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Case report

Surgical resection of a thymoma developed in a case with isolated persistent left superior vena cava

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ABSTRACT

Introduction: Persistent left superior vena cava (PLSVC) is one of the most common vascular abnormalities in the chest. In approximately 10 % of cases, the right superior vena cava is missing, which is called isolated persistent left superior vena cava (IPLSVC).

Presentation of case: The case is an 85 years-old female. An anterior mediastinal tumor was accidentally revealed when the patient was admitted after a traffic accident. As the tumor became larger within four months, a thymectomy was planned. The anterior mediastinal tumor was in front of the ascending aorta, which was close to the confluence of the left and right brachiocephalic veins in normal anatomy. However, in this case, the right superior vena cava was missing, and the right brachiocephalic vein flowed into the left superior vena cava by the chest computed tomography. Preoperative examinations found no accompanying cardiac abnormality. Robot-assisted thymectomy was performed. No tumor infiltration was observed in the right brachiocephalic vein. No abnormality was found in either phrenic nerve. The tumor could be safely resected, and her postoperative course was uneventful. The pathological diagnosis was a thymoma.

Discussion: A case of thymectomy with IPLSVC is quite rare. A careful observation of the preoperative computed tomography images helps to diagnose IPLSVC. Technically, thymectomy was not much different from normal, other than the reversed location of the veins. However, it should be noted that IPLSVC cases may have cardiac malformations.

Conclusion: Thymectomy for thymoma with IPLSVC can be safely performed when the left and right veins are reversed.

1. Introduction

IPLSVC is a venous anatomical abnormality found in 0.3–0.7 % of the general population [2,3]. It is considered to be the most common venous abnormality in the chest and is often found in combination with other cardiac malformations. Many cases have been reported in which this disease was discovered during cardiac catheterization or insertion of a pacemaker [2,4,5]. However, a case of thymoma with IPLSVC is quite rare.

2. Case

The case is an 85-year-old female nonsmoker with hypertension and diabetes mellitus who was involved in a traffic accident. A chest computed tomography image showed multiple right rib fractures and lung contusion. A 20 mm anterior mediastinal tumor was found incidentally. The tumor was solid with an irregular surface, but invasion seemed absent. The tumor was suspected to be a thymoma. She had no family history of the disease and wasn't taking any medications other than antihypertensives and oral diabetes medications. Because the patient was senile, course observation in the outpatient department was selected. However, the tumor enlarged rapidly to 34 mm in four months. Therefore, surgical resection of the tumor was planned.

The tumor was in front of the ascending aorta where the left and right brachiocephalic veins meet in normal anatomy (Fig. 1A). However, in this case, the right superior vena cava was missing. The right brachiocephalic vein ran from the right to the left, crossing the anterior of the aortic arch. It drained into the isolated left persistent superior vena cava (IPLSVC) (Fig. 2A). The IPLSVC was connected to the vein of Marshall and entered the pericardium (Fig. 1B and C). The vein of

Abbreviations: IPLSVC, isolated persistent left superior vena cava; PLSVC, persistent left superior vena cava; SVC, superior vena cava.

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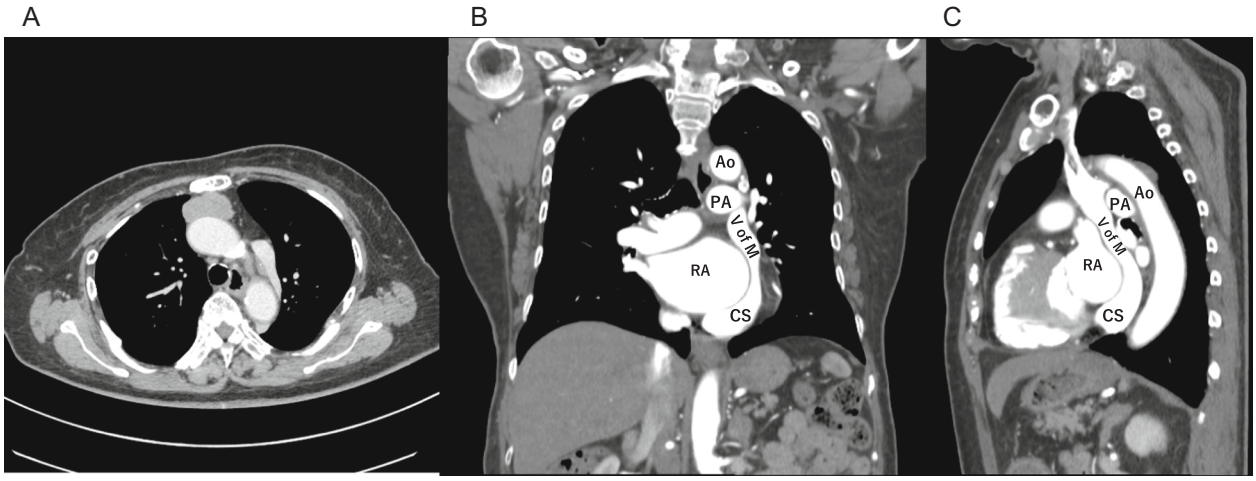

