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Unilateral absence of the pulmonary veins: an unusual diagnosis with characteristic imaging findings

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ABSTRACT

Background: Congenital unilateral absence of the pulmonary vein (UCAPV) is a rare entity with characteristic clinical and imaging findings. Despite its congenital nature, the radiographic findings and symptoms of UCAPV may not be recognized at birth and patients may present in childhood or early adulthood with findings that may mimic other diagnoses.

Methods: The evolution of imaging findings in UCAPV is presented through two cases, one of which demonstrates the progression of findings over several years. The embryologic basis of this entity is reviewed and the clinical presentation and characteristic imaging findings including radiographs, nuclear scintigraphy, computed tomography, magnetic resonance imaging and cardiac catheterization are demonstrated.

Results: Characteristically, normal at birth, radiographs demonstrate the gradual development of a small lung and ipsilateral pulmonary artery over time. In addition to unilateral absence of the pulmonary veins on CT or MRI, a mediastinal "soft tissue mass" reflecting the development of mediastinal collaterals is a common finding and should be recognized as secondary to the absent ipsilateral pulmonary veins rather than as a primary process causing occlusion of the pulmonary veins. Scintigraphy will show absent perfusion to the affected lung.

Conclusion: Awareness of the distinctive imaging findings in this unusual condition is critical to avoid misdiagnosis and to prevent the consequences of UCAPV which include pulmonary hypertension and extensive venous collaterals with or without hemoptysis, both of which may prevent definitive repair.

1. Introduction

Congenital unilateral absence of the pulmonary vein (UCAPV) is a rare entity that follows a typical evolution of radiographic and clinical findings. Since the condition is often not appreciated at birth, its congenital basis may not be recognized at presentation. Imaging findings may be confusing, leading to missed or delayed diagnosis. Herein we present the characteristic evolution of clinical and imaging findings in a patient with this unusual condition.

2. Case 1

A 4 year old girl with a history of mild persistent asthma, allergic rhinitis and multiple hospital admissions for recurrent pneumonias since age 2 years (prompting an extensive evaluation for aspiration, primary immunodeficiency and autoimmune disease), presented with hemoptysis.

A chest radiograph demonstrated patchy bilateral parenchymal opacities.

Radiographic findings at each prior admission varied from those suggestive of bronchiolitis to unilateral focal parenchymal opacities to multifocal bilateral parenchymal opacities. Although a chest radiograph obtained immediately after birth for retained fetal fluid was normal and demonstrated no asymmetry in lung size, subsequent radiographs demonstrated progressive asymmetry of the size of the lungs with the right lung and the right pulmonary artery smaller than the left (Fig. 1).

Bronchoscopy performed to determine the cause of the hemoptysis, revealed hemosiderin laden macrophages. Cultures were negative. A contrast enhanced CT showed a slightly small right lung, diminished caliber of the right pulmonary artery (RPA), normal bronchi, patchy areas of ground glass opacity were present in the right lower lobe and a

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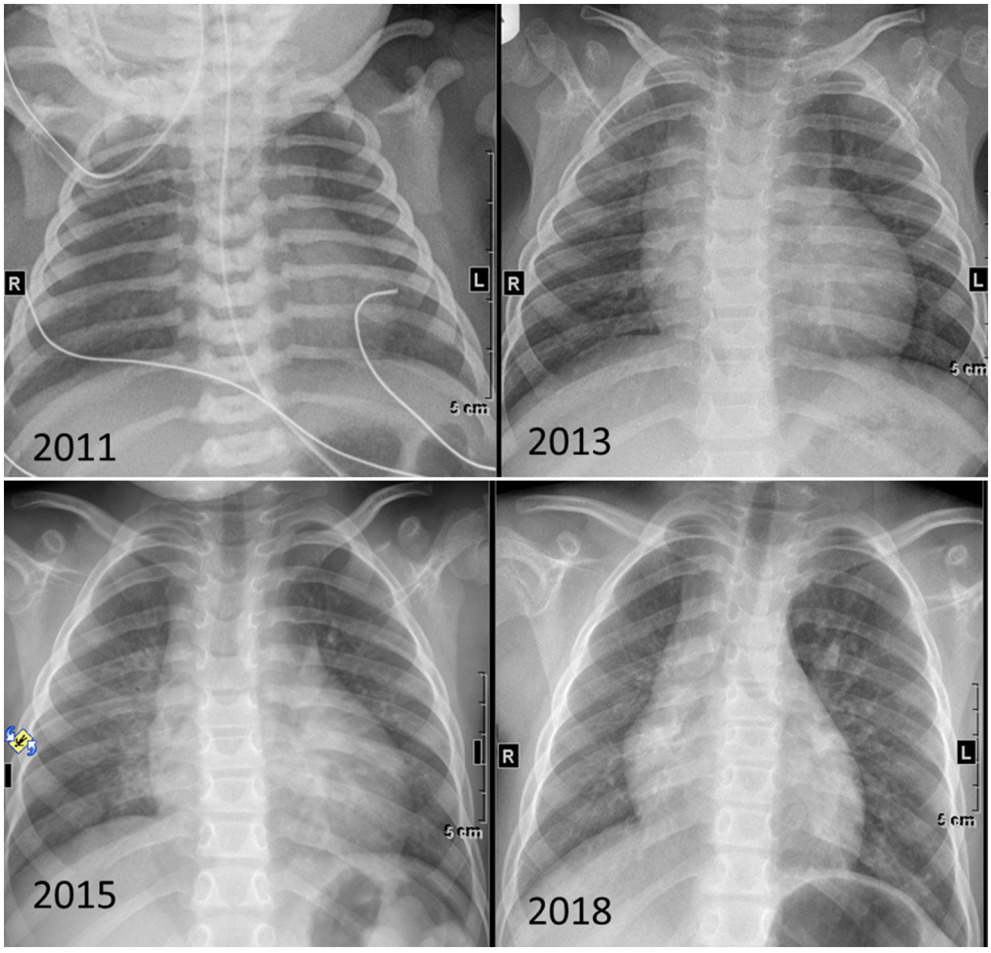

