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Two surgical cases of juvenile posterior mediastinal tumors with difficulty in preoperative diagnosis

Kazuyuki Komori, Hiroshi Hashimoto, Kotaro Yoshikawa, Koji Kameda, Shinichi Taguchi, Yuichi Ozeki

ABSTRACT

Introduction: Mediastinal tumors develop in specific locations. Neurogenic tumors appear most frequently in cases of posterior mediastinal tumors, followed by teratomas or benign tumors. Here, we have described two surgically treated cases of juvenile posterior malignant tumors with similar clinical courses of tumor growing and difficulty in preoperative diagnosis.

Case Series: Case 1: A 32-year-old woman presented to our hospital with a right posterior mediastinal tumor detected during a medical checkup on computed tomography. Positron emission tomography-computed tomography and magnetic resonance imaging showed a neurogenic tumor. Tumor resection was performed via video-assisted thoracic surgery for the retrospectively evident growth. Pathological examination revealed a solitary fibrous tumor with immunohistochemical findings (vimentin/B-cell lymphoma 2 protein/STAT6 positivity). The patient was recurrence free at 72 months postoperatively. Case 2: A 32-year-old woman presented to our hospital with a chest abnormality revealed on medical examination. Computed tomography showed that the right posterior mediastinal tumor was a neurogenic tumor. Evident growth on radiograph and abnormal fluorodeoxyglucose accumulations in the spleen and tumor on positron emission tomography-computed tomography were observed; hence, video-assisted thoracic surgery biopsy was performed to differentiate malignant lymphoma. A solitary fibrous tumor was suspected on immunohistochemical examination; the biopsy findings revealed B-cell lymphoma 2 protein/CD34 positivity and vimentin negativity. Subsequently, tumor resection was performed using video-assisted thoracic surgery. Final examination revealed chondrosarcoma based on immunohistochemical findings (CD99/B-cell lymphoma 2 protein positivity and CD34 negativity). The patient was recurrence free at 66 months postoperatively.

Conclusion: Solitary fibrous tumors and chondrosarcomas should be considered in the differential diagnosis of posterior mediastinal tumors. Their diagnosis is difficult for clinical courses, operative findings or even biopsy, and made only on histopathological findings after resection.

Keywords: Chondrosarcoma, Posterior mediastinal tumor, Solitary fibrous tumor, Surgery

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INTRODUCTION

Differential diagnosis of a mediastinal mass can be performed by examining the location of the lesion, its appearance on radiographic studies, and the patient's clinical data, including age, sex, symptomatology, and the results of fine-needle aspiration cytology studies [1]. Most posterior mediastinal masses in young adults represent tumors of nerve cell origin. Here, we have reported two juvenile cases of rare chondrosarcoma and solitary fibrous tumor (SFT) in the posterior mediastinum with similar clinical courses.

CASE SERIES

Case 1

A 32-year-old woman visited our hospital due to an apparently growing abnormal shadow in the chest over 6 years, detected on medical checkup (Figure 1A and B). Computed tomography (CT) showed a tumor measuring 3.5 cm with heterogeneous contrast in the right posterior mediastinum (Figure 1C and D). Positron emission tomography (PET)-CT showed a slightly abnormal uptake (maximum standardized uptake value [SUV_{max}] = 2.87 [60 min]/2.46 [120 min]) in the tumor (Figure 2), and magnetic resonance imaging (MRI) showed a low-signal intensity on T1-weighted imaging, high-signal intensity on T2-weighted imaging, and a heterogenous pattern on gadolinium-enhanced imaging (Figure 3). The tumor was suspected to be a neurogenic tumor.