Fig. 1. A) Computed tomography image of the thymoma in front of the ascending aorta, B) a coronal image of the IPLSVC, and C) a sagittal image of the IPLSVC. The coronary sinus was connected to the right atrium. Ao: aorta, CS: coronary sinus, IPLSVC: isolated pulmonary left superior vena cava, PA: pulmonary artery, RA: right atrium, v of M: vein of Marshall.

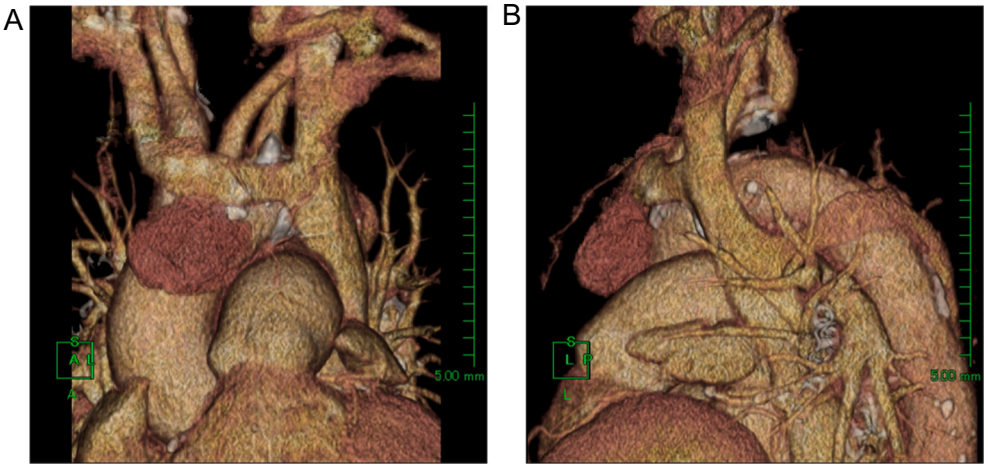

Fig. 2. 3D reconstruction image of the IPLSVC.
A) A frontal view. The thymoma was attached to the right brachiocephalic vein.
B) A lateral view. The azygos vein drains from the dorsolateral aortic arch to the IPLSVC.

Marshall ran in front of the left main pulmonary artery downward and was connected to the right atrium through the coronary sinus. The azygos vein was found only on the left side and flowed into the IPLSVC from the posterior after straddling the aortic arch from the dorsal lateral side (Fig. 2B). Her interviews confirmed that she could tolerate more than 4 metabolic equivalents of exercise. No abnormality was found in the electrocardiogram. The patient's ultrasound cardiography revealed a normal ejection fraction without congenital abnormalities other than dilatation of the coronary sinus.

