Rare Case of Left Upper Lobe Partial Anomalous Pulmonary Venous Connection

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ABSTRACT

Partial anomalous pulmonary venous connection is a rare abnormality with failure of connection between the initial draining system of the lungs and the common pulmonary vein. Right sided anomalous return is the most common form of anomalous connection, with left sided anomalous return uncommon. Presented is a case of left upper lobe partial anomalous pulmonary venous connection that was diagnosed incidentally on computed tomography (CT). This is an example of the utility of CT, in particular coronal, sagittal and 3-d reconstructions, in assessment of cardiopulmonary anatomy.

CASE REPORT

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A 66 year old man with no significant past medical history presented to the emergency department with a cough. His only prior thoracic imaging consisted of a chest x-ray 3 weeks prior, which demonstrated a pulmonary nodule measuring 8 mm (Figure 1). A contrast enhanced CT of the thorax was performed in the radiology department for further evaluation. Helical images were obtained of the thorax following administration of 100cc of Optiray300 intravenously. Axial, coronal, sagittal and 3-dimensional reformats were performed (Figures 2,3). The pulmonary nodule measure 8 mm and was calcified, consistent with a benign granuloma. Incidental note was made of the left upper lobe pulmonary veins draining into a vertical vein, which then drained into the left brachiocephalic vein. This represents a partial anomalous left upper lobe pulmonary venous connection. The remaining pulmonary veins and cardiovascular structures were normal in appearance and connection. No signs of cardiac strain or heart failure were noted. No further imaging of the patient has been obtained at our institution.

DISCUSSION

Partial anomalous pulmonary venous return (PAPVC) is a rare entity. The reported incidence is between 0.4 - 0.7% (1). Of these, only approximately 10% are left sided (4). Only 3% of cases have been reported with drainage from the left lung into the innominate vein (7). The most common presentation is a right upper lobe vein draining into either the right atrium or superior vena cava (4). PAPVC most commonly presents with an atrial septal defect (ASD), reportedly in 80-90% of cases (7). Of these, 85% are reportedly sinus venosus type, while 10-15% are secundum type (3). An intact atrial septum is rare. No definitive data is available to establish whether PAPVC is more common in males or females. No associated risk factors have been identified for its development.

Embryonic development of the pulmonary veins occurs early in cardiovascular development. The prevailing theory is that initial drainage is via the splanchnic plexus into the cardinal and umbilicovitelline veins (1). A craniocaudal outpouching forms in the sinoatrial region of the heart with extension to the lung buds (1,6). With caudal regression, the cranial portion develops into the common pulmonary vein, which then incorporates into the left atrial wall (6). Partial anomalous pulmonary venous return occurs due to failure of connection between the common pulmonary vein and the splanchnic plexus (1,6). When occurring in the left upper lobe, a vertical vein is seen collecting the left upper lobe pulmonary veins, and then drains into the brachiocephalic vein (8) (Figure 4).

PAPVC forms a left to right shunt, which is often clinically silent until adulthood (3,5). The patient is predisposed to right-sided volume overload, pulmonary

hypertension, right ventricular dysfunction, tricuspid regurgitation and volume overload (3,5).

There are two thought processes regarding the management of left side PAPVC. The first is to perform a repair before symptoms develop (5). The second is to perform surgical correction only when symptomatic, as surgical complications include atrial fibrillation, complete heart block, cardiac arrest and pulmonary venous obstruction (5). Surgical correction is often performed when analysis shows mild to moderate tricuspid regurgitation, right ventricular dilatation or hypertensive pulmonary vascular disease (3,5). This will lead to symptoms including fatigue, dyspnea, exercise intolerance and palpitations. The goal of the surgical procedure, performed via anterolateral thoracotomy, is to anastomose the vertical vein to the left atrial appendage (5). According to the Mayo Clinic, this may be performed with or without cardiopulmonary bypass (5).

The morbidity and mortality of PAPVC is thought to be low given that most are identified incidentally or upon autopsy; however, no hard data is available (9). Morbidity and mortality is reportedly low with surgical correction, as the previously described complications are less likely with improved technique (5). However, hard data is, again, not available.