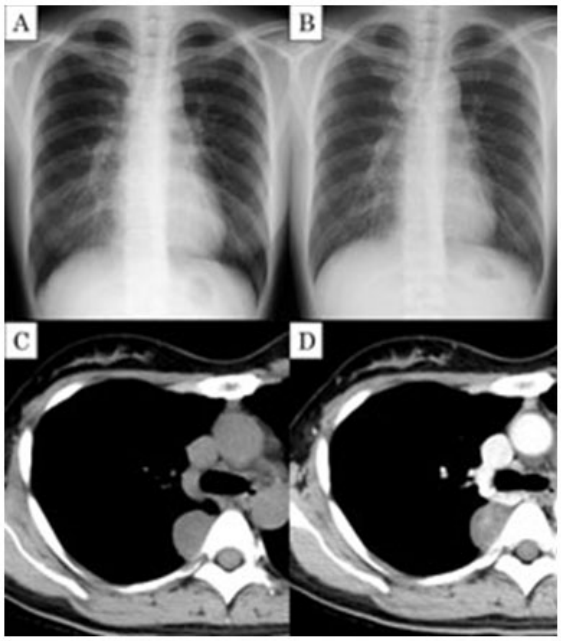

Figure 1: Chest radiography and computed tomography in case 1. (A, B) Abnormal shadows exhibiting apparent growth over 6 years in the right first cardiac arch. (C, D) A well-defined and heterogeneously enhanced tumor (size: 3.5 cm) in the right posterior mediastinum.

Tumor resection was performed via video-assisted thoracic surgery (VATS). The mass was an apparently circumscribed, gray, and myxoid tumor measuring 3.3 × 2.3 × 1.5 cm and had peripheral bleeding. Microscopically, the tumor demonstrated proliferation of oval nuclei and eosinophilic spindle-shaped vesicles, with myxoid stroma and peripheral collagen fibrous deposition (Figure 4). Immunohistochemical analysis showed that the tumor cells were positive for STAT6, vimentin, and B-cell lymphoma 2 protein (bcl-2); slightly positive for CD34; and negative for S-100 protein, desmin, CD31, glial fibrillary acidic protein, neurofilament, and CD99 (Figure 5). The Ki-67 index was 0.8%. These features indicated a benign solitary fibrous tumor (SFT) of the myxoid variant. The patient was followed up with no additional postoperative treatment, and there was no evidence of recurrence at 72 months postoperatively.

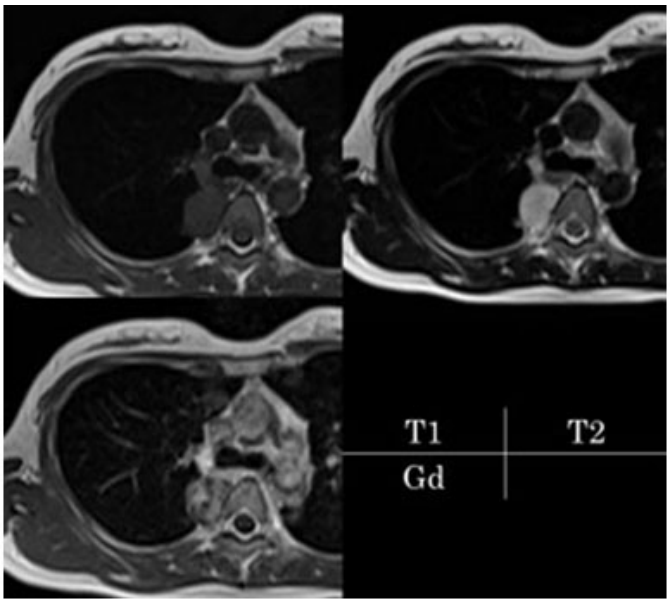

Figure 2: Magnetic resonance imaging in case 1 showing low-signal intensity on T1-weighted imaging, high-signal intensity on T2-weighted imaging, and a heterogenous pattern on gadolinium-enhanced imaging.

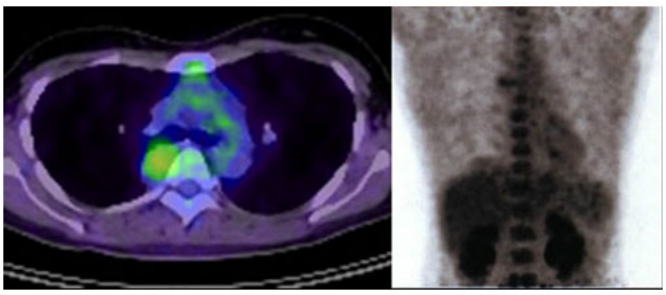

Figure 3: Positron-emission tomography in case 1. Abnormal slight uptake (SUV_{max} = 2.87 [60 min)]/2.46 [120 min]) is seen in the tumor.

