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Complete Resection of Mediastinal Undifferentiated Pleomorphic Sarcoma

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A 70-year-old man was referred to our hospital for mediastinal malignant neoplasm. Chest computed tomography revealed a 20-cm mass in the middle mediastinum with displacement of the superior vena cava, left innominate vein, and right upper lobe. Tumor resection, right upper lobectomy, combined resection of the left brachiocephalic vein and superior vena cava, and revascularization were performed. It was pathologically diagnosed as an undifferentiated pleomorphic sarcoma, and the patient experienced no recurrence for 4 years. Although the prognosis of mediastinal pleomorphic sarcoma cases is poor, complete surgical resection may lead to long-term survival.

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Undifferentiated pleomorphic sarcoma (UPS), formerly called malignant fibrous histiocytoma, is an aggressive type of soft tissue sarcoma.[1] Owing to limited efficacy of the current treatment, UPS has a poor prognosis, with a 5-year overall survival rate of 30% to 50%. However, because of its rare situation, these reports did not include mediastinum lesions.[1] The prognosis of malignant fibrous histiocytoma/UPS of mediastinal origin is even worse, with a half-year survival rate of 57%.[2] Herein, we present a rare case of 4-year disease-free survival after right upper lobectomy and tumor and superior vena cava resection with revascularization for mediastinal UPS.

A 70-year-old male smoker with an 80-pack-year history presented to our hospital for further workup after an endobronchial ultrasound-guided transbronchial needle aspiration diagnosed a mediastinal malignant neoplasm. His chief complaint was right-sided back pain. His medical history included hypothyroidism and hypertension.

Chest radiography revealed a mass shadow and reticular shadows in the right upper lung field, and chest computed tomography (CT) revealed a 20-cm mass in the middle mediastinum with displacement of the superior vena cava and left innominate vein and obstructive pneumonia in the right upper lobe (Figure 1). A positron emission tomography/CT scan revealed a tumor with a maximum standard uptake value of 10.3, indicating fluorodeoxyglucose and 2-[fluorine-18] fluoro-1-deoxy-D-glucose accumulation. Blood tests showed that the soluble interleukin 2 receptor level was slightly high (632 pg/mL), although there were no abnormalities in the acetylcholine receptor antibody and any other tumor marker, such as carcinoembryonic antigen, squamous cell carcinoma antigen, cytokeratin 19 fragment, and pro-gastrin-releasing peptide.

Because tumor invasion of the superior vena cava and right upper lobe was suspected in addition to the difficulty of dissection between tumor and surrounding organs such as the aorta, the pericardium, the right lung, and the trachea by single approach, a combined resection of the superior vena cava and right upper lobe by the median sternotomy approach and posterolateral approach was planned. First, the patient was placed in the supine position. The tumor was dissected from the aorta. To avoid total clamping during reconstruction of the superior vena cava, the bypass between the left brachiocephalic vein and the auricle of the right atrium with an 8-mm

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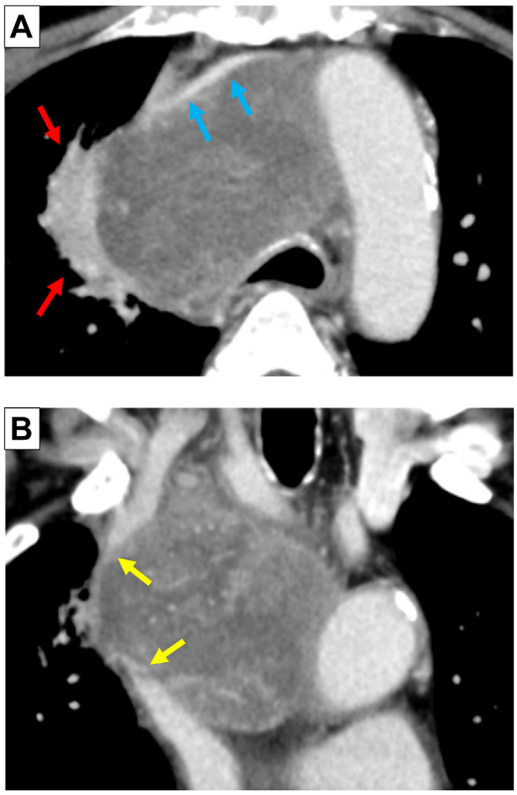

FIGURE 1 Representative images of radiologic findings. (A) Axial and (B) sagittal computed tomography images showing a 20-cm mass with displacement of the superior vena cava, left innominate vein, and right upper lobe. The red, blue, and yellow arrows indicate the right upper lobe, left innominate vein, and superior vena cava, respectively.

