Surgery of Thymic Tumor with Persistent Left Superior Vena Cava

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Because a persistent left superior vena cava (PLSVC) is a rare congenital malformation in the thoracic venous system, surgery of the thymus in such patients has rarely been reported. We herein present a case involving a 68-year-old woman who was treated for a thymic tumor adhering to a PLSVC. She underwent complete resection of the thymic tumor through median sternotomy with left-sided video-assisted thoracic surgery. Although the tumor was close to the left phrenic nerve, this nerve was safely preserved. The pathological diagnosis was mucosa-associated lymphoid tissue (MALT) lymphoma of the thymus. Thoracoscopy combined with median sternotomy enabled us to perform a safe surgical procedure for this patient.

Keywords: thymus neoplasm, thoracic surgery, persistent left superior vena cava, vessel anomaly

Introduction

In thoracic surgery, surgeons often encounter vessel abnormalities that make the surgical procedure much more challenging. A persistent left superior vena cava (PLSVC) is one of these thoracic vessel anomalies.

PLSVC is a rare congenital thoracic venous malformation that occurs in approximately 0.3%–0.5% of the general population.1 This venous anomaly originates from failure of closure of the left anterior cardinal vein during cardiac development.2 The PLSVC usually drains into the coronary sinus with no hemodynamic effects.

In most cases, it is asymptomatic and detected as an incidental finding.

Although surgical treatment of lung cancer in patients with a PLSVC has been reported,3,4 few reports have described surgical treatment of thymic tumors in patients with a PLSVC. In addition, surgical treatment would be challenging if the thymic tumor were adhered to the PLSVC. We herein report the first case of successful complete resection of a thymic tumor adhered to a PLSVC.

Case Report

A 68-year-old woman with no medical history presented to our hospital because of an abnormal chest radiograph. She had no complaints. Laboratory data showed that her soluble interleukin-2 receptor concentration was 168 U/mL, which was within the reference range. The patient was positive for anti-SS-A antibody but negative for anti-SS-B antibody. She was also negative for anti-acetylcholine receptor antibody. We did not check her serum immunoglobulin level before surgery. All other laboratory data were within the reference ranges. Chest computed tomography revealed an irregularly shaped, 42-mm-diameter anterior mediastinal mass with multiple cystic changes, which raised suspicion for...
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A thymic tumor (Fig. 1A). On a T2-weighted magnetic resonance axial image, the ventral part of the tumor was hypointense and the dorsal part of the tumor was hyperintense, suggesting that the tumor was composed of a solid and cystic component (Fig. 1B). Preoperative positron emission tomography showed abnormal uptake in the mediastinal mass, which suggested a malignant tumor. Biopsy was not performed. No detectable metastases were present in other organs.

Chest computed tomography also showed that the left brachiocephalic vein passed laterally on the left side of the aorta (white arrow). The patient also had a bridging innominate vein (black arrow). (D) The lateral view of the three-dimensional CT image showed the vein flowing into the coronary sinus (arrow). The vein was the persistent left superior vena cava.

Fig. 1  (A) Chest computed tomography (CT) showed an anterior mediastinal mass (arrow) on the persistent left superior vena cava (arrowhead). (B) A T2-weighted magnetic resonance axial image showed a mass lesion comprising a hyperintense area (arrow) and hypointense area (arrowhead). (C) Three-dimensional reconstructed CT showed that the left brachiocephalic vein passed laterally on the left side of the aorta (white arrow). The patient also had a bridging innominate vein (black arrow). (D) The lateral view of the three-dimensional CT image showed the vein flowing into the coronary sinus (arrow). The vein was the persistent left superior vena cava.

Because we suspected that she had a thymoma, we conducted an extended thymectomy through median sternotomy with the aid of left-sided video-assisted thoracic

Fig. 2  The intraoperative view revealed the persistent left superior vena cava passing on the caudal side in front of the descending aorta (white arrow) and the left phrenic nerve descending laterally toward the persistent left superior vena cava (black arrow). The thymic tumor was adhered to the phrenic nerve; however, the tumor did not invade the nerve.
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surgery. The intraoperative findings showed that the tumor was adherent to the PLSVC but had not invaded it (Fig. 2). To ensure safe completion of the surgical procedure, we placed two thoracic ports in the left fifth intercostal space. With the aid of thoracoscopy, we dissected the adhesion between the tumor and PLSVC. We also identified the left phrenic nerve passing lateral to the PLSVC (Fig. 2) and safely preserved the phrenic nerve. We cut the bridging innominate vein because it was involved in the tumor. Thymic veins were not identified because they were too thin to visualize. We successfully achieved complete resection of the tumor. The patient’s postoperative course was uneventful, and she was discharged without left phrenic nerve palsy or other complications.

