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Pictorial Essay: Understanding of Persistent Left Superior Vena Cava and Its Differential Diagnosis 임상화보: 지속성 좌측상대정맥의 이해와 감별 진단

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Pictorial Essay

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Persistent left superior vena cava (PLSVC) is a rare congenital, thoracic, and vascular anomaly. Although PLSVCs generally do not have a hemodynamic effect, several types of PLSVC and some cardiac anomalies may manifest with clinical symptoms. The presence of PLSVC can render catheterization via left subclavian access difficult when placing a pacemaker or central venous catheter. As such, recognizing a PLSVC that is typically incidentally discovered can prevent complications such as vascular injury. Differentiating vessels found in a similar location as PLSVC is necessary when performing thoracic vascular procedures. This pictorial essay explains the multi-detector CT findings of a PLSVC, and provides a summary of other blood vessels that require differentiation during thoracic vascular procedures.

Index terms Persistent Left Superior Vena Cava; Cardiovascular Abnormalities; Differential Diagnosis; Computed Tomography, X-Ray

INTRODUCTION

Developmental anomalies are often unexpectedly revealed on CT scans. Among them, persistent left superior vena cava (PLSVC) is the most commonly detected thoracic vascular anomaly, and is observed in 0.2%–2.9% of the general population and 1.3%– 11.0% of patients with congenital heart diseases (1). PLSVC is more common in newborns with anatomical malformation such as heart disease, which can also lead to spontaneous abortion or premature death in fetus (2, 3). Although PLSVC may be asymptomatic, various clinical symptoms may occur depending on the location of the drainage and accompanying malformations (4). Nearly 40% of PLSVC patients have various cardiac anomalies, such as bicuspid aortic valve, atrial septal defect (ASD), or coarctation of the Received December 10, 2021 Revised February 1, 2022 Accepted February 21, 2022

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aorta (2). These anomalies are revealed during central venous catheter placement or thoracic surgery. A relatively tortuous course makes the manipulation of central venous catheterization difficult, which can cause hemodynamic instability, arrhythmias (5). Understanding of embryologic features and anatomical location is important when evaluating other thoracic vessels for differentiation. In this essay, we have explained the imaging findings and developmental processes of PLSVC. In addition, we reviewed the CT appearance of several vessels and summarized their routes to allow differentiation from PLSVC.

EMBRYOLOGICAL DEVELOPMENT OF THE PLSVC

PLSVC develops through complex stages during embryological development (2, 6, 7). The embryonic thoracic venous system consists of two pairs of primary veins. The superior cardinal vein returns the blood from the cranial part of the embryo, while the inferior cardinal vein returns the blood from the caudal part. At 5 weeks of gestation, both superior and inferior cardinal veins merge to form common cardinal veins, which connect to form the two horns of the sinus venosus (Fig. 1A). The sinus venosus receives blood from the vitelline veins and umbilical veins. The caudal portion of the right superior cardinal vein and the common cardinal vein combine and develop into the right superior vena cava (SVC). At 8 weeks of gestation, the inferior transverse venous plexus regresses, and the superior transverse venous plexus becomes the normal left brachiocephalic vein (BCV) (Fig. 1B). The cranial part of the right superior cardinal vein and left superior cardinal vein form the subclavian veins and internal jugular veins, whereas the caudal parts of the right anterior and right common cardinal vein form the normal right-sided SVC. Usually, the caudal part of the left superior cardinal vein and the left common cardinal vein merge to form the Marshall ligament. When the left common cardinal vein and left anterior cardinal vein are continuously maintained in the venous system during early development, they persist as a PLSVC (Fig. 1C). The bridging vein appears to be a vessel connecting the right SVC and PLSVC or may undergo idiopathic regression. The right horn of the sinus venosus is integrated into the right atrium structure, while the left horn forms the oblique vein and coronary sinus (CS) of the left atrium, which drain into the major cardiac vein. The inferior vena cava (IVC) develops from the cranial part of the right vitelline vein, while the left vitelline vein and both umbilical veins regress.