Imaging modalities beyond echocardiography are used to assess the cardiopulmonary system. CT allows for accurate characterization of cardiopulmonary anatomy including pulmonary venous drainage, as demonstrated in this case. Coronal, Sagittal and 3d volume rendered reformatted images aid in the assessment of pulmonary veins allowing for additional vantage points. CT demonstrates failure of connection of pulmonary veins to the left atrium. Tubular structures proven to be pulmonary veins drain into a vertical vein (8). MRI will demonstrate the abnormal pulmonary venous connection as well, but can better depict an associated ASD (7). Chest radiography is often normal; however, secondary signs of a left to right shunt, such as cardiomegaly, pulmonary vascular prominence and pulmonary artery hypertension can be visualized (7). Radiographs may also demonstrate tubular opacities within the left upper lobe, which represent the pulmonary veins (8).

A differential consideration of prominent pulmonary veins is a pulmonary varix. A pulmonary varix occurs with stenosis or atresia of one of the four pulmonary veins, resulting in varicose dilatation of another (8). On chest radiography, tubular or round opacities are demonstrated, which vary in size with Valsalva maneuver (8). On CT and MRI, saccular or tubular dilatation of the affected vein is demonstrated (8). Differential considerations of an enlarged vertical vein include an enlarged left superior intercostal vein and persistent left superior vena cava. A left superior intercostal vein courses along the lateral convexity of the aortic arch to drain into the left brachiocephalic vein (8). This can demonstrate a similar appearance to the vertical vein associated with PAPVC on CT. On plain film, an "aortic nipple" is demonstrated measuring approximately 2-3 mm in diameter at the convexity of the aortic arch (8). A final differential consideration is persistent left superior vena cava, which is an incidental finding. In 65% of cases the left brachiocephalic vein is absent and the right SVC is smaller (9). Left Superior Vena Cava (SVC) courses along the left mediastinum. On CT, a tubular structure is noted along the left superior mediastinum without feeding vessels from the lung. The coronary sinus is usually enlarged and can help lead to the diagnosis of left SVC (9).

TEACHING POINT

Partial anomalous pulmonary venous connection is a rare entity, particularly in the left upper lobe, with the potential for significant clinical consequences such as right-sided volume overload, pulmonary hypertension, right ventricular dysfunction and tricuspid regurgitation. Coronal, sagittal and 3d volume rendered reformats are particularly useful in fully assessing the pulmonary venous anatomy.

