

An interrupted right pulmonary artery with stenotic right pulmonary veins

Stenotik sağ pulmoner venler ile sağ pulmoner arter kesintisi

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ABSTRACT

Interrupted pulmonary artery is a very rare congenital abnormality. It may be detected at an early age or in adulthood. It may be associated with congenital cardiovascular defects. However, it is usually isolated and detected incidentally in adults. In this article, we present clinical and radiological findings of a 50-year-old female patient with interrupted right pulmonary artery and stenosis of the inferior pulmonary veins in the affected lung, in light of the literature review.

Keywords: Interrupted pulmonary artery; pulmonary vein stenosis; radiological findings.

Unilateral interruption of the pulmonary artery (formerly known as the absent pulmonary artery) is rare and may be associated with cardiac anomalies. However, it can sometimes be an isolated finding. Pool et al.^[1] studied 78 patients with an absent pulmonary artery and found that approximately 40% of the cases had an isolated anomaly. In this case report, we present the clinical and computed tomography (CT) findings of a patient with an interrupted right pulmonary artery and stenosis of the right inferior pulmonary veins. To the best of our knowledge, this is the first case of pulmonary vein stenosis (PVS) in association with an interrupted pulmonary artery in the literature.

CASE REPORT

A 50-year-old woman with a backache, chest pain, and shortness of breath was referred to our clinic. She

ÖZ

Pulmoner arter kesintisi çok nadir bir doğuştan anomalidir. Küçük yaşlarda veya yetişkinlikte saptanabilir. Doğuştan kardiyovasküler defektler ile ilişkili olabilir. Ancak genellikle izoledir ve yetişkinlerde tesadüfen saptanır. Bu yazıda, sağ pulmoner arter kesintisi ve etkilenen akciğerde alt pulmoner venlerde darlık olan 50 yaşında bir kadın hastanın klinik ve radyolojik bulguları literatür incelemesi ışığında sunuldu.

Anahtar sözcükler: Pulmoner arter kesintisi; pulmoner ven darlığı; radyolojik bulgular.

had been receiving asthma treatment for five years, and chest auscultation revealed decreased breathing sounds. A chest X-ray showed an elevated right hemidiaphragm, an asymmetrically small right lung, shifting of the mediastinum, and hyperinflation of the left lung. On contrast-enhanced multislice computed tomography (Aquilion™ 64, Toshiba Medical Systems, Otawara-shi, Tochigi-ken, Japan), the right lung had a small volume, and only the small proximal portion of the right pulmonary artery was visible. The pulmonary truncus and left pulmonary artery were normal. In addition, the right intercostal arteries were prominent, and a right infradiaphragmatic branch arising from the celiac trunk supplied the right lung (Figure 1). Furthermore, the right inferior pulmonary veins were stenotic (Figure 2). In the lung window settings, there was widespread ground glass opacity and peripheral



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Figure 1. Contrast enhanced coronal three-dimensional volume-rendered image shows the interrupted right pulmonary artery and the collateral artery (arrow) arising from the celiac trunk and supplying the right lung.

reticulations in the mid and lower lobes of the right lung, and peribronchial thickening and bronchiolectasis were also present in the posterior basal segment.



Figure 2. Axial maximum intensity projection image shows the hypoplastic inferior pulmonary veins in the right lung.

Moreover, fissures were diffusely thickened in the right lung. After questioning the patient, she revealed that she had been evaluated via pulmonary angiography nine years earlier and that an absent pulmonary artery had been diagnosed (Figure 3). Next, transthoracic echocardiography was performed, and no pulmonary hypertension or cardiac anomalies were detected.

DISCUSSION

Involution of the proximal sixth aortic arch, may result in an absent extrapulmonary pulmonary artery. The intrapulmonary arteries are usually intact and have normal distribution. The mediastinal portion of the affected pulmonary artery may be totally absent or may terminate within 1 cm of its origin.^[2]

An interrupted pulmonary artery may be a single anomaly, or it can be associated with congenital cardiovascular defects. Ventricular and atrial septal defects, tetralogy of Fallot, coarctation of the aorta, a right aortic arch, transposition of the great arteries, subvalvular aortic stenosis, mitral valve prolapse, and patent ductus arteriosus have been reported with this condition, which is usually asymptomatic. However recurrent pulmonary infections, shortness of breath, hemoptysis, high-altitude pulmonary edema, congestive heart failure, and pulmonary hypertension may occur.^[3]

The affected lung parenchyma is supplied by collaterals from the bronchial, intercostal, internal



Figure 3. Pulmonary artery digital subtraction angiography shows the absent right pulmonary artery and normal left pulmonary artery.

mammary, subclavian, innominate, or the celiac axis.^[3] Transpleural branches of the collateral arteries may appear as multiple linear opacities perpendicular to the pleural surface, and serrated pleural thickening may be detected on soft tissue window settings because of the enlarged intercostal collaterals. Furthermore, ground glass attenuation, reticular opacities, septal thickening, subpleural consolidation, cystic lung changes, pleural thickening, bronchial dilation, and bronchial wall thickening has been detected on high-resolution CT of the lung parenchyma of the interrupted pulmonary artery.^[2,3] Unlike Swyer-James syndrome, no trapped air is visible on the expiratory chest radiograph with this condition. The normal bronchial branching pattern differentiates it from hypogenetic lung syndrome. Additionally, the causes of the acquired pulmonary artery occlusion (i.e., a chronic pulmonary thromboembolism, fibrosing mediastinitis, or Takayasu's arteritis) should be present in the differential diagnosis of the interrupted pulmonary artery.^[2]

Pulmonary vein stenosis is an extremely rare abnormality with a prevalence rate of 1.7 per 100,000 in children younger than two years old.^[4] It can be either primary or secondary in nature, and there is a belief that the primary form stems from the abnormal incorporation of the common pulmonary vein into the left atrium in the later stages of cardiac development.^[5] Pulmonary vein stenosis may be focal, or it can involve a long segment of one or more pulmonary veins.^[6] Acquired cases of PVS may be the result of mediastinal neoplasms, sarcoidosis, or fibrosing mediastinitis, but this condition can also arise after anomalous pulmonary vein surgery or radiofrequency ablation procedures for atrial fibrillation. Increased back pressure proximal to the stenosis decreases the ipsilateral pulmonary artery flow and gas exchange in the affected lung,^[4] leading to

the eventual appearance of the hypoplastic pulmonary artery along with the ipsilateral PVS.

In conclusion, while the absence of the pulmonary artery in combination with different cardiac anomalies has previously noted, the literature has made no mention of a case with PVS accompanied by this rare anomaly.

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