Fig. 1. Serial frontal chest radiographs from birth (top right) to age 7 years (bottom left). While the initial film shows normal symmetry of the hila and lungs, over time asymmetry of the lungs as well as the hila is apparent, with the right side smaller than the left.

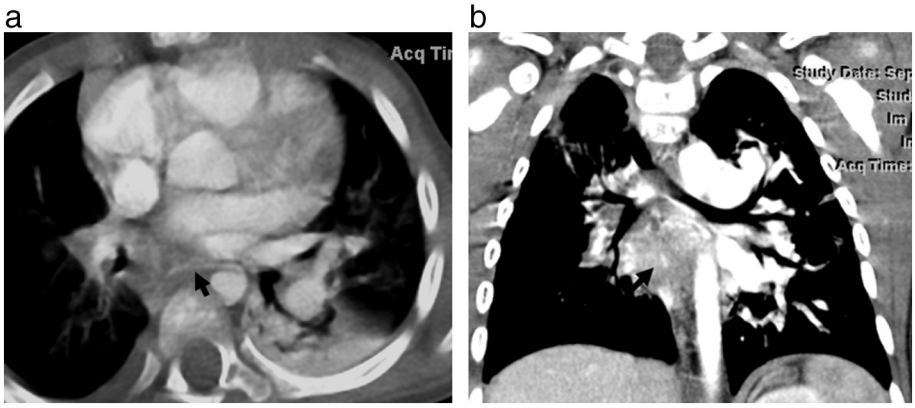

Fig. 2. (a,b): Axial (a) and Coronal(b) contrast enhanced CT images of the chest at age 4 years demonstrate soft tissue density in the right middle mediastinum and right hilum. The right sided bronchi are patent. Note the enhancing vessels coursing through the mediastinal soft tissue process (arrows). On the axial image this represents a large bronchial artery.

middle mediastinal soft tissue infiltrative mass (Fig. 2). Enlarged bronchial arteries were noted coursing through the mass and a diagnosis of a vascular malformation was entertained. Nonvisualization of the right pulmonary veins was considered secondary to compression by the soft tissue mass.

As the patient was stable, it was elected to follow her. A repeat CT performed at age 6 years again demonstrated a stable middle mediastinum infiltrating mass. Decreased right lung volume, narrowing of the RPA and absence of the right pulmonary veins (confirmed on echocardiogram) were noted (Fig. 3); left pulmonary veins were normal.

Given the presence of a presumed slow growing infiltrative mass narrowing the RPA and obstructing the right pulmonary veins, the patient was scheduled for biopsy. Prior to surgical biopsy, additional imaging was requested included a PET scan (no abnormal radiotracer uptake) and a lung perfusion scan which revealed absence of right lung perfusion (Fig. 4). Core needle biopsy revealed fragments of fibrovascular and adipose tissue. Subsequent thoracotomy with open biopsy was performed. The pathology report described a reactive node with mild interfollicular hyperplasia and no evidence of lymphoma.

3. Case 2

A Middle Eastern male infant presented at 6 months of age with episodic respiratory distress.

