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Case report

A rare combination of cardiopulmonary anomalies demonstrated on ventilation-perfusion scan

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Abstract

In this case report we describe an unusual appearance seen on a ventilation-perfusion (V/Q) scan in a woman with pulmonary hypertension. Although the pulmonary hypertension was not caused by pulmonary emboli, the V/Q scan suggested several cardiac anomalies which may lead to pulmonary hypertension. Most of the cardiac anomalies, including right-sided aortic arch and right-to-left shunt, can be deduced from careful examination of the V/Q scan. A subsequent cardiac MRI scan confirmed the anomalies.

Key words: V/Q scan, pulmonary hypertension

Introduction

Isolated unilateral agenesis of a pulmonary artery is rare and is usually encountered as an associated condition in 0.39% of congenital heart diseases, such as right-sided aortic arch, septal defects, truncus arteriosus and tetralogy of Fallot [1].

In this report we describe a patient who was referred to nuclear medicine for ventilation-perfusion scintigraphy for further investigation of pulmonary hypertension. This revealed evidence of a right-to-left shunt, a right sided aortic arch and a small left lung. These findings were all confirmed on subsequent CT and MR imaging, which also revealed the absence of a left pulmonary artery.

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Case report

A 46-year-old woman diagnosed with perimembranous ventricular-septal defect (VSD) and severe pulmonary hypertension on echocardiography was referred to the nuclear medicine department for further investigation. Initially, a chest radiograph had demonstrated a mediastinal shift to the left, a small left hemithorax, raised left hemidiaphragm, cardiomegaly and a right-sided aortic arch (Figure 1).

Ventilation-perfusion scintigraphy using Tc^{99m} MAA and $Krypton^{81m}$ was performed to exclude thromboembolic disease as the cause for the pulmonary hypertension. This revealed decreased tracer uptake in both ventilation and perfusion scans in the left lung

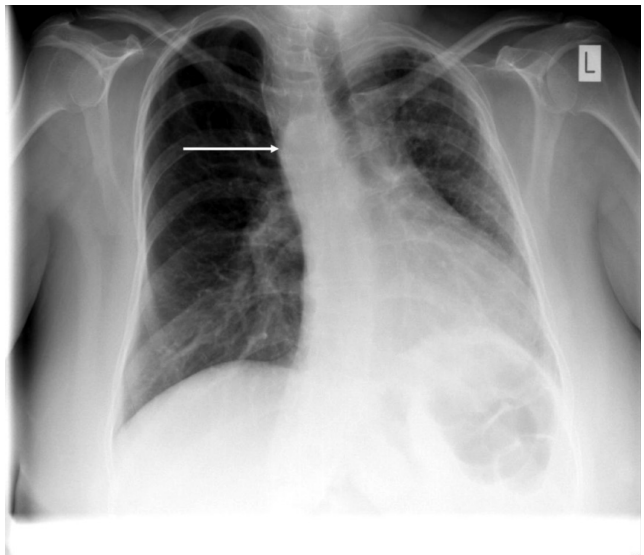

Figure 1. Chest radiograph demonstrating mediastinal shift to the left, a small left hemithorax, raised left hemidiaphragm, tracheal deviation to the left and a right-sided aortic arch (arrow).

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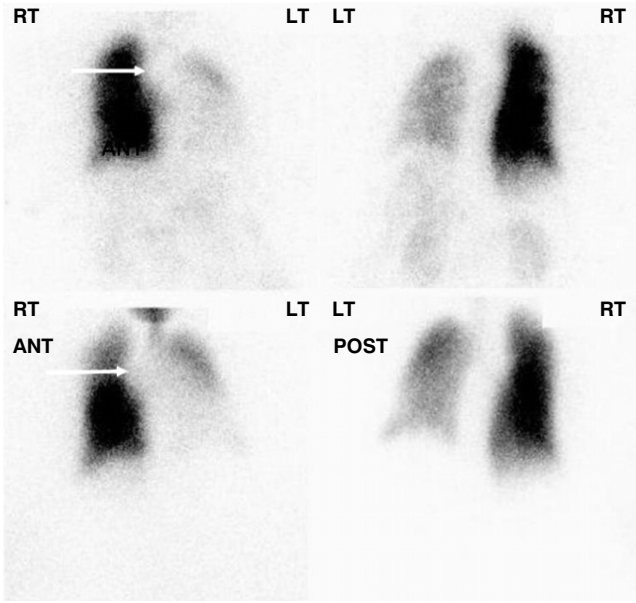

Figure 2. Ventilation-perfusion scintigraphy (perfusion images above, ventilation images below) demonstrating a small left lung with decreased tracer uptake in both anterior and posterior views, evidence of a right-sided aortic arch (arrow) and uptake bilaterally in the kidneys indicating the presence of a right-to-left shunt.