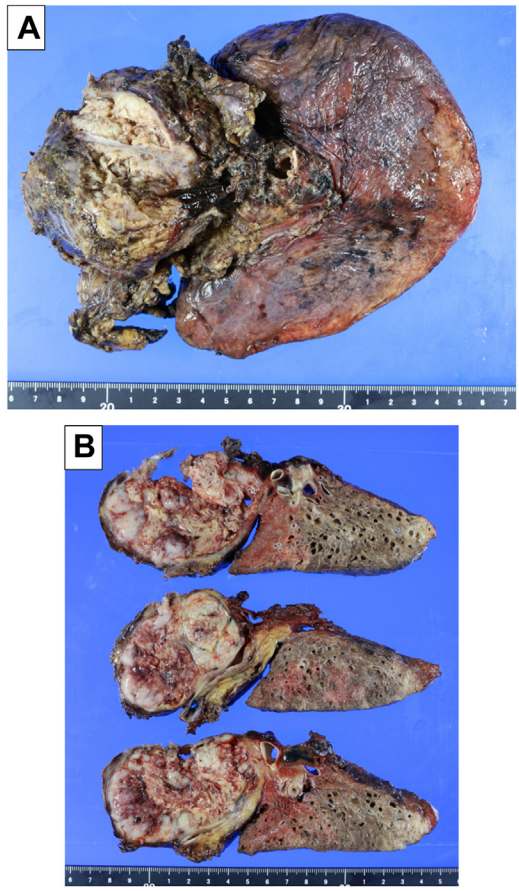

FIGURE 2 Representative macroscopic images of pathologic findings. (A) Overall image. (B) Sectioning image.

ringed polytetrafluoroethylene tube was constructed following the median sternotomy approach. Next, the patient was placed in the right lateral decubitus position. A right upper lobectomy, combined resection of the superior vena cava, and reconstruction of the right brachiocephalic vein-superior vena cava bypass with a 12-mm ringed polytetrafluoroethylene tube were performed through a posterolateral incision. Pathologic findings revealed a diagnosis of anaplastic polymorphous sarcoma (Figures 2, 3). The immunohistochemical assays revealed positive staining for vimentin; however, they showed negative staining for AE-1/3, CAM 5.2, EMA, LCA, synaptophysin, chromogranin A, SALL4, S-100, HMB-45, D2-40, TTF-1, MDM2, CDK-4, NUT, CD34, desmon, SMA, Bcl-2, CD30, c-kit, AFP, HCG-b, and glypican3. In addition, no SMARCA4 or INI1 deficiency was observed. Therefore, pathologic examination confirmed a mediastinal UPS.

Owing to a thrombus in the left brachiocephalic vein graft, edoxaban was administered on postoperative day 10, and the patient was discharged on postoperative day 18. Follow-up enhanced CT revealed thrombus in the graft of the left brachiocephalic vein and perfusion in the graft of the right brachiocephalic vein. Therefore, edoxaban was discontinued 1 month after the operation. Follow-up chest CT performed 4 years after the surgical resection did not show evidence of recurrence, and his general condition was good.

COMMENT

Accurate diagnosis of mediastinal UPS requires exclusion of other tumors through pathologic examination as UPS itself does not have distinct morphologic characteristics. During resection, the specimen should undergo extensive sampling to identify lipoblasts, osteoid matrix, and well-differentiated liposarcoma components.

