Isolated Unilateral Pulmonary Artery Agenesis

Eva Perez-Pampin 1, Joaquin Campos-Franco 2, Anxo Martinez-de Alegria 3 and Pedro Rigueiro 4

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A 33-year-old woman with a history of psoriasis presented with asymmetric oligoarthritis. She had no cardiovascular symptoms. Naproxen and steroids were started with the diagnosis of psoriatic arthritis. Routine chest X-ray showed a small left hemithorax with a right-sided aortic arch and decreased left-sided pulmonary vasculature. A CT-scan (Picture A) demonstrated the absence of the left pulmonary artery (PA). A 3D CT angiography with volume rendering revealed a hidden PA at the hilum (black arrow) and collateral circulation provided by enlarged bronchial arteries, and by the left internal mammary artery (Picture B, white arrow) and the left subclavian artery (Picture C, white arrow). Transthoracic echocardiography excluded other heart anomalies. To date, she remains asymptomatic.

Unilateral agenesis of a PA is a rare anomaly with an estimated prevalence of 1 in 200,000 (1, 2). CT and MRI facilitates the diagnosis, reserving angiography for selected cases that require embolization or revascularization surgery (2).

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References