RADIOLOGIC FINDINGS AND CLINICAL IMPLICATIONS OF THE PLSVC

In approximately 90% of cases, PLSVC is present with the right SVC, also known as a double SVC (6). Cases of PLSVC without a right SVC are known as isolated PLSVC, which occur in 0.09%–0.13% of patients with other cardiac malformations (8, 9). This occurs when the caudal part of the right superior cardinal vein regresses in the gestational period. In this case of isolated PLSVC patients, cardiac anomalies can be seen in almost 50%, such as ASD, atrioventricular septal defect (AVSD) or tetralogy of Fallot (TOF) (10).

In 65% of the cases, double SVCs run on either side of the area lateral to the mediastinum without connecting blood vessels (Fig. 2) (11). There are cases where two blood vessels are

Fig. 1. Embryological development of venous system and PLSVC.

A. At 5th week of embryological development, the posterior aspect of the heart of an embryo shows the double horn of the sinus venosus with both CCVs, the VVs, and the UVs. There are transverse venous canals called the STVP and ITVP, respectively, which arise around the primitive aorta, connecting the right SCV and left SCV.

B. Normal development of the thoracic venous system. In the 8th week, both SCVs form the internal jugular, subclavian, and BCVs. The caudal part of right SCV and right CCV merge to form the SVC, and the UVs and left VV regress. The right VV persist as an IVC. The left SCV and right SCV connect through the STVP that eventually forms the left BCV. As the left ICV regresses, the right ICV forms the azygos vein. Finally, the left CCV becomes the ligament of Marshall.

C. Development of PLSVC. If the left CCV and the caudal part of left SCV fail to regress, they will persist as PLSVC. A right-sided SVC connects with the PLSVC via a bridging vein, which develops from the STVP.

BCV = brachiocephalic vein, CCV = common cardinal vein, CS = coronary sinus, ICV = inferior cardinal vein, ITVP = inferior transverse venous plexu, IVC = inferior vena cava, PLSVC = persistent left superior vena cava, RA = right atrium, SCV = superior cardinal vein, STVP = superior transverse venous plexu, SVC = superior vena cava, UV = umbilical vein, VV = vitelline vein



connected through the left BCV, which is also known as the bridging vein (Fig. 3).

PLSVC originates from the junction of the left subclavian and jugular veins and passes through the mediastinum lateral to the aortic arch (12). Several variations of the drainage site of PLSVC have been reported. The vessel usually transverses along the ligament of Marshall and drains into the right atrium through the CS, or directly into the right atrium (13-15). In a normal CS, PLSVC flows downward and dilates the CS. However, within the CS ostial atresia, the blood from the coronary vein flows retrograde to the right SVC and into the right atrium via a bridging vein. Additionally, the diameter of the PLSVC in the presence of a bridging vessel is smaller than without a bridging vessel, as it carries only coronary venous blood (16). In about 10%, PLSVC drains into the left atrium directly through the left atrial appendage or via the left pulmonary vein or CS (17). In the case of PLSVC with ASD due to the unroofing of the CS, it is known as Raghib syndrome (6). There are many cardiac anomalies associated with PLSVC, the

Fig. 2. PLSVC without bridging vessel in a 69-year-old female patient.

A. Axial CT image at the level of aortic arch shows the PLSVC (arrow) without a bridging vessel.

B. Axial CT image shows the location of the PLSVC (arrow) at the level of the left main bronchus, with two vessels anterior to the left main bronchus, the PLSVC and left superior pulmonary vein (arrowhead).

C. Axial CT image shows the PLSVC (arrow) draining into the dilated CS, which connects to the right atrium. D. Coronal reformatted CT image depicts the PLSVC (arrow) coursing on the left side of the mediastinum, originating from the left subclavian and jugular vein junction, and no blood vessel connecting the two su-

perior vena cava.

CS = coronary sinus, PLSVC = persistent left superior vena cava



most common being single ventricle, AVSD (Fig. 4), and TOF (1). It is known that heterotaxy such as situs inversus or situs ambiguus are also frequently accompanied by PLSVC (18).

Clinical symptoms of PLSVC vary according to its drainage site and the presence of accompanying anomalies. PLSVC without any cardiac anomaly is usually asymptomatic and is found incidentally. However, if the PLSVC draining into the right atrium expands the CS, the dilated CS can compress the atrioventricular node or his bundle leading to cardiac arrhythmias such as atrial or ventricular fibrillation. Previous studies have reported that PLSVC plays a substantial role in the occurrence and maintenance of atrial fibrillation in about half of cases (8). In addition, an enlarged CS may press the left atrium, decreasing the left ventricular preload (10, 19). PLSVC with left atrium drainage may induce a large right-to-left shunt. Increasing blood flow through this shunt can lead to desaturation, which may cause cyanosis, syncope, and progressive fatigue.

