Unilateral absence of the left pulmonary artery with patent ductus arteriosus and interrupted inferior vena cava

Sol pulmoner arterin tek taraflı yokluğu ile birlikte patent duktus arteriyozus ve kesintili inferior vena kava

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Summary– Unilateral absence of the pulmonary artery and interrupted inferior vena cava are rare congenital disorders. The clinical presentation is variable, and many patients can be asymptomatic for many years and even throughout their lives. We report the case of a 44- year-old female patient with a history of hemoptysis. She was referred to our clinic with a diagnosis of pulmonary artery agenesis. Computed tomography revealed absence of the left main pulmonary artery and long-tract patent ductus arteriosus (PDA). Blood supply to the left lung was provided by major aortopulmonary collateral arteries (MAPCAs). Right heart catheterization through the right femoral vein was problematic, as the catheter could not be negotiated from the inferior vena cava to the right atrium. We revealed the interrupted inferior vena cava, which continued as a dilated azygos vein to the superior vena cava. The magnitude of the shunt flow from the PDA was too small, and the calculated shunt fraction was not significant. Hemoptysis was possibly due to MAPCAs. However, as the patient had no active bleeding when she was referred, the exact source of the hemoptysis could not be identified.

Özet– Tek taraflı pulmoner arter yokluğu ve kesintili inferior vena kava nadir rastlanılan konjenital anomalilerdir. Klinik tablo değişkendir. Birçok hastanın yıllar boyunca ve hatta hayat boyu hiçbir yakınması olmayabilir. Bu yazıda, 44 yaşında hemoptizi öyküsü olan bir kadın hasta sunuldu. Hasta kliniğimize pulmoner arter agenezisi tanısıyla gönderilmişti. Bilgisayarlı tomografi sol pulmoner arterin olmadığını ve uzun patent duktus arteriyozus (PDA) varlığını göstermekteydi. Sol akciğerin kanlanması majör aorto pulmoner kollateral arterler (MAPCAs) ile sağlanmaktaydı. Sağ femoral ven yoluyla yapılan sağ kalp kateterizasyonu zorlu geçti; kateter inferior vena kavadan sağ atriyuma ilerletilemedi. Genişlemiş bir azigos veni yoluyla superior vena kavaya bağlanan kesintili inferior vena kava bulunduğu belirlendi. PDA şant akımı oldukça az olup şant oranı kayda değer değildi. Hemoptizinin muhtemelen MAPCAs kaynaklı olduğu düşünüldü. Ancak, hastanın hastanemize yönlendirildiği dönemde aktif kanaması yoktu ve hemoptizinin kesin kaynağı belirlenemedi.

The unilateral absence of a pulmonary artery (UAPA) is a rare congenital anomaly and is frequently associated with other cardiovascular anomalies, such as tetralogy of Fallot, septal defects, right aortic arch, and patent ductus arteriosus (PDA).[1,2] Many patients with isolated UAPA usually survive into adulthood experiencing only minor, if any, symptoms, while about 20% of the patients with UAPA develop inconsequential hemoptysis.

Abbreviations:

CT	*Computed tomography*
IVC	*Inferior vena cava*
MAPCAs	*Multiple major aortopulmonary collateral arteries*
PDA	*Patent ductus arteriosus*
SVC	*Superior vena cava*
UAPA	*Unilateral absence of a pulmonary artery*

We describe a case presenting with hemoptysis. To the best of our knowledge, this is the first time the combination of unilateral agenesis of the pulmonary artery, interruption of the inferior vena cava (IVC) with azygous continuation, PDA, and right aortic arch have been reported in one patient.

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CASE REPORT

A 44-year-old female with a history of two episodes of hemoptysis in the last two months was referred to our outpatient clinic from the pneumology department of another hospital with a diagnosis of pulmonary artery agenesis documented with computed tomography (CT), which revealed absence of the left pulmonary artery and multiple major aortopulmonary collateral arteries (MAPCAs) as a possible cause of hemoptysis. The patient was in good general condition except for mild dyspnea and coughing. No active bleeding was seen. She had never been a cigarette smoker. Her physical development was normal, and there was no family history of congenital cardiovascular disease.

At the initial evaluation, she looked pale. There were no clinical signs of edema, cyanosis or clubbing of fingers. The cardiac auscultation and physical examination were unremarkable. Hemodynamic parameters were stable. Routine hematological and biochemical profiles were within normal ranges except for a hemoglobin level of 6.8 mg/dl. Further blood tests showed severe iron deficiency. She had no active bleeding, and hemoptysis history did not comply with massive hemoptysis. She had not required hospitalization or blood transfusion. We thus thought that the severe anemia could not be explained solely by hemoptysis.

