

Recurrent pulmonary embolism in an asthmatic patient who had interrupted inferior vena cava with azygous continuation

Azigos ven ile devamlılığı olan kesintili vena kava inferiyora sahip astımlı hastada tekrarlayıcı pulmoner emboli

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Summary– A 45-year-old woman with a history of recurrent pulmonary embolism was admitted to the emergency clinic with dyspnea, wheezing and tachypnea. Partial deep vein thrombosis of the popliteal vein was seen on Doppler sonography. On the contrast-enhanced thorax computed tomography (CT) scan, a clot was detected in the right main pulmonary artery and its major descending branch. Moreover, the azygos vein was prominently dilated. Abdominal multi-slice computed tomography (MSCT) scan revealed absence of the hepatic segment of the inferior vena cava (IVC) with continuation of the IVC as a dilated right-sided azygos vein. The hepatic veins were draining directly into the right atrium. Thus, we discuss herein this rare anatomic variant presented with recurrent pulmonary embolism, together with the findings on MSCT.

Interruption of the inferior vena cava (IVC) is a congenital anomaly. The venous drainage of the lower extremities is achieved through a compensatory dilated vena azygos system. Computed tomography (CT) and magnetic resonance imaging (MRI) are non-invasive imaging techniques that can be used to detect these vascular anomalies. Awareness of the existence of IVC anomalies is important for preoperative planning. These anomalies are rarely associated with thrombosis of the iliac and femoral veins, especially in young

Abbreviations:

CT	Computed tomography
CTA	Computed tomography angiography
IVC	Inferior vena cava
MSCT	Multi-slice computed tomography
PTE	Pulmonary thromboembolism

Özet– Tekrarlayıcı pulmoner emboli hikayesi olan 45 yaşında kadın hasta dispne, hırıltılı solunum ve taşipne şikayetleri ile acil kliniğine başvurdu. Doppler ultrasonografide, popliteal vende kısmi trombüs saptandı. Kontrastlı toraks bilgisayarlı tomografisinde (BT), sağ ana pulmoner ven ve büyük dallarında trombüs mevcuttu. Ayrıca, azigos ven belirgin olarak genişlemişti. Abdominal çok kesitli bilgisayarlı tomografide (ÇKBT), vena kava inferiyor'un (VKİ) hepatic segmentinin olmadığı ve VKİ'nin genişlemiş azigos ven olarak devam ettiği saptandı. Hepatik venler, doğrudan sağ atriya boşalmaktaydı. Böylelikle, tekrarlayan pulmoner emboli ile ortaya çıkışı nadir olan bu anatomik varyasyonu, ÇKBT bulguları ile tartışmayı amaçladık.

patients.^[1] Here, we report a patient with interrupted IVC anomaly who presented with deep vein thrombosis of the femoral veins and recurrent pulmonary thromboembolism (PTE).

CASE REPORT

A 45-year-old woman who had been followed since 2009 presented to the emergency department with dyspnea, wheezing and tachypnea in January 2010. She was admitted to the hospital and received asthma medication but did not recover. Thus, she underwent thorax CT scan, which revealed a thrombus in the right pulmonary artery (Figure 1). Lower extrem-

Received: May 20, 2013 Accepted: October 02, 2013

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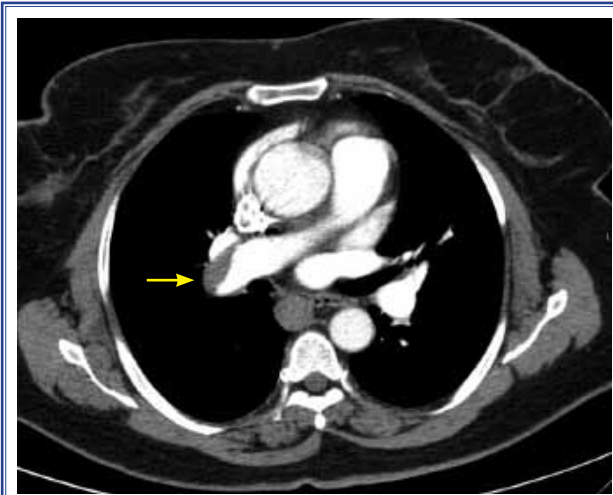


Figure 1. CT scan shows the thrombus in the right main pulmonary artery (arrow).