Robot-assisted thymectomy was performed. A 3 cm median subxiphoid incision was placed for the video port. Two 8 mm ports were added to the mid-clavicular line in the sixth intercostal space. As the patient was 140 cm in height and 45 kg in body weight, her thoracic cavity was small, and the visualization was poor. It was found that the right phrenic nerve ran across the right subclavian vein to the anterior side of the right hilum of the lung (Fig. 3A). The left phrenic nerve was also found adjacent to the IPLSVC, almost equivalent to normal anatomy (Fig. 3B). The IPLSVC crossed in front of the main pulmonary trunk and entered the pericardium (Fig. 3C). Two thymic veins were found to drain into the right brachiocephalic vein. A small infrathyroid vein was found and preserved. The tumor did not invade either the ascending aorta or the right brachiocephalic vein. The tumor was completely removed, and the patient's postoperative course was uneventful. She was discharged on day seven after surgery. The pathological diagnosis was type B1 thymoma of WHO stage I. She was followed up in the outpatient department with no sign of recurrence.

3. Discussion

We report a case of thymectomy with IPLSVC. Although a case with double persistent superior vena cava with anterior mediastinal tumor has been reported, a case with IPLSVC is quite rare [6]. A careful observation of the preoperative computed tomography images helps to diagnose IPLSVC [7–9]. The technical aspect of the thymectomy was not much different from typical surgery other than the reversed location of the veins.

The superior vena cava arises from common cardinal veins that flow into the sinus venosus during the first five weeks of intrauterine life. Four branches flow into the common cardinal veins from the superior, inferior, right, and left (right superior cardinal vein, right inferior cardinal vein, left superior cardinal vein, and left inferior cardinal vein). Around the primitive aorta are transient venous canals that laterally

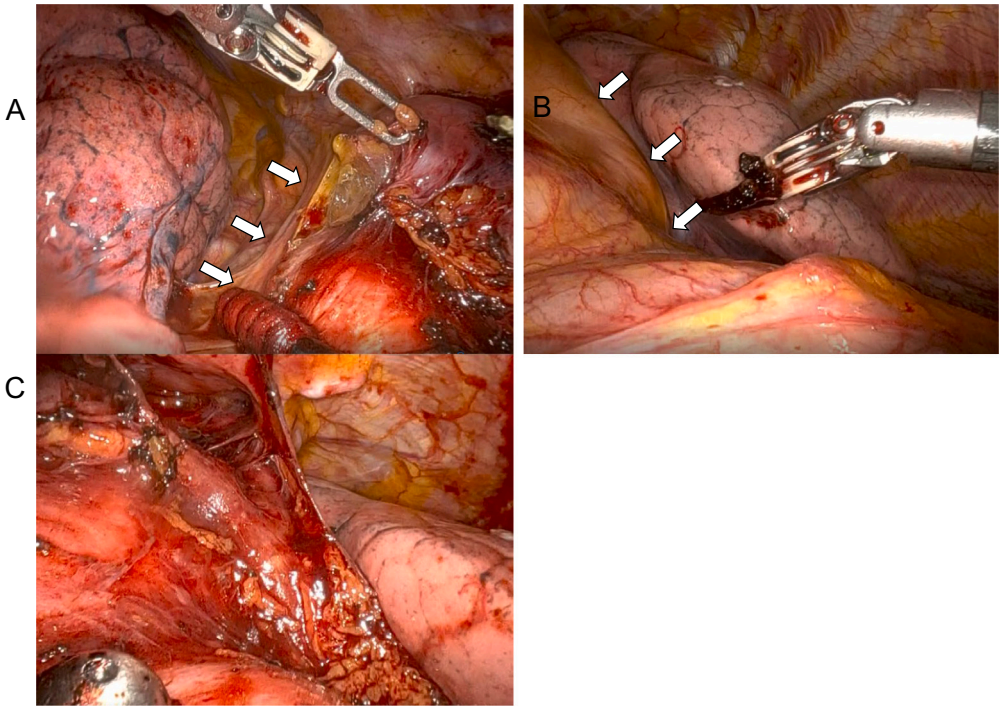

Fig. 3. Intraoperative view.
A) The right phrenic nerve was in its normal position (white arrows). The thymoma was pushed leftward with fenestrated bipolar forceps. There was no invasion of the ascending aorta.
B) The left phrenic nerve was also in its normal position (white arrows).
C) The right brachiocephalic vein and the IPLSVC were merged and crossed in front of the main pulmonary artery.

anastomose the left and right superior cardinal veins. The transverse venous canals merge into a left brachiocephalic vein in normal development in the eighth week of intrauterine life. The left superior anterior cardinal vein, which is more central than the bifurcation of the left brachiocephalic vein, causes obliteration and retracts into the ligament of Marshall. The sinus venosus remaining on the central side develops into the coronary sinus. On the right side, the right superior vena cava is formed from the right superior cardinal vein and the common right cardinal vein.

