Case report

Unilateral pulmonary artery agenesis

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Case

A middle aged male presented with a one-week history of progressively worsening dyspnea upon exertion, with no prior respiratory symptoms. Contrast-enhanced computed tomography of the chest showed multiple collateral vessels within the expected region of the left main pulmonary artery with absence of the left main pulmonary artery and hypoplasia of the left lung and compensatory hyperinflation of the right lung, as well as a patent ductus arteriosus (Fig. 1A). Ventilation-perfusion scintigraphy demonstrated normal ventilation in both with the absence of perfusion in the left lung (Fig. 1B). Unilateral pulmonary artery agenesis (UPAA) is a very rare congenital condition, with approximately 150 cases reported since 1868. Although the disease can be associated with other congenital cardiovascular abnormalities, isolated UPAA may not be diagnosed until adulthood, presenting as dyspnea and/or with abnormal chest imaging [1]. Treatment options can include pneumonectomy, closure of collateral arteries or revascularization. It is suggested to follow asymptomatic patients with echocardiography to monitor for early development of pulmonary hypertension, which would then require a cardiac catheterization to measure pulmonary artery pressures. Unfortunately, there are no guidelines or consensus regarding treatment for these patients [2].
Conflicts of interest

The author declares that no conflicts of interest exist.

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Author contributions

ASS, JMS and PJ: conception and design, acquisition of radiological and pathological data, drafting the article, critical revision of intellectual content and final approval of the version to be published.

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References