Pathological examination of the resected specimen revealed mucosa-associated lymphoid tissue (MALT) lymphoma of the thymus. The resected mass was 28 × 13 mm. Macroscopically, the tumor was solid with multiple cystic lesions (Fig. 3A). A histopathological examination revealed lymphoepithelial lesions formed by the infiltration of atypical lymphoid cells and expansion of Hassall’s corpuscles (Fig. 3B). On immunochemistry, the thymic epithelial cells were positive for CD 20 (Fig. 3C). The thymic epithelial cells also demonstrated positive staining with anti-cytokeratin antibody (Fig. 3D). The cystic lesions were confirmed to be thymic cysts, which are characteristic findings in MALT lymphoma. The tumor did not microscopically invade the bridging innominate vein or PLSVC.

The patient developed no recurrence for 6 months after surgery. She underwent no additional therapy because the oncologists did not recommend chemotherapy. Postoperative examination did not reveal *Helicobacter pylori* infection. The postoperative laboratory data revealed an elevation of the serum immunoglobulin A (IgA) concentration (1984 mg/dL), which was carefully followed up by the oncologists. Although the patient was positive for anti-SS-A antibody, the rheumatologists did not diagnose her with Sjogren’s syndrome because she was clinically asymptomatic.

**Discussion**

We have herein reported a case of successful resection of a thymic tumor that had extended to the PLSVC.
Previous reports have shown that a PLSVC may pose difficulties not only in performing central venous catheterization or intracardiac electrode placement, but also in conducting thoracic surgery. However, surgery for treatment of thymic tumors in patients with a PLSVC has not been previously reported; this is the first such case report.

In this rare case, we needed to obtain a detailed understanding of the patient’s anatomy to ensure safe performance of the surgical procedure. The left phrenic nerve was particularly important for this surgery. Sparing the phrenic nerve has great significance in thymectomy. Because the tumor had spread to the PLSVC and we had feared that we might misunderstand the anatomy of the left phrenic nerve, we used thoracoscopy to safely perform the thymectomy. Thoracoscopy via the intercostal spaces provided us an overview of the PLSVC and left phrenic nerve. Thoracoscopic surgery also enabled us to preserve the left phrenic nerve. Indeed, left phrenic nerve palsy did not develop after surgery.

The preoperative diagnosis of a PLSVC is of importance. classified bilateral superior vena cava into five groups. The current patient had bilateral superior vena cava with an intercommunicating vein. The intercommunicating vein was hemodynamically insignificant, and this is why we cut the bridging vein during surgery. The left superior vena cava must be differentiated from supracardiac total anomalous pulmonary venous return because the latter requires specific treatment. Enhanced chest computed tomography or magnetic resonance imaging would be useful for differential diagnosis of the anomaly of bilateral superior vena cava.

Treatment of MALT lymphoma of the thymus requires complete surgical resection. Thymic MALT lymphoma is a rare type of lymphoma that develops in the thymus, and it was first reported in 1990. Because of the rarity of thymic MALT lymphoma, its treatment has not yet been optimized. However, previous reports have supported the effectiveness of surgical treatment. Long-term survival of patients with thymic MALT lymphoma after complete surgical resection without adjuvant chemotherapy or radiotherapy has been reported. Because the oncologists did not recommend chemotherapy in the present case, the patient did not receive adjuvant chemotherapy. She was still alive without recurrence at the time of this writing (6 months postoperatively). We believe that the radical resection described herein led to the patient’s favorable prognosis.

Conclusion

In summary, a patient with thymic MALT lymphoma adhered to a PLSVC was successfully treated by thymectomy. Thymectomy with complete resection of the tumor is a promising treatment for patients with thymic MALT lymphoma. In addition, thoracoscopy was useful to observe the detailed anatomy of the PLSVC and thus greatly contributed to safe surgical removal of the thymic tumor in this patient with a PLSVC.

Disclosure Statement

The authors declare that no grants, financial support, or technical or other assistance was received. There is no conflict of interest related to this study.

References