



Unilateral proximal interruption of a pulmonary artery

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A previously healthy, 43-year-old woman presented to the emergency department with dry cough and hypoventilation at the right lung base. A chest X-ray showed a small right lung with an inconspicuous hilum and mediastinal shift (Figure 1A). Chest CT angiography allowed the diagnosis of proximal interruption of a pulmonary artery (PIPA) on the right side, with multiple collateral vessels supplying the ipsilateral lung (Figure 1B-D). Her echocardiogram was unremarkable, and no other abnormality was found.

PIPA is a rare vascular anomaly with an estimated prevalence of 1 in 200,000 people.⁽¹⁾ The term interruption is preferred to the term absence because the

intrapulmonary arteries remain intact despite the blind end of the ipsilateral pulmonary artery at the hilum.⁽²⁾ Right-sided PIPA tends to be an isolated finding and is more common than left-sided PIPA, which is usually associated with other congenital cardiovascular anomalies.⁽¹⁾

Oxygenated blood is supplied to the lung through collateral systemic vessels, such as bronchial arteries and transpleural branches of the internal mammary, intercostal, subclavian, and innominate arteries,⁽³⁾ or directly from the aorta.

Clinical presentation can be asymptomatic or include dyspnea, chest pain, recurrent infections, and

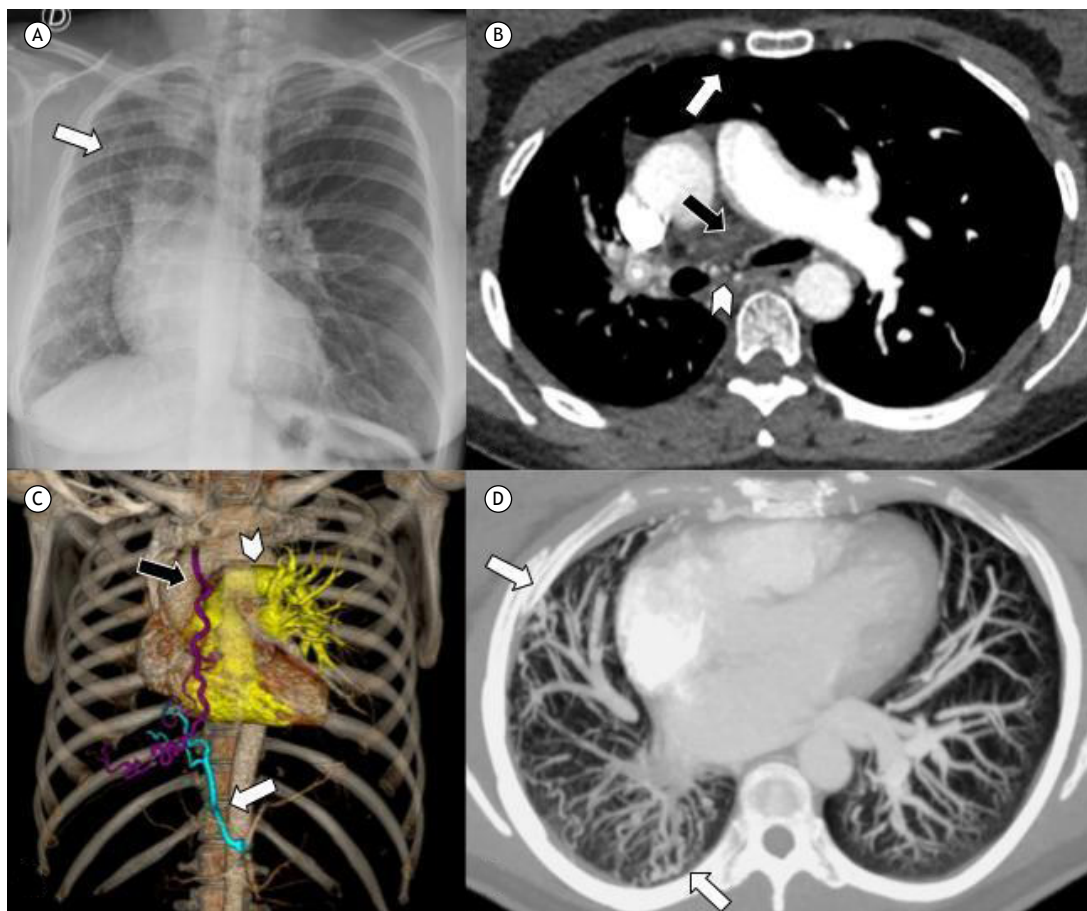


Figure 1. In A, chest X-ray showing an inconspicuous right hilum, a small right lung with reticular opacities, mediastinal shift, and a hyperinflated left lung with herniation to the right hemithorax, represented by a displaced anterior junction line (arrow). In B, axial CT angiographic image showing interruption of the right pulmonary artery (black arrow), as well as hypertrophied bronchial arteries (arrowhead) and right internal mammary artery (white arrow). In C, three-dimensional reconstruction showing the dilated right internal mammary artery (in purple; black arrow), a branch of the celiac artery (in light blue; white arrow), and a normal left pulmonary artery (in yellow; arrowhead). In D, maximum intensity projection showing multiple tortuous transpleural vessels (arrows) throughout the right lung.

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hemoptysis.^(1,3) Pulmonary hypertension is the most feared complication.

Chest X-ray and CT findings can lead to the correct diagnosis. Awareness of this anomaly is important

in order to rule out alternative diagnoses and allow appropriate management.

CONFLICTS OF INTEREST

None declared.

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