

Unmasking of Agenesis of Right Pulmonary Artery with Unilateral Absence of Perfusion on Lung Scintigraphy

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We present a rare finding on lung ventilation–perfusion (V/Q) scintigraphy for a woman with longstanding dyspnea. CT of the chest showed volume loss on the right side, which raised concern about possible bronchiolitis obliterans or Swyer–James–MacLeod syndrome; however, the right pulmonary artery could not be visualized. A subsequent V/Q scan showed absence of perfusion and decreased ventilation to the entire right lung, consistent with agenesis of the right pulmonary artery. The patient’s clinical course and imaging features mimicked Swyer–James–MacLeod syndrome, which usually presents with a matched perfusion defect in a single lung or lobe on V/Q scanning. This case highlights the importance of a multimodality imaging approach to achieve a diagnosis.

Key Words: pulmonary artery agenesis; ventilation–perfusion scintigraphy; Swyer–James–MacLeod syndrome; CT

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Right pulmonary artery agenesis is a rare developmental anomaly of the pulmonary artery trunk and is usually accompanied by unilateral pulmonary hypoplasia (1). Many of the reported cases are discovered in early childhood; however, a few have been reported in adults. Although infant patients may present with respiratory distress or be asymptomatic, adult patients may have variable nonspecific manifestations, that is, chest pain, dyspnea, recurrent pulmonary infection, and even hemoptysis. The clinical presentation mimics that of Swyer–James–MacLeod syndrome (SJMS) and can lead to misdiagnosis. We report the classic lung ventilation–perfusion (V/Q) scintigraphic and accompanying radiographic findings in a woman with agenesis of the right pulmonary artery who had been symptomatic since childhood and had been misdiagnosed as having SJMS.

CASE REPORT

A 59-y-old woman with a past medical history notable for stage 3b chronic kidney disease and mitral regurgitation secondary to childhood rheumatic disease presented to the pulmonology clinic with longstanding dyspnea. Pulmonary function testing revealed decreased functional capacity. The

patient reported a history of chronic asthma, exertional dyspnea since childhood, recurrent pulmonary infections, and a diagnosis of SJMS. Chest radiography revealed an elevated right hemidiaphragm, a right tracheal deviation, and a mediastinal shift (Fig. 1). Non-contrast-enhanced chest CT revealed early fibrotic changes, architectural distortion, and absence of the right pulmonary artery (Fig. 2). A lung V/Q scan revealed absence of perfusion to the entire right lung and decreased ventilation (Fig. 3). A diagnosis of right pulmonary artery agenesis was established on the basis of the imaging features and clinical manifestations.

DISCUSSION

Agenesis of the right pulmonary artery is a rare congenital anomaly caused by failure of the primitive sixth aortic arch to migrate and rotate (2). The overall mortality rate is 7%, with 30% of adult patients having no symptoms (3). Asthma and recurrent respiratory tract infections unresponsive to conventional treatment are commonly seen, with hemoptysis seen in 10% of adult patients (4). Treatment is based on the severity of symptoms. Surgical intervention with revascularization and pneumonectomy can be considered in cases of recurrent hemoptysis, recurrent pulmonary infections, and pulmonary hypertension (4).

CT angiography of the chest is the imaging modality of choice for diagnosis and might be able to depict the supplying aberrant right bronchial artery (5,6). This modality may not be plausible in all patients because of concurrent comorbidities. Non-contrast-enhanced chest CT may demonstrate pulmonary parenchymal changes, in addition to the nonvisualization of the pulmonary artery. On lung V/Q scanning, right-sided pulmonary artery agenesis presents with global absence of right lung perfusion and normal

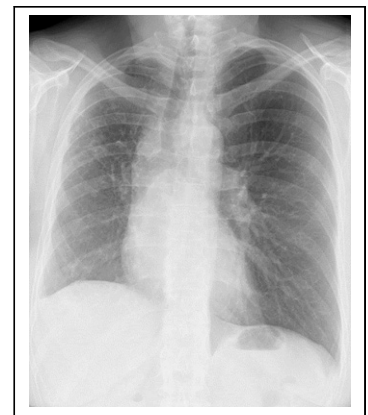


FIGURE 1. Chest radiograph (anterior view) depicts elevated right hemidiaphragm, right tracheal deviation, and mediastinal shift.

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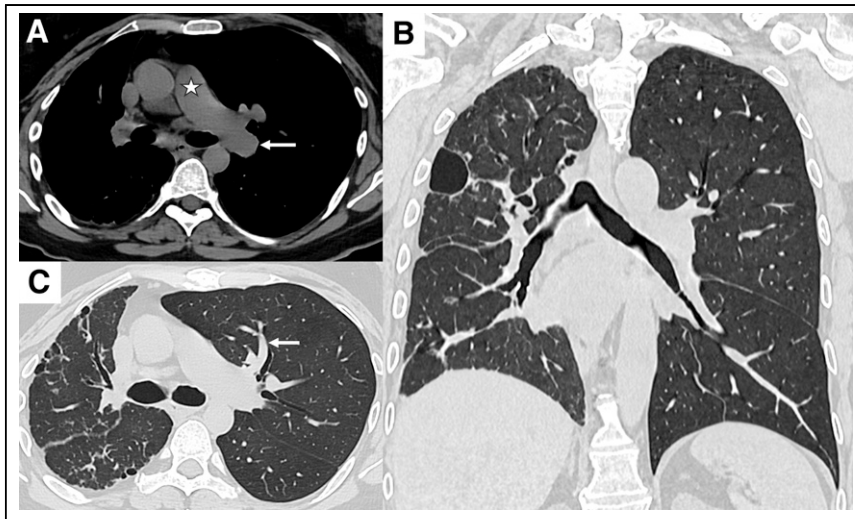


FIGURE 2. Non-contrast-enhanced CT of chest. (A) Soft-tissue window image shows pulmonary trunk (star) and left pulmonary artery (arrow) but not right pulmonary artery. (B and C) Coronal (B) and axial (C) lung window images show subpleural bullae, peribronchial thickening, decreased right lung volume, and left-side peribronchial pulmonary artery (arrow).

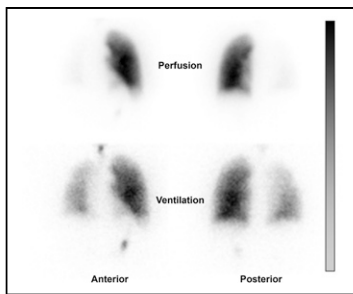


FIGURE 3. V/Q scan reveals complete absence of perfusion to right lung (top), with homogeneously decreased ventilation (bottom).

or diminished ventilation secondary to hypoplasia of the right lung. The mismatched global perfusion defect in the entire unilateral lung differs from matched perfusion defects observed in SJMS due to pulmonary artery hypoplasia as a sequela of bronchiolitis obliterans. Furthermore, the imaging features and clinical presentation do not support

the diagnosis of pulmonary embolism (7,8).

CONCLUSION

This case demonstrates the value of lung V/Q scanning in the diagnosis of unilateral agenesis of the pulmonary artery. The unique absence of perfusion to the entire lung should be distinguished from SJMS. Given the inconsistency between the patient's clinical presentation and pulmonary embolism, and the accompanying imaging features on chest CT, the diagnosis of unilateral pulmonary artery agenesis was achieved.

DISCLOSURE

No potential conflict of interest relevant to this article was reported.

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