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CASE REPORT

Anomalous single pulmonary venous trunk

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ABSTRACT

We present a rare case of a single pulmonary venous trunk that drains all pulmonary segments to the left atrium. This anomaly may mimic pulmonary arteriovenous malformation on imaging. This report briefly discusses the embryology of the pulmonary veins and the classification of the pulmonary veins anomalies.

Key words: Anomaly of the pulmonary veins, single pulmonary venous trunk, pulmonary arteriovenous malformation

INTRODUCTION

The pulmonary vascular bed drains into left atrium through four individual pulmonary veins; one superior and one inferior vein from the right and left lung connect directly to the corresponding side of the left atrium.[1] This normal arrangement occurs in 82% of the population.[1] In anatomic terms, pulmonary venous anomalies are classified as anomalous connections, anomalous drainage, or abnormal numbers of pulmonary veins.[2,3] Anomalous pulmonary venous connection refers to the situation when some or all pulmonary veins abnormally connect to systemic veins instead of the left atrium, resulting in abnormal pulmonary venous drainage and a left to right shunt. Abnormal pulmonary venous drainage occurs when some or all pulmonary veins connect normally to the left atrium but drain abnormally, most typically due to atrial septal malalignment, resulting in a left to right shunt.[2,3] Finally, the number of pulmonary veins draining into the left atrium can vary from one to five veins.[2,3] There is wide variation in the anatomy, clinical presentation, and expected outcome of these anomalies.[1-3] The most common variation of the pulmonary veins number is the presence of a common pulmonary vein on either the right or left side with an incidence of 23.9%.[2] The next most common variation is the increase in their numbers which is mainly observed as an additional vessel called the right middle lobe pulmonary vein with a prevalence of 1.6%.[2]

In this case, a single pulmonary vein drains all pulmonary segments and connects to the left atrium. In this unusual arrangement, the right upper and right lower pulmonary veins combine in the right hilum, pass through the mediastinum, and follow a tortuous course in the left lung before combining with a tortuous left upper and lower pulmonary vein, ultimately draining the entire pulmonary venous return through a single orifice into the left atrium.

CASE REPORT

A 4-month-old previously healthy female was referred to our institution for evaluation of recurrent respiratory distress which was initially treated as bronchiolitis. A chest radiograph showed a left perihilar density, which was suspicious for a left-sided pulmonary arteriovenous malformation (PAVM) [Figure 1].

A chest CT scan with contrast and 3D reconstruction revealed an extensive pulmonary vein abnormality [Figure 2]. The right pulmonary veins joined to form a common right trunk which traveled posterior to the left atrium and under the left pulmonary artery. The common right trunk then passed anterior and superior to the left pulmonary artery where the left upper pulmonary vein joined the common right pulmonary vein. The new common pulmonary vein then followed a tortuous course in the parenchyma of the left lung and ultimately traveled the normal course of a dilated left lower pulmonary vein before entering the left atrium through a single orifice. A small patent ductus arteriosus (PDA) was also identified.

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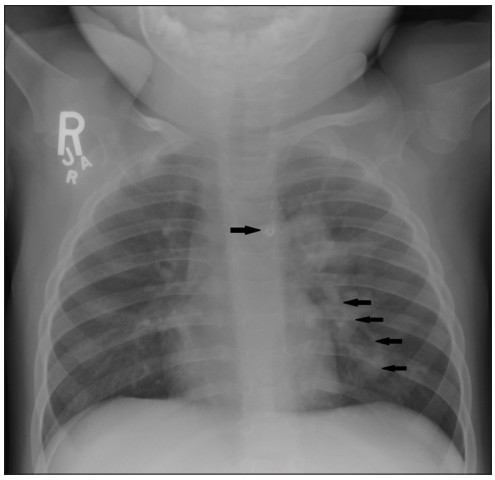

Figure 1: Plain radiograph demonstrating rounded left perihilar densities created by the abnormal pulmonary veins (arrows). A coil is also seen in the PDA