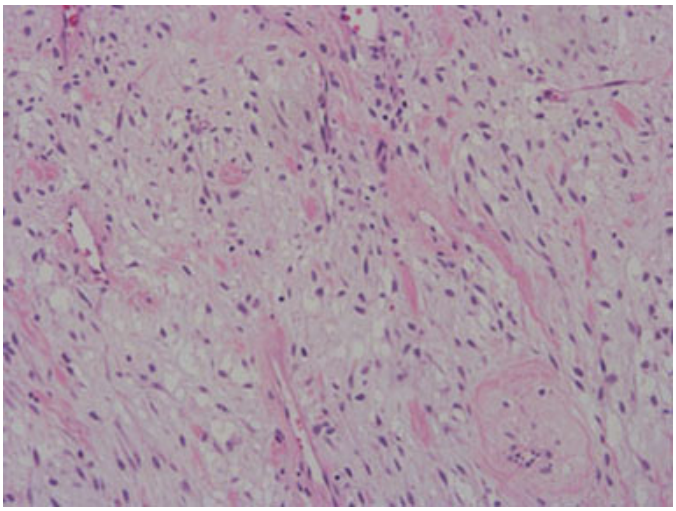

Figure 4: Microscopic findings of case 1. The tumor demonstrates proliferation of oval nuclei and eosinophilic spindle-shaped vesicles with myxoid stroma and peripheral collagen fibrous deposition.

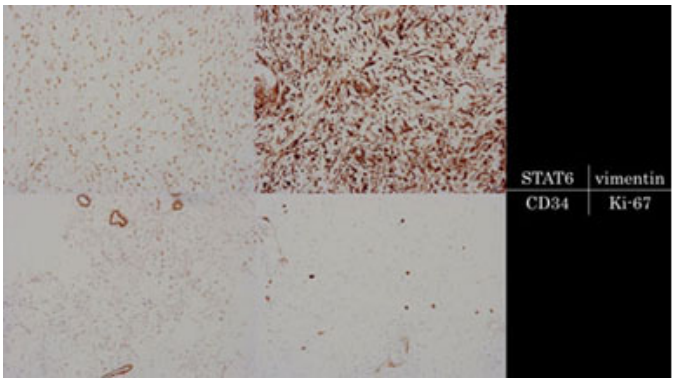

Figure 5: Immunohistochemical findings of case 1. The tumor cells are positive for STAT6, vimentin, and B-cell lymphoma 2 protein; slightly positive for CD34; and negative for S-100 protein, desmin, CD31, glial fibrillary acidic protein, neurofilament, and CD99; Ki-67 index = 0.8%.

Case 2

A 32-year-old woman visited our hospital due to an abnormal shadow in the right lower chest detected on medical checkup roentgenogram. The lesion had apparently grown over 3 years (Figure 6A and B), and CT revealed a tumor measuring 5.0 cm with calcification and heterogeneous contrast in the right posterior mediastinum (Figure 6C and D). MRI showed low-signal intensity on T1-weighted imaging, heterogeneously high-signal intensity on T2-weighted imaging, and contrast enhancement on gadolinium-enhanced imaging (Figure 7). PET-CT showed an abnormal uptake (SUV_{max} = 11.0 [60 min]/15.9 [120 min]) in the tumor and the spleen (Figure 8). The tumor was suspected to be a malignant lymphoma, germ cell tumor, or malignant SFT.

First, a tumor biopsy via VATS was performed. Immunohistochemically, the tumor was positive for bcl-2 and CD34 and negative for cytokeratin, S-100, synaptophysin, vimentin, desmin, and CD31/99. These features were suggestive of a SFT.