ity color Doppler sonography revealed deep venous thrombosis. She had no evident risk factors for thromboembolic disease, such as trauma, surgery, obesity, or history of a long journey (longer than 6 hours), or any other acquired risk factor. Standard therapy with heparin was given for six months without complication. The follow-up CT scan revealed resolution of the thrombus. One year later, the patient presented with symptoms of dyspnea, hemoptysis and swelling of the right lower extremity. Partial deep vein thrombosis of the lower extremity (popliteal vein) was seen on Dop-

pler sonography. Genetic tests were performed since she had a history of recurrent PTE. Tests for thrombophilia such as protein S, protein C, antithrombin III, homocysteine, and antiphospholipid antibody were found to be normal. Mediastinal widening was seen on the X-ray evaluation. On the contrast-enhanced thorax CT scan, a clot was detected in the right main pulmonary artery and its major descending branch. The CT scan revealed a prominently dilated azygos vein (Figure 2a, b). An abdominal CT scan was performed for the evaluation of the distal vascular structures. On the 64-slice multi-slice computed tomography (MSCT) and computed tomography angiography (CTA), absence of the hepatic segment of the IVC was noted. The IVC continued as a dilated, right-sided azygos vein. The hepatic veins were draining directly into the right atrium (Figure 3). The left renal vein was retro-aortic. No additional congenital anomalies, including heart disease or polysplenia, were seen. The patient was treated using unfractionated heparin and oral anticoagulants. A standard regimen was used including initial intravenous bolus of 80 IU/kg heparin followed by continuous infusion with 18 IU/kg/h. We measured activated partial thromboplastin time value every six hours to determine whether the therapeutic threshold was reached. Warfarin was added and maintained after 24 hours of heparin therapy. At the time of the first attack, warfarin therapy had been continued for nine months. After a second attack, a new therapeutic plan

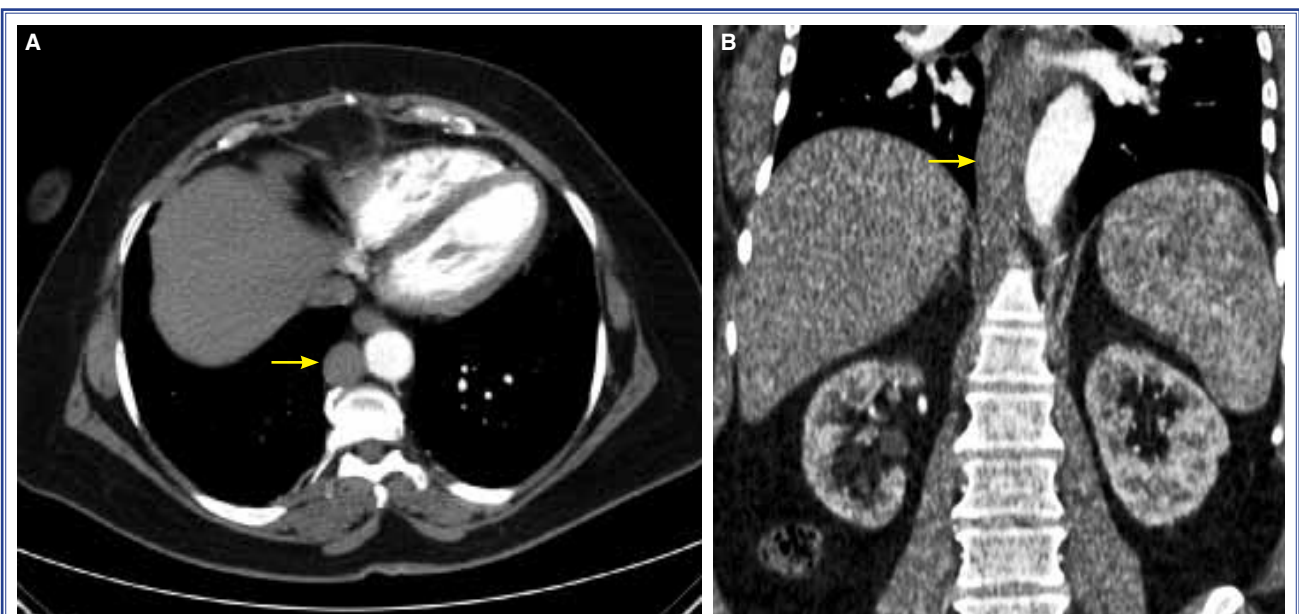


Figure 2. (A) Axial and (B) coronal CT scans demonstrate the enlarged azygos vein at the hepatic level (arrow).