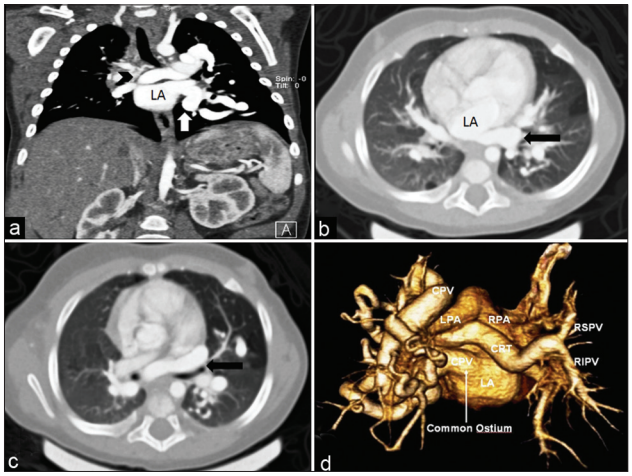

Figure 2: (a) Coronal image. Single pulmonary vein (arrowhead) crosses the midline superior to the left atrium to join a vessel in the left lung. Single pulmonary vein joins the left atrium (arrow). (b) Transverse image. Single pulmonary vein joining the left atrium. (c) Transverse image. Pulmonary vein crosses midline. (d) 3D image. The course of the common pulmonary vein. LPA - Left pulmonary artery; RPA - Right pulmonary artery; RSPV - Right superior pulmonary vein; RIPV - Right inferior pulmonary vein; CPV - Common pulmonary vein; CRT - Common right trunk; LA - Left atrium

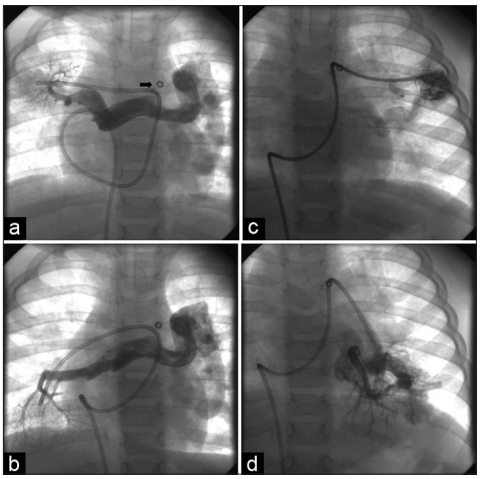

Figure 3: Pulmonary artery wedge injections in the right upper lobe pulmonary artery (a), right lower lobe pulmonary artery (b), left upper lobe pulmonary artery (c), and left lower lobe pulmonary artery (d), demonstrating the course of the individual pulmonary veins. A coil can be seen in the ductus arteriosus (small arrow)

Due to the history of unexplained recurrent respiratory distress, cardiac catheterization was performed to evaluate for pulmonary venous obstruction that was not found on the CT scan. This confirmed the anatomy identified on the CT scan [Figure 3]. There was no evidence of pulmonary venous obstruction (normal pulmonary arterial and venous pressure with no gradient on pull back from pulmonary vein into left atrium through trans-septal access via the patent foramen ovale). The small PDA was occluded by coils. No further management was required.

DISCUSSION

The pulmonary venous anomalies are best understood by examining its embryology. The common pulmonary vein develops around the embryonic day 24 at the sinoatrial region to the left of the septum primum.[2,3] By embryonic day 32, the majority of pulmonary venous return drains into the common pulmonary vein.[2,3] Through differential growth, the common pulmonary vein becomes incorporated into the left atrium and connections to the systemic veins are lost resulting in the usual arrangement of four pulmonary veins draining into the left atrium with separation of the systemic and pulmonary venous systems.[2,3]

Pulmonary venous anomalies are thought to result, in part, from abnormal development of the common pulmonary vein.[2,3] Atresia of the common pulmonary vein that occurs before obliteration of the pulmonary to splanchnic venous plexus connections results in total anomalous pulmonary venous return.[2,3] Stenosis of the common pulmonary vein can result in cor triatriatum.[2,3] Anomalous incorporation of the common pulmonary vein into the left atrium can result in an abnormal number of pulmonary veins.[3] In this case, one could hypothesize that abnormal incorporation of the pulmonary vein into the left atrium with the persistence of interpulmonary venous connections, but regression of pulmonary to splanchnic connections, could result in the anatomy we see in this case.

A decreased number of pulmonary veins with abnormal interpulmonary venous structures has been described.[2,4-7] These anomalies are typically asymptomatic and are incidental findings found on imaging studies. The most common variation is the presence of three pulmonary veins, with a single pulmonary vein draining

the ipsilateral lung.[2,4-6] Other anomalies have been documented, including pulmonary venous structures crossing the mediastinum and anastomosing with veins in the contralateral lung.[7] To our knowledge, a single pulmonary vein draining into the left atrium has been rarely described.[2,8]

The anomalies of the number of pulmonary veins, as in this case of the single pulmonary venous trunk, may mimic PAVM on the roentgenographic imaging of the chest.[9] The classic roentgenographic appearance of a PAVM is that of a round or oval mass of uniform density, frequently lobulated but sharply defined, more commonly in the lower lobes, and ranging from one to 5 cm in diameter.[10] Currently, helical contrast-enhanced chest computed tomography or contrast-enhanced magnetic resonance angiography seems to be a very useful nonionizing and noninvasive procedure for the diagnosis of PAVM and the anomalies of the number of pulmonary veins like in our case.[11] Despite advances in the techniques mentioned thus far, contrast pulmonary angiography remains necessary when invasive therapy is being considered for PAVM and the anomalies of the number of pulmonary veins.[12]

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