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FIGURES

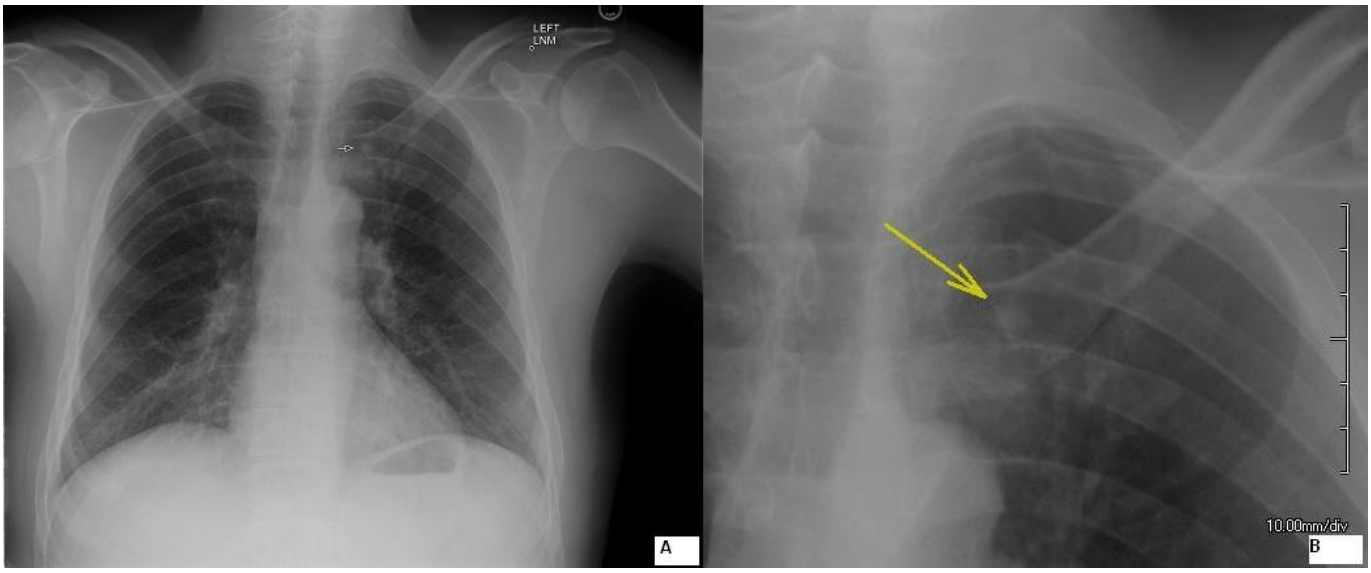

Figure 1: 66 year old Male with Left Upper Lobe Partial Anomalous Pulmonary Venous Connection. Image A: PA Chest X-Ray demonstrates an 8mm left upper lobe pulmonary nodule (arrow). The lungs are otherwise clear.
Image B: Magnification view better demonstrates the nodule. (Phillips CR)

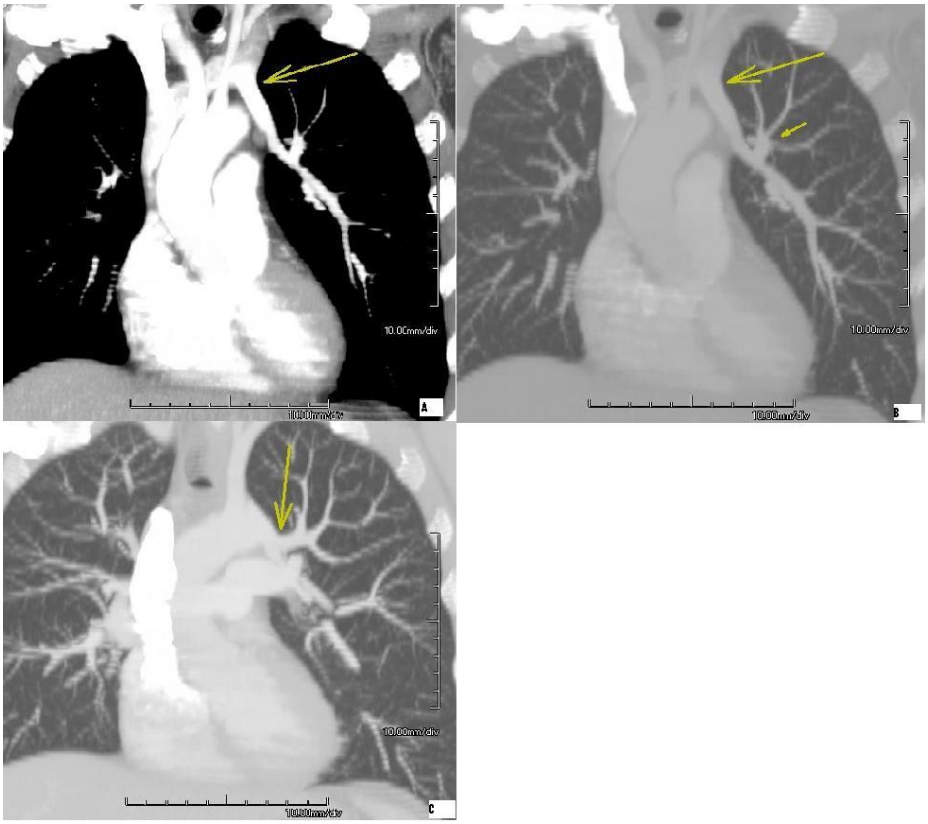

Figure 2: 66 year old Male with Left Upper Lobe Partial Anomalous Pulmonary Venous Connection.
Image A: A coronal oblique CT image denotes a Vertical Vein (arrow) along the left aspect of the mediastinum draining into the Brachiocephalic Vein. The left upper lobe Pulmonary Veins attach to the Vertical Vein instead of the Left Atrium creating a Left to Right shunt.
Image B: A coronal oblique CT image denotes a Vertical Vein (large arrow) along the left aspect of the mediastinum draining into the Brachiocephalic Vein. The left upper lobe Pulmonary Veins attach to the Vertical Vein (small arrow) instead of the Left Atrium, creating a Left to Right shunt.
Image C: A coronal oblique CT image denotes a Vertical Vein along the left aspect of the mediastinum, which drains into the Brachiocephalic Vein. The left upper lobe Pulmonary Veins attach to the Vertical Vein (arrow) instead of the Left Atrium creating a Left to Right shunt.
(Protocol: Phillips CT scanner, 170 mA, 120 kV, 3 mm slice thickness, 100 cc IV of Optiray 300).