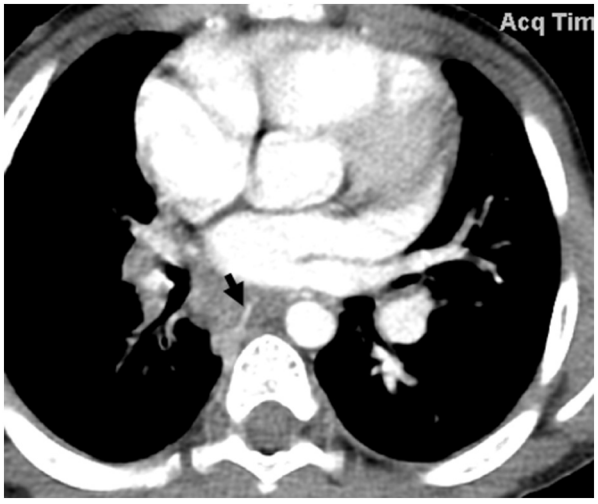

Fig. 3. Axial contrast enhanced CT image of the chest at age 6 years. The soft tissue process in the posterior mediastinum and involving the right hilum is stable. A prominent vessel (arrow) is again seen coursing through the posterior mediastinal soft tissue process. While the left pulmonary veins are present, the right pulmonary veins are absent.

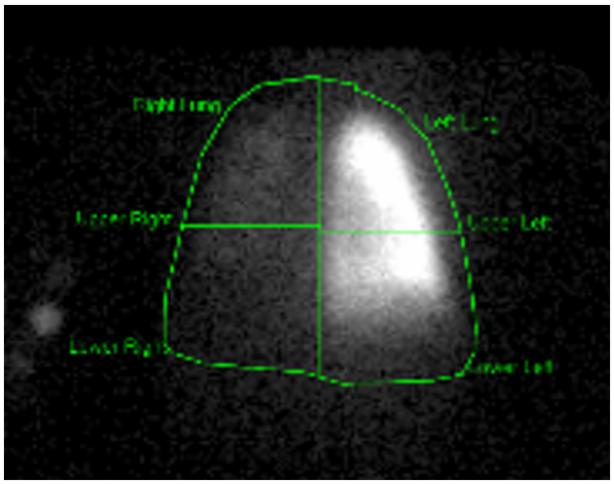

Fig. 4. Anterior 99mTc Macroaggregated albumin perfusion scan performed at age 6 years demonstrates absent perfusion to the right lung.

The patient also had a history of hypotonia, developmental delay, microcephaly, right choanal atresia, left ptosis, and right clubfoot; chromosomal microarray was negative.

A chest radiograph (Fig. 5a) revealed a hypoplastic right lung with hyperinflation of the left lung and decreased pulmonary vascularity in the right lung compared to the left. Prominence of the inferior aspect of the right hilum was noted. Symptoms persisted and at 9 months of age, a CT angiogram was performed (Fig. 5b). This revealed absence of the right pulmonary veins and prominent hypodense tissue in the middle mediastinum extending to the right hilum. There was no compression of the right sided bronchi. Reconstructioned images at 1.25 mm (not shown) demonstrated numerous tiny vessels within the hypodense tissue. Septal thickening was present throughout the right lung, especially in the lower lobe posterolaterally. A cardiac catheterization (Fig. 5c) showed non-visualization of the right superior and inferior pulmonary veins with stasis in multiple right lower lobe venous collaterals. The right pulmonary artery was relatively small as were its branches.

At surgery, the right superior and inferior pulmonary veins were represented by fibrotic cords proximal to the left atrium; peripheral

