Unilateral absence of the pulmonary artery and cavernous transformation of portal vein: a rare combination

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DESCRIPTION

A 32-year-old woman was referred to the liver transplant centre, for pretransplant evaluation. Her medical history revealed portal hypertension diagnosed at the age of 6 years, oesophageal varices with history of banding and sclerotherapy by the age of 12 years, as well as multiple episodes of pneumonia. Contrast-enhanced CT of the thorax was performed as part of the routine preoperative workup for liver transplant, which demonstrated unilateral absence of the right pulmonary artery (UAPA) associated with a small right lung and rightward mediastinal shift (figure 1). Additionally, numerous right subpleural cysts were present with diffuse pleural thickening (figure 1). Collateral vessels were noted along the right hemidiaphragm supplied by a single branch coming off the aorta at the level of the coeliac trunk, likely a hypertrophied right phrenic artery (figure 2). There were also prominent intercostal, bronchial and internal mammary arteries suggesting additional collateral flow (figure 3). MRI of the abdomen with and without contrast performed 1 month later revealed

Figure 2 Coronal CT with intravenous contrast (A and B) demonstrates a normal variant of the right phrenic artery (yellow arrow), which arises directly from the aorta just superior and lateral to the superior mesenteric artery. The phrenic artery is also dilated likely as a result of the congenitally absent right pulmonary artery. This is more clearly visualised on three-dimensional volume-rendering image (C).

small 'ghost' vessels within the liver, suggestive of cavernous transformation of the portal vein as well as a splenorenal portosystemic shunt (figure 4). Other MRI findings included splenomegaly and evidence of an absent portal vein (figure 4). In the absence of cirrhosis, extrahepatic portal vein obstruction or non-cirrhotic portal fibrosis were considered in the differential.¹ Congenital UAPA—found in <0.3% of the population—is a rare anomaly that results from a unilateral involution of

Figure 1 Axial CT with intravenous contrast soft tissue (A) and lung (B) windows demonstrate a congenitally absent right pulmonary artery. The main pulmonary artery is normal in size. The right lung is visually smaller than the left and there is rightward mediastinal shift. The lung window image (B) demonstrates right-sided subpleural cysts (yellow arrow) and diffuse pleural thickening. Three-dimensional volume-rendering image (C) clearly demonstrates absence of the right pulmonary artery.

Figure 3 Coronal CT with intravenous contrast (A and B) demonstrates dilated right-sided intercostal arteries (yellow arrowhead), which potentially provide arterial supply to the right lung.

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Figure 4 The coronal and axial MRI liver acquisition with volume acquisition (LAVAs): postcontrast images of the abdomen (A and B) reveal no trace of a portal vein (depicted by red arrow in A and dotted line in B), which may be due to congenital absence or from portal vein thrombosis in the paediatric years. The axial image (C) reveals a splenorenal shunt (yellow arrow). Also of note is the severely enlarged spleen, which is larger than the liver in the craniocaudal dimension (A).

the proximal sixth aortic arch.² While UAPA has been documented in association with other congenital cardiovascular anomalies, an association between UAPA and paediatric development of cavernous transformation of the portal vein has not been previously described.^{2 3}

Learning points

- Unilateral absence of the pulmonary artery is a rare foregut malformation. This possibility should be considered in the differential in individuals presenting with recurrent bronchopneumonia and/or haemoptysis. Additional anomalies should be looked for in such individuals.
- The present case highlights a unique association of unilateral absence of the pulmonary artery with development of cavernous transformation in childhood.

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REFERENCES

- 1 Khanna R, Sarin SK. Non-cirrhotic portal hypertension—diagnosis and management. *J Hepatol* 2014;60:421–41.
- 2 Pool P, Vogel J, Blount S Jr. Congenital unilateral absence of a pulmonary artery. The importance of flow in pulmonary hypertension. *Am J Cardiol* 1962;10:706–32.
- 3 Newman B. Congenital bronchopulmonary foregut malformations: concept and controversies. *Pediatr Radiol* 2006;36:773–91.

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