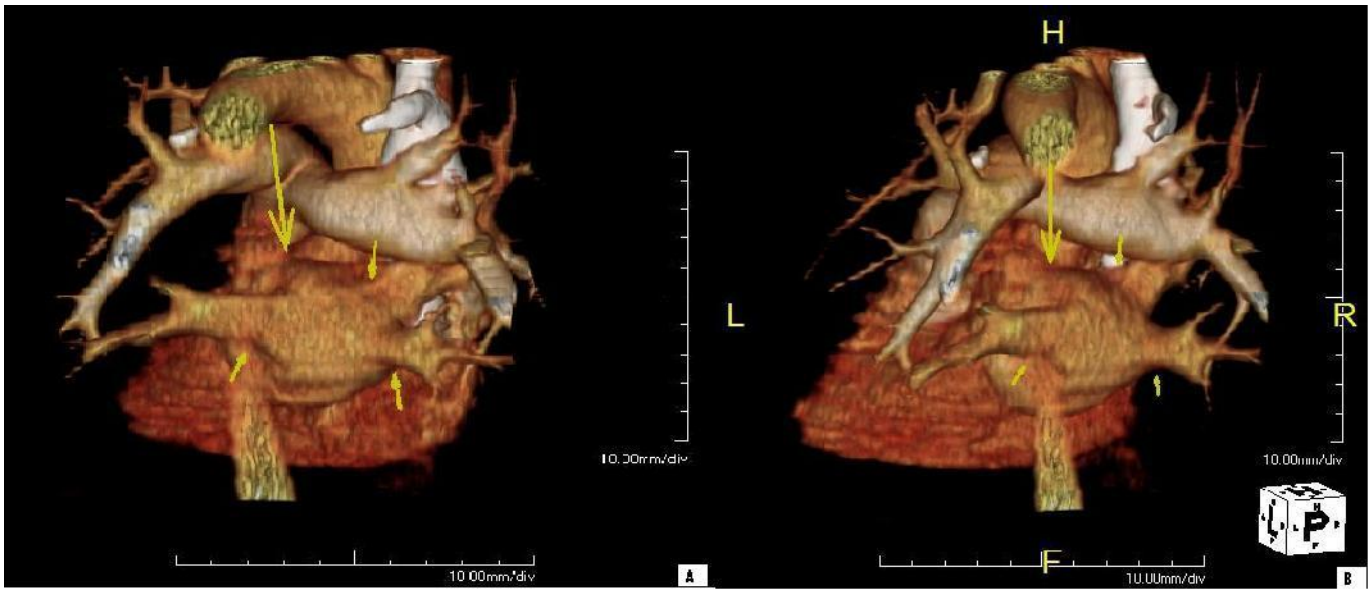

Figure 3: Image A: 66 year old Male with Left Upper Lobe Partial Anomalous Pulmonary Venous Connection. A 3d volume rendered image from a posterior coronal oblique position denotes the normal Pulmonary Veins connecting to the Left Atrium (small arrows). The left upper lobe Pulmonary Vein is not seen connecting to the Left Atrium (large arrow).
Image B: A 3d volume rendered image from a posterior coronal oblique position denotes the Normal Pulmonary Veins connecting to the Left Atrium (small arrows). The left upper lobe Pulmonary Vein is not seen connecting to the Left Atrium (large arrow).
(Protocol: Phillips CT scanner, 170 mA, 120 kV, 3 mm slice thickness, 100 cc IV of Optiray 300).