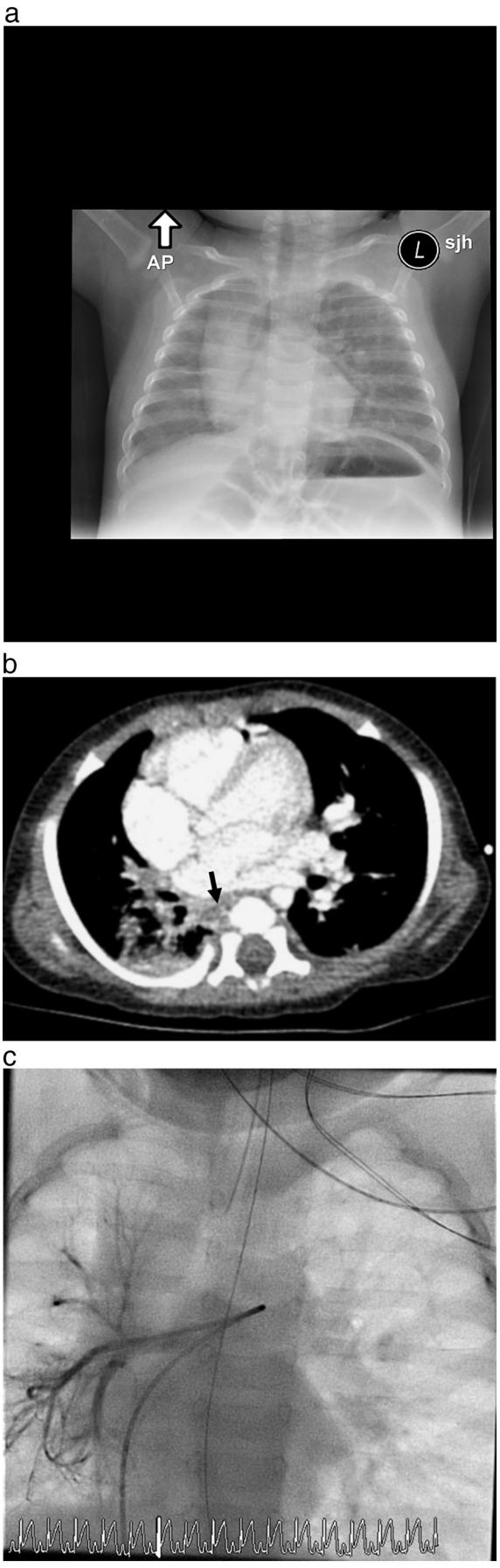

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Fig. 5. (a, b, c): Six-month old infant with developmental delay, microcephaly, right choanal atresia, left ptosis, and right clubfoot who presents with episodic respiratory distress.
a: The chest radiograph demonstrates a small right lung with diminished vascularity, and hyperinflation of the left lung.
b: CT angiography at age 9 months shows absence of the right pulmonary veins and subsegmental atelectasis at the right lung base. Prominent soft tissue is noted in the mediastinum (arrow) which proved to be venous collaterals. The left pulmonary artery was incidentally noted to be duplicated (not shown).
c: Cardiac catheterization shows decreased right pulmonary arterial vascularity. Stasis was present in multiple right lower lobe venous collaterals and there was non-visualization of the right superior and inferior pulmonary veins.

pulmonary venous branches were patent. The pulmonary veins were opened proximal to the area of fibrous occlusion and a patch of main pulmonary artery was used to connect the two right pulmonary veins to the left atrium.

4. Discussion

The lungs arise from the foregut. Prior to the development of the pulmonary veins, the foregut is intimately related to the splanchnic plexus, the capillary plexus which develops into the pulmonary vascular bed. The splanchnic plexus itself, communicates with other venous systems in the embryo including the cardinal and vitelline venous systems. Early in fetal development, venous drainage of the lungs occurs via these systemic veins.

The primitive common pulmonary vein (CPV) forms as a protrusion arising from the back wall of the primitive left atrium. It undergoes canalization and subsequently fuses with the venous plexus draining the lungs. Once fusion of the CPV and the intraparenchymal venous networks of the primitive lungs occurs, the connections with the systemic venous systems are obliterated and the site of venous drainage of the lungs becomes the left atrium. The common pulmonary vein is subsequently incorporated into the left atrium with the development of four distinct pulmonary veins and ostia [1,2], although atypical anatomic patterns of venous drainage occur in 38% of the population [1].

Varying types of pulmonary vein obstructive abnormalities exist depending on the embryologic error in pulmonary vein development. If a developmental error in the relationship between the CPV and left atrium occurs early in embryologic development when connections between the splanchnic plexus and systemic venous systems exist, the result will be total anomalous pulmonary venous return. If however, the developmental error between the CPV and the left atrium occurs once the connections to the systemic vessels no longer exist, and no collateral pathways remain, the result is complete or partial pulmonary vein atresia [1,3].

Complete pulmonary vein atresia is fatal [4].Unilateral pulmonary vein atresia is rare and may be right or left sided. Fifty percent of patients have associated cardiac anomalies (usually VSD or PDA) [5]. In the setting of unilateral absence of pulmonary veins, intimal fibrosis and hypertrophy of the small pulmonary veins develop and there is reduction in their luminal diameter. Interstitial fibrosis and interlobular septal thickening may develop in the affected lung due to infection or infarction [6]. As a result of pulmonary venous obstruction and increased pulmonary venous pressure, venous collaterals develop between pulmonary and bronchial veins which allow drainage of the pulmonary venous system into the systemic system. These include submucosal bronchial varices which may be seen in the affected lung on bronchoscopy. They may be absent if other collateral channels to the azygous system are present [5].