Detection of PLSVC is important prior to cardiovascular procedures such as central venous catheter insertion or pacemaker implantation. In patient with PLSVC, the central venous catheter tip can be located on the left side of the mediastinum, which may be misinterpreted as being misplaced on chest radiography (Fig. 5). PLSVC should also be considered when the CS is dilated during transthoracic echocardiography (4). If PLSVC is not recognized before the

Fig. 3. PLSVC with a bridging vessel in a 59-year-old female patient.

A. Axial CT scan demonstrates a bridging vessel (arrow) that communicates with both SVC (asterisks), coursing anterior to several arteries (brachiocephalic trunk, common carotid artery, and subclavian artery).

B, C. Axial (B) and coronal (C) reformatted images show a tortuous form of PLSVC (arrowheads) that descends along the left side of the mediastinum.

D. Three-dimensional volume rendered image demonstrates a bridging vessel (arrow) connecting the two SVCs (asterisks).

PLSVC = persistent left superior vena cava, SVC = superior vena cava



procedure such as central catheter insertion, it can cause complications such as angina, cardiovascular perforation, arrhythmia, and tamponade (2, 8, 20).

DIFFERENTIAL DIAGNOSES

There are several blood vessels around the mediastinum that are visible in a position similar to the PLSVC. Several anatomical features that help with proper differentiation are the origin site, drainage site, relationship with the other vessels surrounding the mediastinum, and direction of blood flow. Therefore, the connection between the vessel and the surrounding structures should be carefully considered for differentiation. Understanding of the characteristics of accompanying cardiac and non-cardiac diseases is also required. Key differentiating features between PLSVC and its common mimics have been described in the subsequent section and are summarized in Table 1. Fig. 4. PLSVC with AVSD in a 1-year-old female patient.

A, B. Axial CT image (A) and four-chamber view (B) show PLSVC (arrows) with an AVSD consisting two defects in the interventricular septum and interatrial septum.

C. Sagittal reformatted image demonstrates PLSVC (arrows) draining into the CS, which shows diffuse dilatation. D. Echocardiography in a four-chamber view demonstrates a complete AVSD. A large ostium primum atrial septal defect and ventricular septal defect are noted.

AVSD = atrioventricular septal defect, CS = coronary sinus, LA = left atrium, LV = left ventricle, PLSVC = persistent left superior vena cava, RA = right atrium, RV = right ventricle



VERTICAL VEIN

The vertical vein (VV) originates from the pulmonary vein and drains the blood into the left BCV in the case of supracardiac type total or partial anomalous pulmonary venous return (PAPVR) (Fig. 6). About 18% of PAPVR cases originate from the left-sided pulmonary veins, which should be distinguished from PLSVC (21). At the level of the aortic arch, the cephalic portion of the PLSVC and VV appear similar. However, at the level of the main bronchus, these two vessels show different appearance; In the case of PAPVR, there is no vessel at the anterior aspect of the left main bronchus due to the absence of a normally located left superior pulmonary vein. However, in the case of PLSVC, the two vessels, the superior pulmonary vein and the PLSVC, are observed in front of the left main bronchus (22). Moreover, the expected flow direction is caudocranial in the VV, while the flow is craniocaudal in the PLSVC.

Fig. 5. PLSVC containing a central venous catheter in a 48-year-old female patient with breast cancer. **A.** Chest X-ray shows the atypical location of the chemoport catheter along the lateral border of the left cardiac edge. It can be confused as being located in the mediastinum or pleura.

B. Axial CT image showing the tip of the catheter (arrow) located near the left atrial appendage. The vessel in which the catheter is located leads to the CS, which drains into the right atrium (not shown). Since the contrast was injected through a catheter located inside the PLSVC, only a portion of the PLSVC was enhanced.

C. Coronal reformatted CT image shows a central venous line leading to the PLSVC (arrow) along the left edge of the mediastinum. A central venous line is observed inside.

