ABNORMAL LEFT SINGLE PULMONARY VEIN: CASE REPORT OF AN ANECDOTAL VARIANT

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We describe here a patient with a very rare ectopic unilateral single pulmonary vein, with normal distal drainage into the left atrium, and thus a non pathologic finding to be distinguished from clinically significant abnormalities.

Key-word: Pulmonary veins, abnormalities.

MDCT of the chest with intravenous injection of contrast is the current gold standard to exclude pulmonary embolism or metastasis. The radiologist should know all the variations of the pulmonary vascularization and recognize those potentially clinically significant and those not. Pulmonary veins may vary in course, in origin or destination, in number or in size. We describe here a patient with a very rare ectopic unilateral single pulmonary vein, with normal distal drainage into the left atrium, and thus a non pathologic finding.

Case report

Contrast chest CT was performed in an 84-year-old lady for persistent infiltrates on plain chest X-ray. A large vein in the left lung showed a very abnormal course through the left lung, (Fig. 1 A-D): it starts as a left superior vein, climbs up, crosses the fissure and goes down through the left lower lobe, joining the inferior pulmonary vein to drain normally into the left atrium (Fig. 2).

Discussion

Pulmonary veins variations can be clinically significant, when there is an abnormal partial venous return with left-to-right shunt. It can be an anecdotal finding, as in this case, where the only abnormality is the trajectory and the number, without shunt.

A review of the literature revealed around 20 cases of anomalous unilateral single pulmonary vein (AUSPV) (1). Four cases similar to our case were reported. The right-sighted cases are more frequent, and a case of bilateral single pulmonary vein was described (2). The first case of left AUSPV has been reported in 1968 (1, 2).

This congenital anomaly could be the result of atresia or hypoplasia of one of the major pulmonary

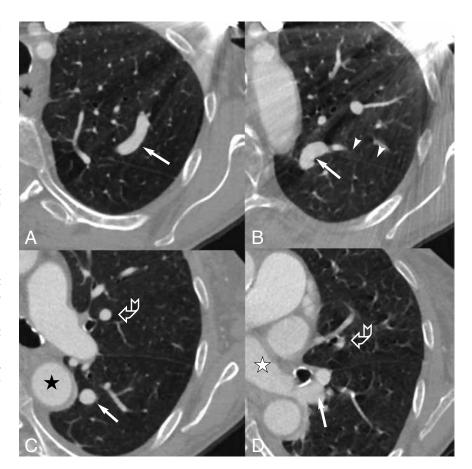


Fig. 1. — MDCT with contrast: from top to below. A shows the top of the abnormal vein (white arrow). B shows the vein (white arrow) crossing the main left fissure (arrow heads). C: ascending course of the vein (black curved arrow) trough the left upper lobe and medial descending course (white arrow) of the vein along the thoracic aorta (black star). D: Origin of the vein in the lingular area, passing near from the hilum of the left lung (curved black arrow) and his end (white arrow) draining into the left atrium (white star) after joining the left inferior pulmonary vein. CT parameters in this case were the following: 64 slices MDCT, 1,25 mm thin slices, 100 kV, 255 mAs resulting in patient in a CTD/vol of 4,16 mGy, and DLP 143,37 mGy-cm.

veins (3). The congenital abnormalities of the pulmonary venous system may vary according to their origin, their drainage, course, or number. Some authors distinguish the AUSPV from a variant called "the meandering pulmonary vein", which has a curved course through the

lung and drains into the inferior vena cava and/or in the left atrium directly, not via the inferior pulmonary vein like in this case (4).

MDCT after intravenous injection of contrast allows to differentiate congenital anomalies of the pulmonary veins which can be classified in three different categories (5): (a) anomalous pulmonary venous drainage, partial or total, with or without abnormal course in the lung, (b) anomalous



Fig. 2. — Thickened-MIP oblique slice showing the end of the AUSPV (white arrow) joining the left inferior pulmonary vein (curved black arrow) to drain into the left atrium (black star).

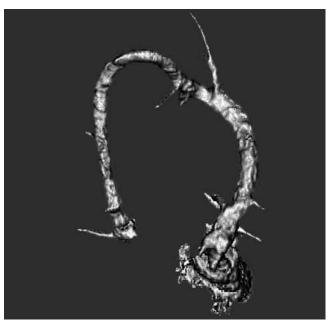


Fig. 3. — Lateral view of a 3D volume rendering-reconstruction of the abnormal vein, starting in the lingular area, passing near from the hilum, ascending to the apex, crossing the fissure, and descending to finally drain into the left atrium, after joining the left inferior pulmonary vein.

pulmonary venous route with normal drainage (like this case), and (c) abnormal venous diameters (varices, stenosis and atresia).

The pattern of anomalous pulmonary venous drainage presents a pulmonary venous blood flowing directly into the right heart or the systemic veins, partially or totally, causing a right-to-left shunt. The total anomalous drainage is uncommon and is associated with an obligatory septal defect causing a right to left shunt. A thirty of different anomalous connections were made, classified by supracardiac, cardiac, infradiaphragmatic or mixed. Those anomalies may be seen on chest Xray, but are more characterized by contrast-enhanced CT. A pulmonary venous hypertension may be caused when the entire venous drainage of an entire lung passes into the right atrium without septal defect.

The main example of anomalous pulmonary venous drainage is the hypogenetic lung (Scimitar) syndrome. It is characterized by hypoplasia of the right lung and anomalous pulmonary drainage from it to the inferior vena cava (IVC), and rarely the suprahepatic portion of the IVC, hepatic veins, portal vein, azygos vein, coronary sinus or right atrium. It may be associated with cardiovascular anomalies, anomalies of the right bronchial tree, diverticula, and, occasionally, with extension of a portion of the right lung across the midline

into the left hemithorax. The abnormal course of the vein is usually visible on chest X-ray with a curved shadow descending to the diaphragm, right to the heart, mimicking a Turkish sword (or scimitar).

In literature (6), different cases of AUSPV were confused with an arteriovenous malformation on plan chest X-ray. The difference must be made with further investigation before any intervention, even in the absence of symptoms due to a right-to-left shunt (reduction in the arterial oxygen saturation, cyanosis, polycythemia or paradoxical emboli). Incidence of AVM rises in context of Osler-Rendu disease, Fanconi's syndrome and polysplenia syndrome.

The tortuous aspect of the AUSPV may be confused with a varix, particularly on angiography, which is a dilated and tortuous aspect of a pulmonary vein normally entering to the left atrium without course abnormality (3).

The AUSPV is very rare, asymptomatic and has no consequence, not being associated with a left-to-right shunt. No further diagnostic test or treatment is needed. The only relevant clinical implication of such a variant would be in case of a lobectomy, because the vein crosses the fissure (7).

Conclusion

Classification of congenital anomalies of the pulmonary venous sys-

tem is complex. The radiologist must be aware of the many variations and distinguish relevant variations (such as those causing left-to-right shunts) from incidental findings.

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