Over time, it is hypothesized that pulmonary arterial blood flow in the affected lung diminishes due to the abnormal gas exchange secondary to ventilation-perfusion alterations in the ipsilateral lung. Additionally, preferential pulmonary arterial perfusion to the contralateral lung and ultimately flow reversal towards the contralateral lung occurs. This results in impaired growth of both the ipsilateral pulmonary artery and ipsilateral lung. This in turn leads to the development of bronchial arterial collaterals [6,7].

The onset of symptoms in cases of UCAPV is often insidious and patients may present in childhood, adolescence or rarely in adulthood [8]. The variability in the age of presentation likely reflects the balance between the systemic arterial collateral supply and the extrapulmonary venous collaterals drainage the affected lung. Symptoms include recurrent pulmonary infections, (some of which may reflect pulmonary hemorrhage secondary to rupture of bronchial varices) asthma, progressive dyspnea or hemoptysis secondary to submucosal bronchial varices. Pulmonary hypertension may also develop [6]. UCAPV is fatal in 35% of affected patients [5].

The chest radiograph in newborn infants with UCAPV is characteristically normal. Over time, radiographs demonstrate asymmetry of the pulmonary arteries with a small pulmonary artery and ipsilateral lung on the affected side. In the absence of a radiograph from the neonatal period, the radiographic findings may mimic a congenital hypoplastic lung. With progression of the disease process and reversal of flow in the ipsilateral pulmonary artery, ventilation scans will confirm absent pulmonary artery flow to the ipsilateral lung, as seen in the first case presented [6].

Typical CT findings include a small lung with hypoplasia of the ipsilateral pulmonary artery and prominence of ipsilateral bronchial arteries, absent ipsilateral pulmonary veins, increased interstitial marking with interlobular nodular septal thickening secondary to enlarged lymphatics and pulmonary veins, peribronchovascular thickening and ground glass opacities [6,9]. Parenchymal fibrosis may occur due to pulmonary venous infarction and chronic pulmonary edema. [9]

The presence of a mediastinal soft tissue “mass”, representing arterial and venous collaterals, is a common radiographic feature of this condition and has been previously reported on CT and MR [8,9] The “mass” is secondary to the primary abnormality of absent ipsilateral veins rather than cause of the absent ipsilateral veins. If the correct diagnosis of UCAPV is not recognized, as occurred in the first case we present, the identification of a mediastinal soft tissue “mass” may lead to a misdiagnosis of fibrosing mediastinitis or neoplasm occluding the pulmonary artery and veins and a lengthy patient work up may ensue. An important imaging feature in distinguishing the mass of collateral veins present in UCAPV from a slow growing neoplastic process obstructing the pulmonary veins and the pulmonary artery is its position. As in the case we present, in UCAPV, the “mass” lies adjacent to but does not surround the affected pulmonary artery and concomitant bronchial obstruction is absent [10]. Additionally, as in our case, enlarged bronchial arteries may be seen coursing through the apparent mass [8].

The presence of large bronchial arteries may prompt a cardiac catheterization which is diagnostic. In addition to demonstrating absence of the pulmonary veins, slow capillary transit, filling of small tortuous venous collaterals draining to the azygous vein and preferential flow to the contralateral pulmonary artery will be demonstrated. The pulmonary wedge pressure will be elevated on the affected side due to venous congestion and may progress to pulmonary hypertension [5]. Other imaging findings include retrograde flow in the pulmonary artery using phase contrast MR and findings suggestive of increased right ventricular pressures on echocardiography [8].

Therapeutic depend of the severity and extent of the abnormality and include observation if symptoms are few or absent, selective embolization of systemic collaterals in those patients with hemoptysis, or pneumonectomy. Pneumonectomy is preferred in patients who have developed pulmonary hypertension or hemoptysis. In those patients in whom collaterals have not yet formed and in whom the size of the pulmonary veins is accessible and they are located close to the atria, a primary repair with anastomosis of the pulmonary veins to the left atrium may be undertaken [5,8,11]. In summary we present the characteristic imaging findings and clinical presentation of UCAPV.

The congenital nature of this abnormality may not be appreciated as presentation often occurs in childhood or early adulthood. Imaging may present a confusing constellation of findings mimicking other diagnoses. Hemoptysis in a child or young adult with a small lung and small pulmonary artery in association with an apparent mediastinal “mass”, absent visualization of the pulmonary veins, and enlarged bronchial arteries should prompt evaluation for UCAPV.

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