D. Transthoracic echocardiogram in parasternal long axis view shows a dilated CS, which is a finding that suggests the existence of PLSVC.

Ao = aorta, CS = coronary sinus, LA = left atrium, LV = left ventricle, PLSVC = persistent left superior vena cava



LEVOATRIOCARDINAL VEIN

The levoatriocardinal vein (LACV) is a rare cardiac malformation associated with left-sided obstructive lesions (Fig. 7) (23). It functions as a left to right shunt and decompress the left heart obstruction (24). Early in life, a permanent communication between the cardinal vein and pulmonary vein induces a secondary left-sided obstructive lesion, such as mitral atresia or hypoplastic left-heart syndrome. LACV can originate directly from the left atrium (68%) or pulmonary vein (32%) (24). It usually drains into one of the systemic venous structures, which are most commonly the left innominate vein or the right SVC.

LACV and PLSVC can be differentiated via several features. First, unlike LACV that connects to left atrium or pulmonary vein, PLSVC connects inferiorly to the dilated CS. Second, the LACV usually drains into a cephalic direction and less commonly in a bidirectional man-

Feature	Direction of Flow	Origin	Drainage Site	Associations
PLSVC	Usually caudal	Lt subclavian vein and internal jugular vein junction	Coronary sinus or RA or LA	Occasionally associated with cardiac anomalies, or occur in normal heart
Vertical vein in APVR	Cephalad	Lt pulmonary vein	Lt BCV	TAPVR or PAPVR
Levoatriocardinal vein	Cephalad, or bidirectional	Lt atrium or pulmonary vein	Lt BCV, or right SVC	Often associated with left heart obstruction, or rarely in normal heart
Pericardiophrenic vein	Caudal	Internal thoracic, BCV or superior intercostal	IVC through inferior phrenic vein	SVC obstruction
Lt superior intercostal vein	Cephalad	Lt 2-4th intercostal veins	Lt BCV	Various venous obstructions
Aberrant Lt BCV	Caudal, aberrant course passing behind ascending aorta	Lt subclavian vein and internal jugular vein junction	Rt BCV	Often associated with cardiac anomalies, such as TOF, septal defects
Vascular structures secondary to surgery	Usually caudal	SVC or subclavian artery	Usually, pulmonary artery	Postoperative finding of various cardiac anomaly such as TOF, pulmonary stenosis, Rt-sided cardiac obstruction, or hypoplasia

Table 1. Summary of the Characteristic Features of PLSVC and Common Mimics

APVR = anomalous pulmonary venous return, BCV = brachiocephalic vein, IVC = inferior vena cava, LA = left atrium, PAPVR = partial anomalous pulmonary venous return, PLSVC = persistent left superior vena cava, RA = right atrium, SVC = superior vena cava, TAPVR = total anomalous pulmonary venous return, TOF = tetralogy of Fallot

ner, whereas PLSVC is generally caudally oriented. Third, the anatomical relationship of the left pulmonary artery helps to differentiate through the two vessels. PLSVC is located anterior to the left pulmonary artery, whereas the LACV is located posteriorly. Finally, the LACV is almost always associated with left heart obstruction, such as mitral stenosis, aortic stenosis as well as hypoplastic left-heart syndrome, which distinguishes it from PLSVC (25).

In addition, it may be difficult to differentiate between VV and LACV in that both connect BCV and pulmonary vein. However, in the case of VV with anomalous pulmonary vein return, there is no normal connection between the pulmonary vein and the left atrium. On the other hand, LACV connects directly to the left atrium or via the pulmonary vein (23).

PERICARDIOPHRENIC VEIN

The pericardiophrenic vein (PCPV) is involved in the venous drainage of the pericardium and diaphragm (26, 27). It courses along either side of the lateral margins of the mediastinum and heart, as well as lateral to the pericardiophrenic arteries and phrenic nerve. This vessel connect to the internal thoracic vein, superior intercostal vein, or BCV. PCPV is not usually observed in CT, however, when SVC obstruction occurs due to lung cancer or nerve sheath tumor, expanded PCPV may appear due to the development of collateral blood vessels (Fig. 8) (28).

Although it may look similar when comparing PCPV and PLSVC at the superior part, these two vessels can be easily distinguished at their inferior parts. While the PCPV descends to

PLSVC and Differential Diagnosis

Fig. 6. Vertical vein with PAPVR in a 53-year-old female patient.