Subsequently, tumor resection was performed via VATS. The mass was circumscribed, gray, and solid and contained hemorrhage. Microscopic analysis revealed a small, round cell tumor with an irregular arrangement and large and small cellular density, forming ossification and chondrification (Figure 9). Immunohistochemically, the tumor was positive for CD99 and bcl-2; partially positive for D2-40; slightly positive for S-100; and negative for cytokeratin, CD31/34, calretinin, α-smooth muscle actin, and desmin (Figure 10). The Ki-67 index was 20%. These features indicated extraskeletal mesenchymal chondrosarcoma with small margins.

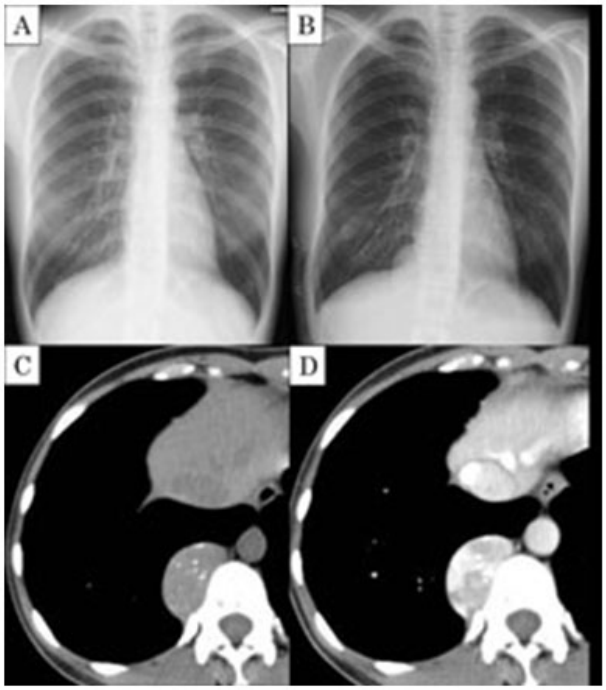

Figure 6: Chest radiography and computed tomography in case 2. (A, B) An abnormal shadow showing apparent growth over 3 years in the right cardiophrenic angle. (C, D) A well-defined and heterogeneously enhanced tumor (size: 5.0 cm) with a scattered calcification in the right posterior mediastinum.

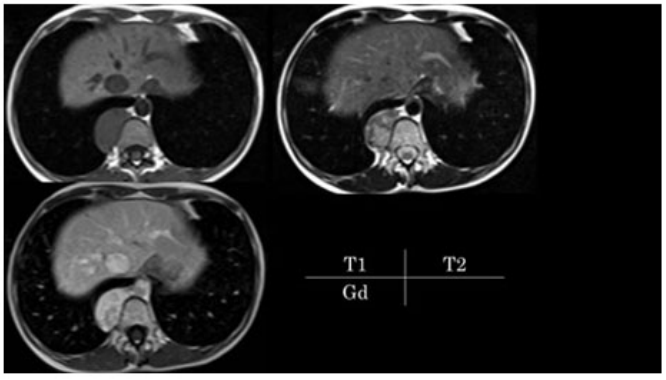

Figure 7: Magnetic resonance imaging in case 2 showing low-signal intensity on T1-weighted imaging, heterogenous intensity on T2-weighted imaging, and a heterogenous pattern on gadolinium-enhanced imaging.

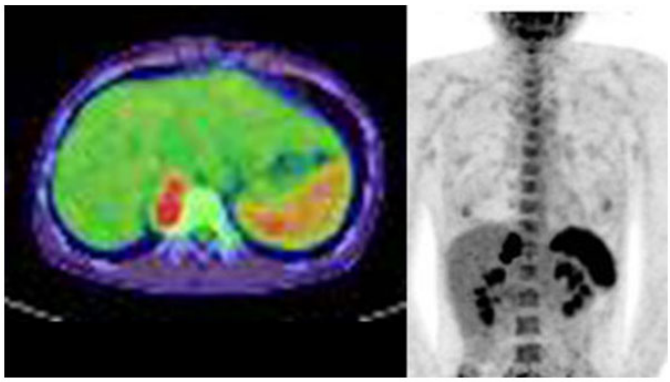

Figure 8: Positron-emission tomography in case 2. Abnormal hyper uptake (SUV_{max} = 11.0 [60 min]/15.9 [120 min]) is seen in the tumor and the spleen.