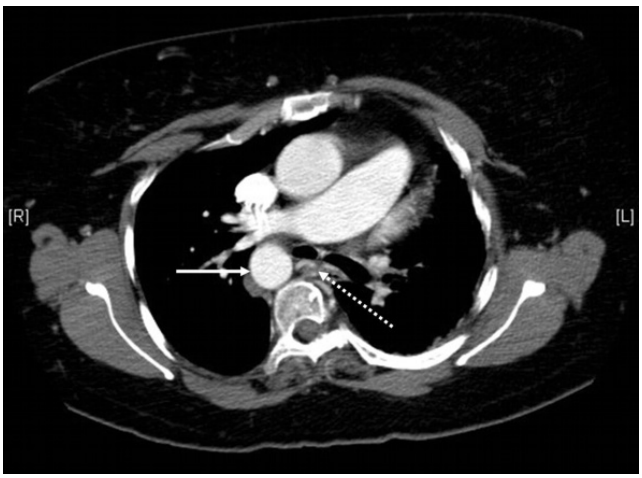

Figure 3. A contrast enhanced CT image demonstrating an absent left main pulmonary artery, right sided aorta (solid arrow) and hypertrophied bronchial arteries (dotted arrow).

field, a small left lung, evidence of a right-sided aortic arch and bilateral tracer uptake in the kidneys, indicating the presence of a right-to-left shunt (Figure 2). There was no evidence of pulmonary embolic disease.

Subsequent CT pulmonary angiogram (CTPA) confirmed the presence of pulmonary hypertension; however, there was no evidence of pulmonary thromboembolic disease. In addition, the scan highlighted the absence of the left pulmonary artery with its sequel including hypertrophied bronchial arteries (Figure 3).

An MRI cardiac study was also performed. This demonstrated the presence of a large semi-membranous VSD posterior and inferior to the crista supraventricularis (Figure 4). Again, the absence of a left-sided pulmonary artery was confirmed together with small volume left lung and compensatory increase in the diameter of the right pulmonary artery. The left lung is supplied only via bronchial arteries and hypertrophied left-sided intercostal arteries.

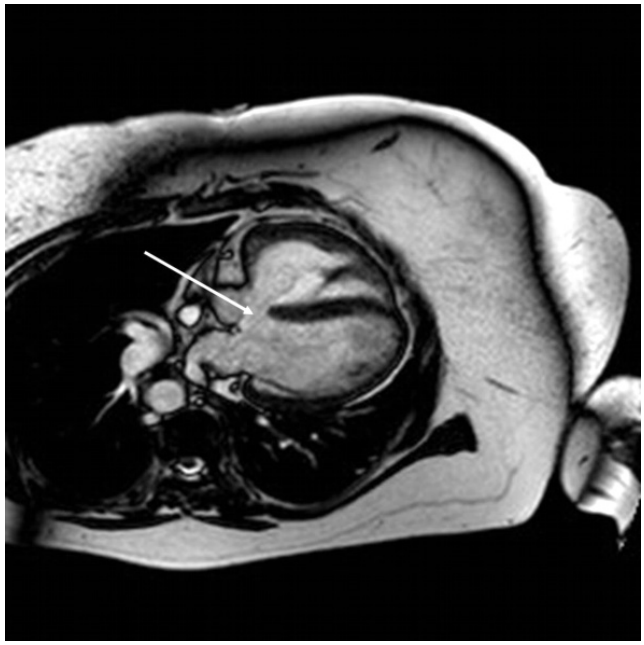

Figure 4. A horizontal long axis cardiac MR image demonstrating the membranous VSD (arrow) with a hypertrophied right ventricle.

Discussion

Congenital unilateral absence of a pulmonary artery is a rare anomaly; a literature review by Ten Harkel et al. [2] revealed only 108 reported cases between 1978 and 2000, more frequently occurring with associated cardiac lesions such as tetralogy of Fallot or septal defects. The most commonly reported symptoms include recurrent respiratory infections, decreased exercise tolerance, dyspnoea on exertion and, less frequently, haemoptysis [2]. Despite this, some patients may remain asymptomatic for a long time, making the prevalence of the absence of a pulmonary artery difficult to estimate accurately. Occasionally, cases may only be recognised by predisposing factors for pulmonary hypertension such as pregnancy or high altitude [3], or as an incidental finding of the characteristic features on chest radiography as first described in 1954 [4].

There is a variety of diagnostic approaches for patients with absence of pulmonary arteries. In the first instance a chest radiograph should be performed, which may show cardiac and mediastinal displacement or absence of the pulmonary arterial shadow; smaller hemithorax; elevation of the hemidiaphragm; and paucity of vascular markings, on the involved side. On the opposite side, there may be hyperinflation and herniation of the lung across the midline [4]. A subsequent echocardiogram may confirm the diagnosis and highlight any other cardiovascular abnormalities. Pulmonary hypertension may also be diagnosed.

Ventilation perfusion scintigraphy has been described in the non-invasive diagnosis of pulmonary artery agenesis in subjects

with hyper lucent lungs [5, 6]. In patients with unilateral absence of a pulmonary artery, there is usually a complete absence of perfusion on the affected side. The presence of reduced tracer uptake in the affected lung of the patient on the perfusion scan can be explained by the compensatory blood supply provided by the bronchial artery, as confirmed on MRI. The presence of compensatory blood supply maintained by the bronchial artery circulation or pulmonary veins has been described previously [7].

With the ever-increasing availability of high resolution CT and Cardiac MRI, these tests are now more extensively used in the evaluation of congenital heart defects. In the scenario that revascularization is proposed, cardiac catheterisation is also vital; including pulmonary venous wedge angiography to discover hilar arteries [2]. Ventilation perfusion scintigraphy should be considered in all patients with pulmonary hypertension to exclude pulmonary emboli. In addition, the V/Q scan may indicate another rarer cause of the pulmonary hypertension, such as this case.

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