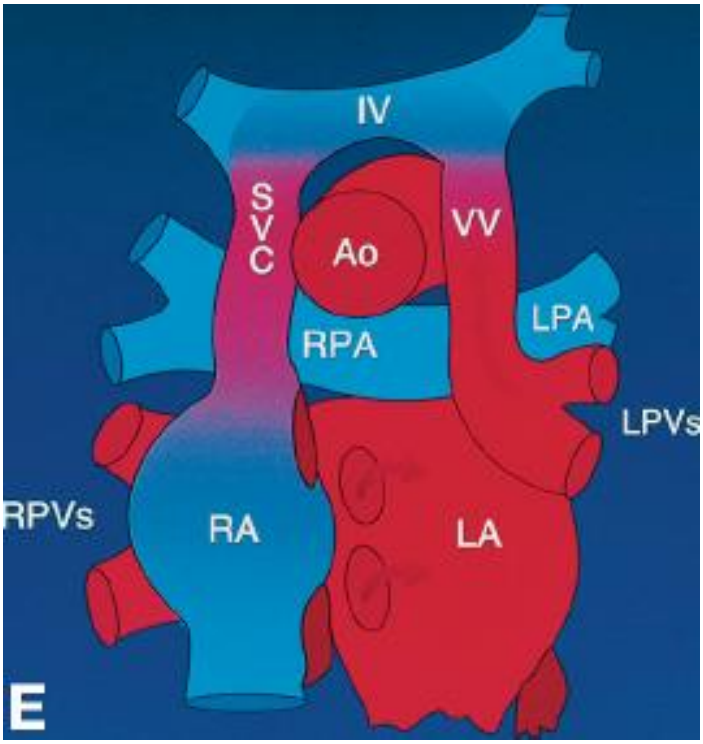

Figure 4 (left): "Frontal view of the heart. The left PVs connect to a vertical vein (VV) that enters the innominate vein (IV). The innominate vein enters the normal SVC and delivers the left pulmonary venous blood to the RA."
Reprinted from Journal of the American College of Cardiology, Vol29 /Issue6, Naser M Ammash, James B Seward, Carole A warnes, Heidi M Connolly, Patrick W O'Leary, Gordon K Danielson, Partial Anomalous Pulmonary Venous Connection: Diagnosis by Transesophageal Echocardiography, 1351-1358, 1997, with permission from Elsevier.

Etiology	Failure of connection between the common pulmonary vein and the splanchnic plexus
Incidence	0.4-0.7%, 10% are left sided
Gender	No definitive data available
Age	Congenital anomaly but often not found until middle age
Risk Factors	None
Treatment	Surgical correction depending on symptomatology or before symptoms develop.
Prognosis	Normal life span
Radiography	Usually normal but may demonstrate cardiomegaly, pulmonary vascular plethora, pulmonary artery hypertension. May see tubular opacities demonstrating the pulmonary veins.
CT	Dilatation of intraparenchymal vessels, right ventricular cavity and right atrium. Pulmonary vein connecting to systemic vein
MRI	Same as CT. Better depicts septal defects.

Table 1: Summary table of Partial Anomalous Pulmonary Venous Connection

	Radiography	**CT**	**MRI**
Partial anomalous pulmonary venous connection	Usually normal but may demonstrate cardiomegaly, pulmonary vascular plethora, pulmonary artery hypertension. May see tubular opacities demonstrating the vertical vein or pulmonary veins.	Dilatation of intraparenchymal vessels, right ventricular cavity and right atrium. Pulmonary vein connecting to systemic vein.	Dilatation of intraparenchymal vessels, right ventricular cavity and right atrium. Pulmonary vein connecting to systemic vein. Better depicts septal defects.
Pulmonary Varix	Round opacities. Diameter varies with Valsalva or Muller maneuvers.	Dilatation of affected vein. Normal pulmonary venous connection.	Dilatation of affected vein. Normal pulmonary venous connection.

Table 2: Differential diagnosis table of dilated pulmonary veins

	Radiography	CT	MRI
Partial anomalous pulmonary venous connection	Usually normal but may demonstrate cardiomegaly, pulmonary vascular plethora, pulmonary artery hypertension. May see tubular opacities demonstrating the vertical vein or pulmonary veins.	Dilatation of intraparenchymal vessels, right ventricular cavity and right atrium. Pulmonary vein connecting to systemic vein.	Dilatation of intraparenchymal vessels, right ventricular cavity and right atrium. Pulmonary vein connecting to systemic vein. Better depicts septal defects.
Left superior vena cava	Left superior mediastinum structure.	Tubular structure left superior mediastinum. No feeding vessels from the lungs. Enlarged coronary sinus.	Tubular structure left superior mediastinum No feeding vessels from the lungs. Enlarged coronary sinus.
Left superior intercostal vein	2-3 mm aortic nipple.	Vein adjacent to the lateral convexity of the aortic arch.	Vein adjacent to the lateral convexity of the aortic arch.

Table 3: Differential diagnosis table of Vertical Vein

ABBREVIATIONS

ASD: atrial septal defect
CT: computed tomography
MRI: magnetic resonance imaging
PAPVC: Partial Anomalous pulmonary venous connection
SVC: superior vena cava

KEYWORDS

Thorax; Anatomy; Partial anomalous pulmonary venous connection