A, B. A series of axial CT images demonstrate a vertical vein (arrow) on the left side of the mediastinum. It connects the left superior pulmonary vein (arrowhead) with the left brachiocephalic vein. At the aortic arch level, it appears similar to PLSVC on a single axial slice.

C. Axial CT image shows that a left superior pulmonary vein is not visible (dotted arrow) in front of the left main bronchus. Two vessels, the PLSVC and the left superior pulmonary vein, are visible in front of the left main bronchus.

D, **E**. Coronal reformatted CT image and three-dimensional volume rendered image depict PAPVR through the vertical vein (arrows) into the left brachiocephalic vein. It also shows a connection with the left pulmonary vein (arrowheads). Unlike PLSVC, which shows caudal flow, it drives cephalad.

PAPVR = partial anomalous pulmonary venous return, PLSVC = persistent left superior vena cava



the diaphragm and runs lateral to the heart, the PLSVC enters the CS or left atrium.

LEFT SUPERIOR INTERCOSTAL VEIN

The left superior intercostal vein (LSICV) occurs in the second to fourth left intercostal veins, and usually drains into the left BCV (Fig. 9). The vein occasionally communicates with the accessory hemiazygos vein. In a healthy person, the erect chest radiograph rarely shows a small nipple lateral to the aortic arch in less than 5% (29). It can be seen in healthy people and is up to 4.5 mm in size. An underlying venous abnormality should be suspected if the nipple exceeds this size. The LSICV can be dilated in several congenital abnormalities, such as the absence of the IVC or hypoplasia of the left BCV. Also, acquired conditions also cause dilatation, including congestive loss of function, hepatic hypertension, and obstruction of SVC or IVC. Although LSICV has similarities with PLSVC, LSICV runs posterior to the aortic arch and descending aorta. The drainage site and frequently connected vessels facilitate the diagnosis (23, 29).

ABERRANT LEFT BRACHIOCEPHALIC VEIN

An aberrant left brachiocephalic vein (ALBV) is a rare venous anomaly, and its incidence in

Fig. 7. LACV in a 77-year-old female patient with aortic stenosis.

A, B. A series of axial CT images show an LACV (arrows) that travels along the left mediastinum and drains into the left brachiocephalic vein. C. On the axial CT image, at the level of the right main pulmonary artery, an LACV is observed behind the pulmonary artery, and it connects to the left superior pulmonary vein (arrowhead).

D, E. Coronal reformatted CT images and a three-dimensional volume rendered image show the tortuous shape of the LACV (arrows), which connects to the left pulmonary vein along the left side of the mediastinum. It is distinguished from the vertical vein as it enters the left atrium through the left pulmonary vein.

LACV = levoatriocardinal vein



children with congenital heart disease is reported to be approximately 0.5%–1.7% (Fig. 10) (30-32). It is often associated with cardiac anomalies, such as TOF and ventricular septal defect with pulmonary atresia.

ALBV arises from the junction of the left subclavian and jugular veins, passing beneath or behind the aortic arch and merges with the right BCV. Although ALBV may be asymptomatic, it must be differentiated from other major vessels before performing a cardiovascular surgery. Careful tracking of these vascular passages through sequential images is key to differentiation (33).

VASCULAR STRUCTURES SECONDARY TO SURGERY

As several cardiac surgeries change the anatomy of the thoracic blood vessels, postoperative images can show vessels in a similar location to the PLSVC. This can be differentiated by checking cardiac surgery records and anatomical alteration of blood vessels. There are vessels found on the left side of the mediastinum after several cardiac surgeries, including left-sided Blalock-Taussig shunt, bicaval Glenn shunt, and collateral vessels after a Fontan surgery (Fig. 11) (34-36). A left-sided Blalock-Taussig shunt is a passage connecting the subclavian artery Fig. 8. PCPV in a 77-year-old male patient with SVC obstruction due to lung cancer.

A, B. Axial CT images depict dilated PCPVs (arrows) descending vertically into the left mediastinum. A large central lung mass (asterisk) involving the right paratracheal mediastinum, SVC, and right upper pulmonary vessels is observed.

