It can be presumed that the masses grow along the dura mater and there is a possibility of the SFT being originated from the dura or leptomeninges lining the spinal canal. Vertebral (osseous) origin of the tumor is unlikely as considering the epicenter of the paravertebral masses and no bony cortical destruction on imaging findings even though a bone marrow SI change was seen in the vertebral body of T6 and the left 6th rib on spine MRI. Only a few cases of paravertebral SFT have been reported, it is difficult to determine the paravertebral anatomic structures that generate these tumors, or to determine the origin of the tumor by imaging studies alone. Therefore, further studies with correlation of radiologic, surgical, and pathological findings are needed for better evaluation and characterization of this type of tumor.

The histopathological diagnosis of SFT is challenging. Histologically, SFT comprises spindle cells arranged in a “patternless pattern” in a variable collagen stroma with variable cellularity, and occasionally, a storiform or herringbone pattern is seen (2). Malignant SFT is characterized by the presence of infiltrative margins, pleomorphism, hypercellularity, a high mitotic index, and necrosis. Immunohistochemistry is essential for differentiating it from other spindle cell neoplasms. Positivity for CD34, CD99, and bcl-2 is an indicator of SFT (2, 5).

However, CD34 is absent in 5%–10% of SFT cases, and malignant SFT may be negative for CD34, which is caused by dedifferentiation of the tumor and indicates poor prognosis (5, 10). The immunohistochemistry results in our patient may correspond to a case of CD34 negative malignant SFT.

In conclusion, a case of malignant SFT presenting as simultaneous bilateral paravertebral soft tissue masses is extremely rare and can be a considerable diagnostic challenge for the radiologists. We think it is beneficial to know that SFTs can be included in a differential diagnosis of malignant tumors in the paravertebral space. Furthermore, a well-performed CT-guided coaxial needle biopsy can be helpful for accurate histopathologic diagnosis.

Author Contributions

Conceptualization, J.J.; data curation, J.J.; formal analysis, J.J.; investigation, all authors; writing—original draft, all authors; and writing—review & editing, J.J.

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

Funding

None

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침습성 양측 척추주위 종양으로 나타난 흉막외 악성 고립섬유 종양: 증례 보고

정제원[1*] · 진소영[2]

고립성 섬유종은 흉막에서 흔히 발생하는 양성 종양으로 알려져 있으나, 신체 어느 곳에서나 발생할 수 있고 10%–30%에서는 악성이다. 전형적으로, 고립성 섬유종은 단일성의 조영증강되는 종괴로 나타나지만, 척추 주위의 양측성 종괴로 나타나는 경우는 보고되어 있지 않다. 이 증례에서는, 등 통증과 만성 농흉의 병력이 있던 88세 남자 환자의 영상 검사에서 침습적인 양측성 척추 주위 종괴가 발견되었고, 만성 농흉과 연관된 흉부 악성 종양을 먼저 의심하였다. CT 유도하 이중구조 바늘 생검을 통해 진단된 양측성의 척추 주위 악성 고립성 섬유종의 증례를 보고한다.

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