Computed tomography (CT) of the chest revealed a right-sided aortic arch, absence of the main left pulmonary artery and long tract PDA (Figure 1). Blood supply to the left lung was provided by MAPCAs.

Echocardiography documented the absence of the left pulmonary artery; however, a PDA could not be shown. The systolic pulmonary artery pressure was estimated to be approximately 40 mmHg.

Right heart catheterization through the right femoral vein was performed to evaluate the severity of PDA and measure the pulmonary artery pressure. The catheter could not be negotiated from the IVC to the right atrium. Injection of contrast agent into the IVC showed hepatic interruption of the IVC. The IVC continued as a dilated azygos vein, which was draining into the superior vena cava (SVC) (Video 1*). The SVC was normal. On manipulation, the catheter was propagated from the IVC to the SVC via the azygos vein and from there to the right atrium and right ventricle (Figure 2a). Because of the sharp angulation of the catheter at the SVC entry, advancement to the pulmonary artery was difficult. The pulmonary artery pressures were 35/5 mmHg /mean 23 mmHg.

The patency of the ductus arteriosus was demonstrated by both aortography and non-selective angiography with a right catheter. The ductus had a conical shape with a large aortic end tapering into the small constricted pulmonary connection (Video 2*). The magnitude of the shunt flow was too small, and calculated shunt fraction was not significant (shunt fraction=1.12).

Multiple MAPCAs originating from the descending aorta and supplying the left lung were identified by selective angiography (Figure 2b).

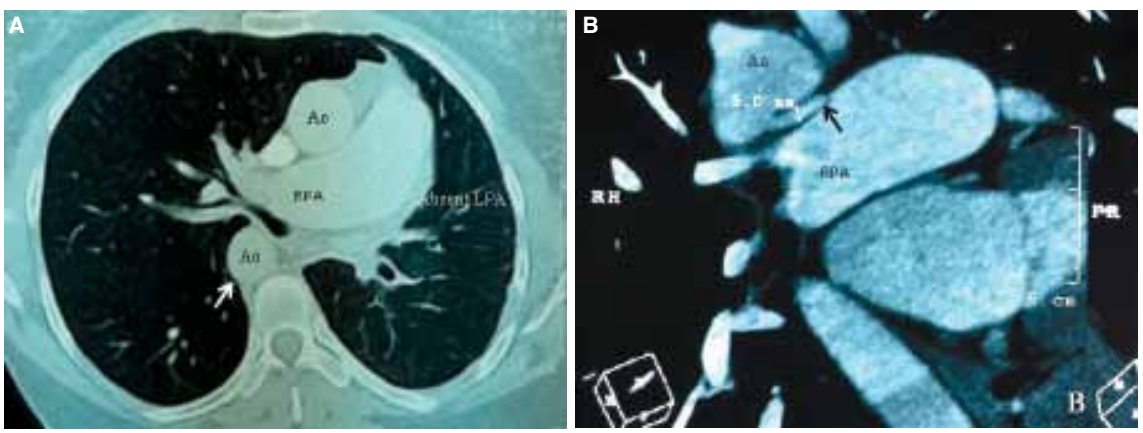

Figure 1. CT scan of the chest shows: **(A)** absence of the main left pulmonary artery and right-sided aorta (white arrow). The main pulmonary artery measures 36 mm. **(B)** Long tract patent ductus arteriosus (black arrow, width: 5 mm). Ao: Aorta, RPA: right pulmonary artery, LPA: left pulmonary artery.