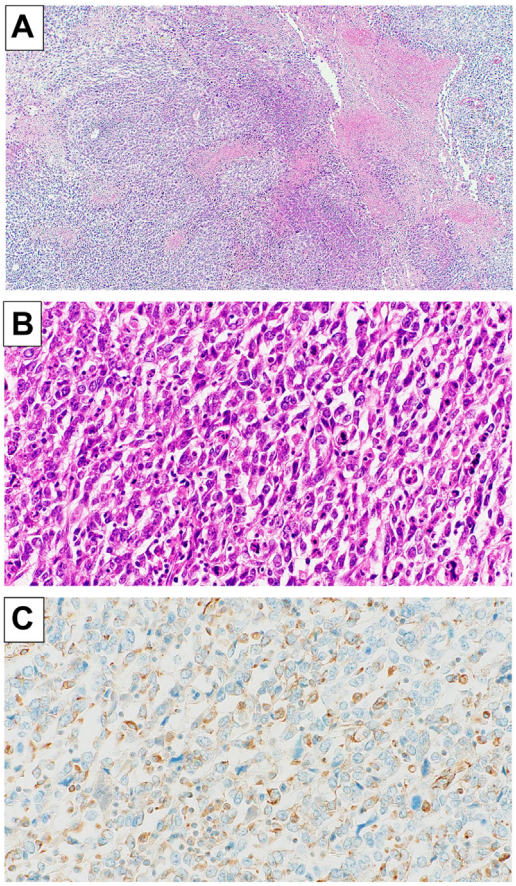

FIGURE 3 Representative images of pathologic findings. The pleomorphic cells were giant or multinuclear cells with large cell sizes, coarse nuclear chromatins, and indistinct nucleoli (hematoxylin and eosin staining: (A), magnification ×10; (B), magnification ×40). On immunohistochemical evaluation, the tumor cells were positive for vimentin (C) but negative for other markers.

Furthermore, the immunohistochemical profile of UPS is nonspecific; therefore, an immunohistochemical analysis should be performed to exclude metastatic sarcomatoid carcinoma, melanoma, lymphoma, nuclear protein of the testis carcinoma, pleomorphic myogenic sarcomas, pleomorphic rhabdomyosarcoma, and dedifferentiated liposarcoma.[3] In this case, these were excluded; therefore, his condition was finally diagnosed as a mediastinal UPS.

Complete surgical resection with a negative margin is the preferred treatment strategy because these tumors are insensitive to chemotherapy or radiotherapy. However, complete resection is more difficult in cases of mediastinal primary lesions than in cases of extremity lesions. We particularly struggled with mediastinal UPS invading the pericardium, heart, great vessels, and lungs, which can cause surgical difficulties or cardiopulmonary failure.[4] Available case studies suggest that patients with mediastinal UPS typically fall within the age range of 50 to 82 years, and the male to female ratio is 1:1. Three of the 6 patients were ineligible for aggressive treatment and unfortunately died within 1 week to 1 month of diagnosis. Surgical treatment was performed in 2 cases excluding this case.[4-8] Okuda and coworkers[5] reported the case of a patient who suffered from a 14.5-cm mediastinal pleomorphic sarcoma and underwent extended surgical resection. However, the patient had locoregional recurrence 1 month after surgical resection, and the patient died 4 months after the second operation for tumor recurrence. In the other case, the tumor was relatively small and did not invade the surrounding area, and the patient had 6 months of disease-free survival after tumor resection.[4] In this case, CT revealed a large mass with a maximum diameter of 20 cm in the middle mediastinum. Although extended resection was necessary in this case as in the case reported by Okuda and coworkers,[5] the patient had a good clinical outcome owing to complete resection. We believe that complete surgical resection plays an important role in the treatment of pleomorphic sarcoma.

In conclusion, we report the case of a successful surgical tumor resection for mediastinal UPS, resulting in a favorable clinical outcome and long-term prognosis. Despite a poor prognosis for mediastinal pleomorphic sarcoma, complete surgical resection can offer long-term survival.

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PATIENT CONSENT

Written informed consent was obtained from the patient for publication of this case report and the patient's anonymity was preserved.

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