Intrapulmonary agenesis of venous system and bronchopulmonary arterial anastomosis

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Abstract

Agenesis of the intrapulmonary and extrapulmonary veins of the right lung was found by angiography and histological examination in a three year old boy. Blood supplied by the intersegmental arteries drained via the pulmonary arteries of the right lung into main and left pulmonary arteries. This caused a shunt between the aorta and pulmonary artery. Agenesis of the pulmonary venous system as a cause of left to right shunt has not been reported before.

Several forms of pulmonary venous abnormalities, such as pulmonary vein stenosis or atresia^{1,2} and total or partially abnormal pulmonary venous connections³ to various sites, have been described. However, the complete congenital absence or agenesis of intrapulmonary and extrapulmonary veins of one lung has not been reported before. There are 17 reports of a fistulous communication between the internal mammary or intercostal artery and the pulmonary artery⁴⁻⁶; but all of these cases had normal pulmonary venous return. We report a child with complete agenesis of the intrapulmonary and extrapulmonary veins of the right lung.

Case report

CLINICAL FEATURES

A three year old boy presented with a history of recurrent lung infections since birth. There were no other cardiovascular or respiratory symptoms. Growth and development were normal. The perinatal and family histories were unhelpful.

He had normal vital signs and no cyanosis. His height and weight were in the tenth percentile. Peripheral pulses were normal with a

blood pressure of 100/50 mm Hg in the right arm. Cardiac findings included an apical impulse in the fourth left intercostal space inside the midclavicular line. No heave or thrill were present. On auscultation first and second heart sounds were normal. No systolic or diastolic heart murmurs were present. Examination of the chest showed decreased breath sounds over the right hemithorax. There was no hepatosplenomegaly. The rest of the physical examination was unremarkable.

INVESTIGATIONS

A complete blood count with differential count, sedimentation rate, urine analysis, hepatic and renal profiles, serum γ globulins, tuberculin skin test, and sweat chloride test were negative. Multiple blood and sputum cultures were sterile. The arterial blood gas profile while he was breathing room air showed pH 7.38, P_{aCO_2} 37, and P_{aO_2} 87 mm Hg.

Chest x ray

The trachea and mediastinum were shifted to the right side. The right hemidiaphragm was raised. There were diffuse infiltrates and pleural thickening in the right lung. Bronchography showed absence of the posterior segment (RB2) of the right upper lobe. The right middle lobe was normal. Except for the basal segment (RB9), the other segmental bronchi of the right lower lobe were clustered together and were much smaller in diameter and length than normal. A ventilation and perfusion lung scan showed decreased ventilation and no perfusion of the right lung. The left side was normal.

Electrocardiogram

There was left axis deviation but the electrocardiogram was otherwise normal for age. Cross sectional echocardiography with Doppler studies showed a reverse flow pattern in the right pulmonary artery and a normal cardiac anatomy.

Cardiac catheterisation

The pressures in the right and left sides of the heart were normal (table). There was a step-up in oxygen saturation in the main and left pulmonary arteries. The oxygen saturation in the right pulmonary artery was 99%. The calculated QP:QS was 1.7:1 (assumed oxygen consumption 140 ml/min). Systemic and pulmonary resistances were normal.

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Results of cardiac catheterisation

Site	Saturations (%)	Pressures (mm Hg)	Mean (mm Hg)
Superior vena cava	73		
Right atrium	66	a-7, v-5	4
Right ventricle	65	27/0-6	
Main pulmonary artery	86	23/12	18
Left pulmonary artery	80	22/12	18
Right pulmonary artery	96	24/11	18
Left ventricle	98	74/0-8	
Aorta	99	73/41	59

Assumed oxygen consumption = 140 ml/min; cardiac index = 3.4 l/min/m²; pulmonary index = 5.9 l/min/m²; QP:QS = 1.7:1; pulmonary vascular resistance (RP) = 3.0 Wood units; systemic vascular resistance (RS) = 17.6 Wood units; RP:RS = 0.17.