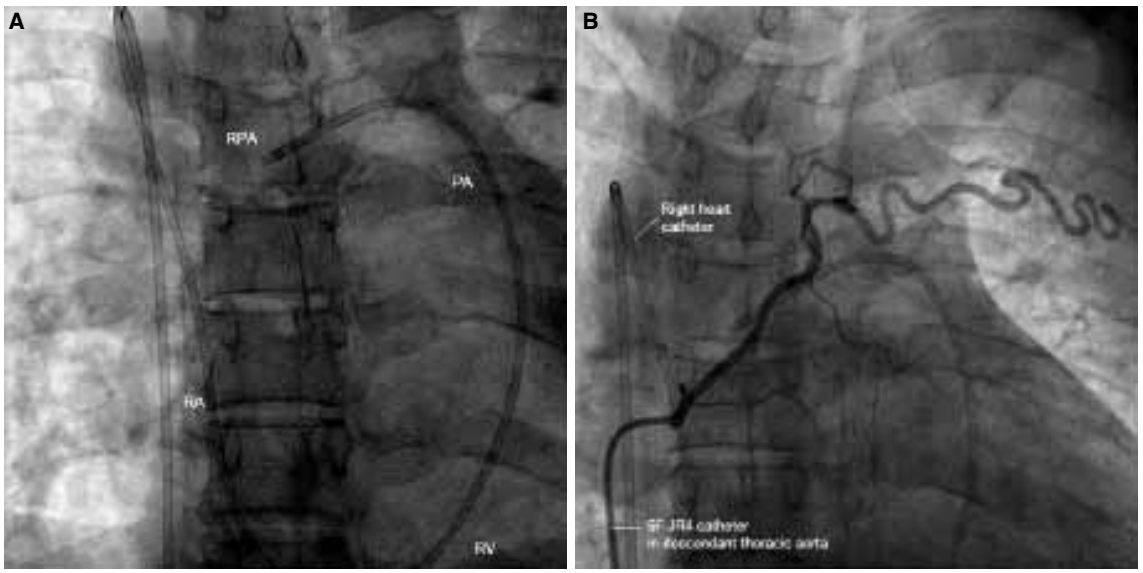

Figure 2. **(A)** Radiograph showing a catheter at the pulmonary artery with an unusual loop because of an interrupted inferior vena cava. **(B)** Selective arteriogram in the anteroposterior view showing a MAPCA originating from the descending aorta.

Since there were no abnormalities in the lung parenchyma and bronchial tree on CT, the source of hemoptysis was thought to be the MAPCAs. The affected lung receives its blood supply through numerous collaterals; therefore, identification of the collateral artery responsible for hemoptysis was not possible. Following a consultation with the thoracic surgeons and interventional radiologists, it was decided to avoid any interventional therapy. She was referred to gastroenterology and hematology departments for investigation of the anemia. She has been asymptomatic without recurrent hemoptysis for one year since discharge.

DISCUSSION

Unilateral agenesis of the pulmonary artery represents a rare developmental anomaly due to a failure in the connection of the sixth aortic arch with the pulmonary trunk.[1] Intrapulmonary vessels and the distal portion of the affected pulmonary artery trunk can develop normally, and blood supply is achieved by systemic collaterals from bronchial, major aortopulmonary collaterals and other systemic arteries.[3] Quiet a few patients appear to remain asymptomatic until adulthood. When symptomatic, patients with isolated UAPA may experience recurrent respiratory infections, dyspnea on exertion, high-altitude pulmonary edema, pulmonary hypertension in the contralateral lung, or hemoptysis.[2,4,5]

Hemoptysis has been described as a clinical symptom in up to 20% of patients from either rupture of hypertrophied bronchial collateral vessels or peripheral arteriovenous fistulas ipsilateral to the pulmonary artery, as well as from rupture of chronically hyperperfused vessels on the contralateral side.[2,6]

On the other side, infrahepatic IVC interruption is a well-recognized but uncommon developmental anomaly resulting in termination of the IVC below the hepatic vein. Its prevalence is 0.6-2.0% in patients with congenital heart disease.[7] During embryogenesis, the IVC is composed of the hepatic, prerenal, renal, and postrenal segments, which by segmental fusion, regression, and midline anastomosis form the IVC.[8] Interruption of the IVC results from agenesis of a segment of the IVC or a fusion defect between the prerenal and hepatic segment of the IVC. The infrahepatic IVC may continue as the azygos vein,[8,9] as in the present case, or may continue as the hemiazygos vein to the left SVC, intrathoracic veins, or anomalous intrahepatic veins. The hepatic segment of the IVC drains directly into the right atrium, as in this case.

This anomaly is usually an incidental finding but may be problematic in certain circumstances. It can

pose technical challenges during invasive procedures, such as right heart catheterization procedures, electrophysiological procedures, IVC filter placement, temporary pacing through the transfemoral route, and abdominal or thoracic surgery. This anomaly may be associated with recurrent deep venous thrombosis of the lower limbs, sick sinus syndrome, and atrial flutter.

From a treatment perspective, neither the interrupted IVC nor PDA required any treatment. Hemoptysis was possibly due to MAPCAs. A therapeutic option in such a case may be a percutaneous or surgical occlusion of the collateral vessels or pneumonectomy of the affected side to avoid hemoptysis. In our case, the patient had no active bleeding when she was referred; therefore, the exact source of hemoptysis could not be identified. Following a consultation with thoracic surgeons and interventional radiologists, it was decided to avoid any interventional therapy.

In conclusion, we have described several rare developmental anomalies in the same patient. Clinicians should be aware of undiagnosed cases of UAPA presenting with hemoptysis and the existence of IVC anomalies before interventional or surgical procedures, which sometimes are critically important to safely perform the procedure.

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****Supplementary video file associated with this article can be found in the online version of the journal.***

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Anahtar sözcükler: Konjenital anormallik/tanı; kardiyovasküler anormallik; hemoptizi.