C, D. Coronal reformatted CT image and 3D volumetric rendering image show the PCPVs (arrows) caused by the development of collateral blood vessels due to the lung cancer (asterisk) that result in obstruction of the SVC. The PCPVs originate from the left brachiocephalic vein and drain caudally into the interior vena cava (arrowhead) through the inferior phrenic vein (dotted arrow). PCPV = pericardiophrenic vein, SVC = superior vena cava



and pulmonary artery, which increases the pulmonary blood flow for cardiac obstruction or hypoplasia. A bicaval Glenn shunt is a direct anastomosis of both SVCs to pulmonary arteries presenting with PLSVC for reduced right ventricular end-diastolic volume. Since the anatomical location of these vessels after surgery is similar to that of PLSVC, it should be carefully differentiated. In Fontan surgery, SVC and IVC are connected to the pulmonary artery, and large collateral blood vessels on the left side of the mediastinum can be confused with PLSVC (37).

SUMMARY

PLVSC is a thoracic anomaly that is often accidentally found during a CT scan. PLSVC occurs when some embryonic vessels that degenerate during development continue to exist. Al-

Fig. 9. LSICV in a 58-year-old female patient.

A-D. Axial (A, B) and coronal CT (C, D) images show curvilinear course of the LSICV (arrows) along the left lateral border of the aortic arch, which subsequently connects to the left brachiocephalic vein and accessory hemiazygos vein (arrowhead). This was discovered incidentally, and the diameter of vessel was measured to be about 4 mm.

LSICV = left superior intercostal vein



though PLSVC is often asymptomatic, it occasionally exhibits symptoms that affect the cardiovascular system. It is important to be aware of the presence of PLSVC and associated cardiac anomalies prior to cardiovascular surgery or catheter insertion, which can prevent serious complications. In addition, differential diagnosis of PLSVC is crucial since there are several blood vessels in similar positions. PLSVC can be differentiated by recognizing its anatomical location and driving route in CT image. It is also helpful to identify the origin and drainage location of the various blood vessels, as well as to evaluate the associated anomalies.

Author Contributions

Conceptualization, K.E.; investigation, J.E.R.; supervision, K.E.; visualization, all authors; writing—original draft, J.E.; and writing—review & editing, J.J.H., K.E.

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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Fig. 10. ALBV in a 52-year-old male patient.

A, B. Sequential axial CT images demonstrate ALBV (arrows) running lateral to the aortic arch. The vein traverses the mediastinum between the ascending aorta and the anterior tracheal wall.

C-E. Coronal reformatted CT images and a three-dimensional volume rendered image show an ALBV (arrows) joining the right brachiocephalic vein to form the superior vena cava.

ALBV = aberrant left brachiocephalic vein



Fig. 11. Vascular structures secondary to Fontan surgery in a 19-year-old male patient.

A-C. Axial and coronal reformatted CT images show both superior vena cava (arrows) connected to both the pulmonary arteries. The previously performed Blalock-Taussig shunt is occluded (arrowheads). The interior vena cava is connected to the left pulmonary artery through the extracardiac conduit after the Fontan procedure (asterisks). There is a right-sided aortic arch and abdominal organ, which manifest situ inversus.



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임상화보: 지속성 좌측상대정맥의 이해와 감별 진단

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지속적 좌측상대정맥은 드문 선천성 흉부 혈관 기형이다. 지속적 좌측상대정맥은 일반적으 로 혈역학적 영향이 없지만 몇 가지 유형의 지속적 좌측상대정맥과 동반되는 심장 기형은 임 상 증상을 유발할 수 있다. 지속적 좌측상대정맥의 존재는 심장 박동기 또는 중심 정맥 카테 터를 배치할 때 왼쪽 쇄골 하 접근을 통한 심장으로의 카테터 삽입을 어렵게 만들 수 있다. 우연히 발견된 지속적 좌측상대정맥을 인식하는 것은 혈관 손상과 같은 합병증을 예방할 수 있다. 또한 지속적 좌측상대정맥과 유사한 위치에 있는 다양한 혈관들을 감별하는 것은 향후 흉부혈관시술을 위해서 필요하다. 이번 임상 화보는 지속적 좌측상대정맥의 다중 검출기 컴 퓨터단층촬영 소견을 설명하고 감별이 필요한 다른 혈관들에 대해 요약해 보고자 한다.

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