PLSVC appears when left-sided regression does not occur during development. Ninety percent of cases of PLSVC are so-called double SVCs in which SVCs are found on both the right and left sides [9]. IPLSVC with a defect in the right upper vena cava, as in this case, is extremely rare. In cases of IPLSVCs, it is reported that the arch of the azygos vein is often on the left, and the right often only has the presence of the hemizygos vein [10].

IPLSVC is frequently associated with cardiac malformations. Bartram et al. reported that cardiac malformations are found in nearly half of IPLSVC cases. Care must be taken before and during perioperative management [11]. In 90 % of IPLSVC cases, superior vena cava blood flows into the right atrium mostly via the coronary sinus, is hemodynamically harmless, and is often asymptomatic [9].

In approximately 10 % of cases, superior vena cava blood flow returns to the left atrium. A pattern of direct inflow to the left atrium, indirect inflow to the left pulmonary vein, and right-to-left shunted flow through the incompletely developed coronary sinus has been reported [12,13]. It has been reported that even cases with right-to-left shunted blood flow can be asymptomatic if the amount of shunt is limited [9].

The coronary sinus is often dilated in IPLSVC, and this dilation causes arrhythmia by stretching the tissue around the atrioventricular node. Lenox et al. reported cases in which the sinoatrial node was affected by IPLSVC, and the patients were predisposed to sick sinus syndrome [14]. A shift of the P wave axis toward the left on the anterior wall electrodes or elongated PR time has been reported as electrocardiographic abnormalities associated with IPLSVC [15]. However, these electrocardiographic abnormalities were not observed in our case.

PLSVC may be accompanied by septal or cardiac cushion defects. Many cases of atrial septal defects, ventricular septal defects, and tetralogy of Fallot with IPLSVC have been reported [3,9]. Furthermore, it has been reported that some patients with aortic diseases, such as coarctation of the aorta and heterotaxy, also have PLSVC [3,16,17]. Further investigation is needed on the frequency of cardiac malformations in IPLSVC cases, especially in adults.

We were able to safely complete thymectomy with robot-assisted surgery. Yanagiya et al. reported a case of thymectomy for mucosa-associated lymphoid tissue lymphoma of the thymus with PLSVC [6]. They chose a median sternotomy approach and were able to safely perform a total thymectomy. Their case was a right SVC-residual double SVC case, with a thin bridging vein connecting the left and right SVCs. In this case, the left superior vena cava flowed into the coronary sinus via the vein of Marshall, similar to our case. No reports of total thymectomy in right SVC-deficient IPLSVC cases such as ours could be found during a literature search. Technically, the left and right veins were reversed, and the operator did not feel any particular difficulty during the surgery. When the subxiphoid approach was selected, the exposure of the right brachiocephalic vein was directed from the central side in the lower left to the peripheral side in the upper right, which seemed to be comfortable for a right-handed surgeon. It should be noted that the LSVC enters the pericardium behind the posterior cranial side of the pulmonary artery trunk, so it is slightly away from the left wall of the ascending aorta. When inspection of the subaortic lymph node is necessary, the visualization will be poor. No particular changes were observed in either phrenic nerve, consistent with the case in the report by Yanagiya et al. [6]

4. Conclusions

Thymoma with IPLSVC is a rare combination of diseases. Thymectomy can be performed safely if a careful preoperative evaluation is performed.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this

journal on request.

Provenance and peer review

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Ethical approval

The study is exempt from ethical approval in our institution.

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Guarantor

Tai Hato.

Research registration number

This is not applicable to this case report.

CRediT authorship contribution statement

TH, HF, KM, and NM performed the surgery. (TH was the console surgeon.) TH, HF, KM, and NM collected data. TH, HF, KM, and NM prepared the manuscript.

Declaration of competing interest

The authors have no conflicts of interest to declare.

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