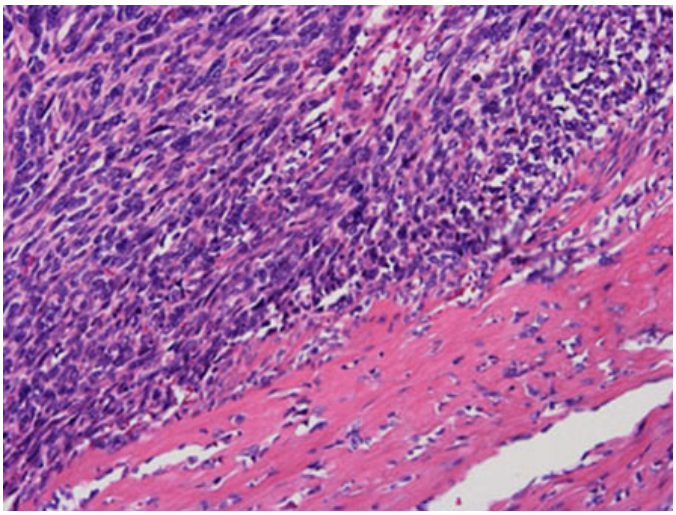

Figure 9: Microscopic findings of case 2. The small round cell tumor demonstrates an irregular arrangement and large and small cellular density with cartilaginous and osseous formation.

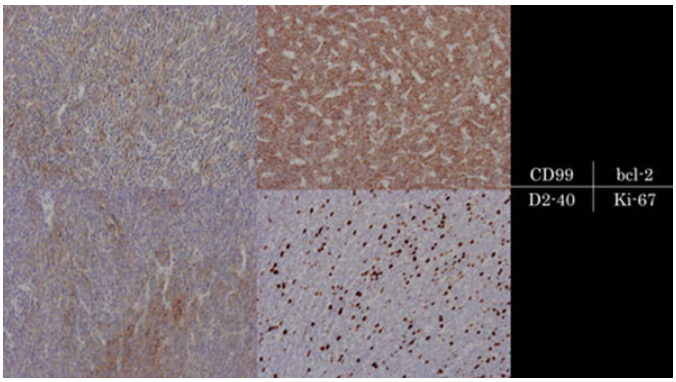

Figure 10: Immunohistochemical findings of case 2. The tumor cells are positive for CD99 and B-cell lymphoma 2 protein; partially positive for D2-40; slightly positive for S100; and negative for cytokeratin, CD31/34, calretinin, α-SMA, and desmin; Ki-67 index = 20%.

The patient was followed up without additional postoperative treatment, and there was no evidence of recurrence at 66 months postoperatively.

DISCUSSION

The posterior mediastinum is the potential space along each side of the vertebral column and the adjacent proximal portion of the ribs [2]. Neurogenic tumors are the most common posterior mediastinal tumors, accounting for 19–39% of all mediastinal tumors and 75% of all posterior mediastinal tumors [3]. Schwannomas and neurofibromas account for >95% of the lesions in these groups [4]. Our patients were initially diagnosed with neurogenic tumors based on their demographics, tumor location, and radiologic findings. In case 2, however, biopsy via VATS was suggestive of a SFT due to CD34 positivity. Therefore, tumor resection was performed via VATS, and the patient was finally diagnosed with mesenchymal chondrosarcoma.