Figure 1 Venous phase of arterial angiograms. (A) Dye was injected into the systemic artery supplying the right lung. This showed the capillary phase and retrograde filling of the pulmonary arterial tree. Dye flowed retrogradely from the right to the left pulmonary artery. (B) Dye was injected into the main pulmonary artery. During laevophase the left pulmonary veins were seen entering the left atrium. No pulmonary veins were seen on the right side.

Cineangiography

The main pulmonary artery angiogram showed that there was a continuous flow of unopacified blood from the peripheral to the central right pulmonary artery and then into the main and left pulmonary arteries. Dye selectively injected into the distal portion of the right pulmonary artery was washed back via the central right pulmonary artery into the main and left pulmonary arteries. There was no forward flow into the capillaries and veins of the right lung. The left lung was normal. An aortic angiogram showed that systemic arteries, two from the thoracic aorta and one from the abdominal aorta, supplied the right lung. The selective angiograms from these arteries showed the flow into the capillary bed of the right lung. During the laevophase of these angiograms (fig 1) dye collected into the pulmonary arterial tree and then left the lung via the right pulmonary artery into the main and left pulmonary arteries. Neither right pulmonary nor systemic arterial injections showed a venous system from the right lung.

Surgery

Right pneumonectomy was performed. Three systemic vessels were identified: two arising from the thoracic aorta entered the hilar region and one from the abdominal aorta entered through the diaphragm into the base of the lung. No pulmonary veins, fibrous strands, or cord connected the right lung to the left atrium.

HISTOLOGY

We examined serial sections made of tissue at various levels of the formalin fixed, uninflated right lung. Systemic elastic arteries entered the right lung, two in the hilar region and one on the diaphragmatic surface of the lung. These vessels ended in a plexus of capillaries close to the airways and acini. The pulmonary arteries also anastomosed with this plexus. The muscular pulmonary arteries, predominantly in the lower lobe, showed changes—Heath and Edwards classification types I and II. An occasional artery showed plexiform lesions and venous-like dilatations. There was

Figure 2 A histological section of the right lung: (A) from the hilar region of the right lung showing no pulmonary veins and (B) from the peripheral area of the right lung showing that the bronchus and the artery had no accompanying bronchial or pulmonary veins. RPA, right pulmonary artery; SA, small artery.

no right pulmonary vein. We found no pulmonary or bronchial veins accompanying the pulmonary arteries and airways (fig 2). The airways and acini seemed to be normal except for occasional foci of early emphysematous lesions. We also found irregularly distributed, mild septal, interstitial, and pleural fibrosis.

Discussion

There are 17 case reports of systemic to pulmonary artery anastomosis; in most a right internal mammary to pulmonary artery fistula affected either a lobe or a segment of a lobe.⁴⁻⁶ These fistulas were mostly congenital. However, systemic to pulmonary artery fistulas secondary to trauma,⁶⁻⁸ inflammation,⁹ or neoplasms¹⁰ have also been described. However, in all these patients, venous drainage of the affected lobe was normal; whereas in our patient we found neither an intrapulmonary nor extrapulmonary venous system of the right lung by angiography or histology.

The venous system of the lungs develops from two different sources.¹¹ Initially, the splanchnic plexus arising from the foregut provides the venous drainage into the systemic veins for the developing pulmonary capillaries. Later, with the development of the common pulmonary vein and absorption of the systemic venous connection, pulmonary venous drainage is established into the left atrium. We speculate that in early stages of development the failure of the splanchnic system to connect with the developing pulmonary capillaries of the right lung mesenchyma resulted in the persistence of the anastomosis between the intersegmental and pulmonary

arteries. Postnatally this anastomosis led to a systemic to pulmonary artery shunt. The absence of pulmonary veins in the lung has not been reported before.

Non-invasive investigations in this child did not show the true nature of the abnormality, whereas cineangiography and cardiac catheterisation did. We recommend cineangiography and cardiac catheterisation in patients with suspected pulmonary vascular malformations.

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