Solitary fibrous tumor is a rare type of soft tissue tumor [5] that was first described by Klemperer and Rabin in 1931 [6]. It was not until 1979 that Scharifker and Kaneko showed that it originated in the subserosal mesenchymal tissue [7]. According to the 2002 World Health Organization classification, SFT is defined as a mesenchymal tumor that may have fibroblastic characteristics and clear peripheral vascular tumor-like branching vessels [8]. It is difficult to ascertain the true incidence and prevalence of SFTs because most patients are asymptomatic [9]. Solitary fibrous tumor are well-defined, homogenously hyperdense masses that form obtuse angles with the pleura on CT [10]. Mediastinal SFTs, such as malignant tumors of the pleura, may show invasive features and have a higher recurrence rate, making them difficult to differentiate from malignant mesothelioma, lymphoma, sarcoma, neurogenic tumors, or thymic neoplasms [11]. Therefore, immunohistochemical analysis plays an important role in the diagnosis. Approximately 70–80% of benign SFTs of the thorax are positive for CD34, a marker for normal endothelium and vascular tumors. Similarly, most benign SFTs are positive for bcl-2, a marker of terminal differentiation, and vimentin. Cytokeratin, S-100, and p53 proteins are upregulated in malignant SFTs [12]. In addition, STAT6 was detected as a diagnostic marker for SFTs in 2013 [13]. In case 1, the tumor cells were positive for STAT6 and vimentin, but they were slightly positive for CD34, and the Ki-67 index was 0.8%, leading to a diagnosis of a benign SFT of the myxoid variant.

Chondrosarcoma is a malignant tumor that produces a cartilage matrix and is the third most common primary malignant tumor of the bone after myeloma and osteosarcoma, constituting 20–27% of all primary malignant osseous neoplasms [14, 15]. Extraskeletal mesenchymal chondrosarcomas (EMCS) are relatively rare neoplasms, representing approximately 1% of chondrosarcomas [16, 17]. The first case of EMCS was described in 1964 [18]. Chondrosarcomas grow with lobular type architecture, and these hyaline cartilage nodules demonstrate high water content and peripheral enchondral ossification, the so called “ring-and-arc”

findings. Imaging features, such as low attenuation on CT and very high signal intensity on T2-weighted MRI, directly reflect this pathological appearance [14]. Microscopically, a characteristic biphasic pattern comprising sheets of undifferentiated round or spindled mesenchymal cells interspersed with islands of well-differentiated hyaline cartilage is observed [17]. Immunohistochemically, similar to other chondrosarcomas, the cartilaginous component is strongly positive for S-100 protein, and undifferentiated cells show scant patchy positivity. Similar to other round cell tumors, the latter cells are positive for CD99, neuron-specific enolase, and Leu-7 [17]. Sox9 is a recently described marker in both cartilage and the cells [19]. In case 2, the presence of a typical biphasic pattern of undifferentiated small round cells blended with islands of hyaline cartilage was observed. Immunohistochemically, undifferentiated cells were partially positive for CD99. Surgery followed by adjuvant chemotherapy is the mainstay of the treatment [17, 20]; however, most tumors (18/20 cases) originate in the bone rather than in the soft tissue. In case 2, the patient was followed up without adjuvant therapy, and there was no evidence of recurrence at 66 months postoperatively.

The two cases described here had a similar clinical course; both cases pertained to young women with tumors located in the posterior mediastinum, which grew in a relatively long period of time. Larger reports or multicenter studies may improve our knowledge of these tumors in the posterior mediastinum.

CONCLUSION

In summary, we have reported two rare cases of posterior mediastinal tumors that had similar clinical courses, but distinct histopathological findings. Our report suggests that SFTs and chondrosarcomas should be considered in the differential diagnosis of posterior mediastinal tumors.

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Author Contributions

Kazuyuki Komori – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Hiroshi Hashimoto – Conception of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

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Conflict of Interest

Authors declare no conflict of interest.

Data Availability

All relevant data are within the paper and its Supporting Information files.

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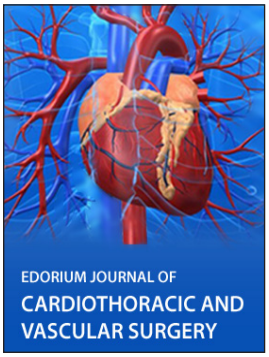

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