Figure 3. Coronal CT scan shows the hepatic veins draining directly into the right atrium (arrow).

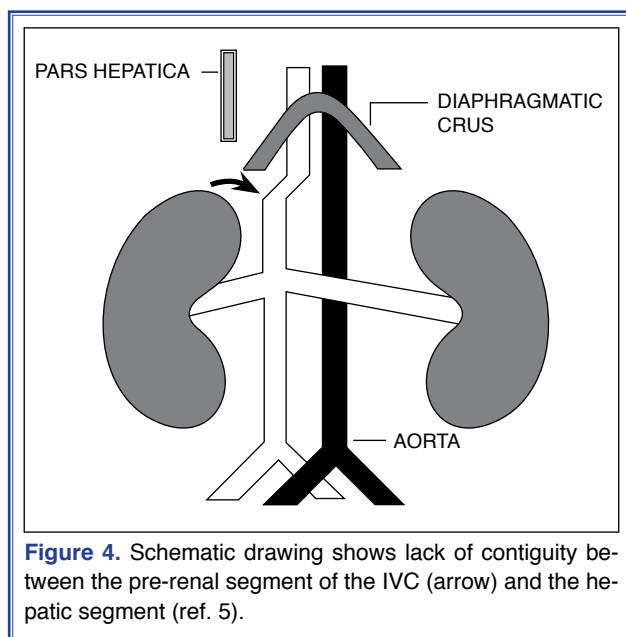


Figure 4. Schematic drawing shows lack of contiguity between the pre-renal segment of the IVC (arrow) and the hepatic segment (ref. 5).

was established. She has had no recurrent symptoms.

DISCUSSION

The embryology of the IVC is complex.^[2] The normal IVC consists of four main segments: hepatic, suprarenal, renal, and infrarenal.^[3] Hepatic segment anomalies occur when the right subcardinal vein fails to converge with hepatic sinusoids.^[3] Thus, blood is shunted from the suprasubcardinal anastomosis through the retrocaval azygos vein.^[4] Azygos continuation of the IVC is defined as absence of the hepatic segment of the IVC with azygos continuation (Figure 4).^[4] This anomaly is a rare clinical condition, with a prevalence

of 0.6%. It is often associated with congenital heart disease, polysplenia, and less commonly, asplenia.^[5] Isolated occurrence without heart disease may be encountered incidentally during radiological imaging studies or vascular interventions. Our patient had no detectable additional congenital anomalies.

The azygos vein generally originates below the diaphragm at the level of the first two lumbar vertebrae by confluence of the ascending lumbar vein and the right subcostal vein.^[6] A normal azygos vein is about 3-7 mm in width and cannot be seen on conventional radiography. When azygos continuation of the IVC is present, the visceral and lower extremity blood returns to the right atrium through the azygos vein. The enlarged azygos vein may manifest as mediastinal widening on chest radiography, and it can be mistakenly interpreted as right-sided paratracheal adenopathy and mediastinal mass.^[7] In the presented case, widening of the right mediastinum was present on chest radiography; however, the final diagnosis was established with CT.

Knowledge of the presence of an interrupted IVC may be important for patients undergoing right heart catheterization, electrophysiological studies, cardiopulmonary bypass surgery, femoral vein catheter insertion, IVC filter placement, and temporary pacing via the transfemoral route. If unidentified, it can lead to life-threatening complications during abdominal surgery.^[5]

Inferior vena cava (IVC) anomalies have been recognized as possible risk factors for deep vein thrombosis. Ruggeri et al.^[8] showed IVC anomalies in four of 75 patients with a first-attack deep vein thrombosis. In another study, 31 patients with iliofemoral deep vein thrombosis were prospectively evaluated by venography and MR angiography, and five anomalous IVCs were found in that study.^[2] An interrupted IVC may cause increased blood pressure in the lower extremity veins in the presence of insufficient collateral vessels. Eventually, venous stasis, which predisposes to deep vein thrombosis and indirectly to PTE, as in our case, may occur. PTE in patients with interrupted IVC with azygos continuation is reported rarely, and most of the publications are case reports.^[3]

Another important feature of our case is the presence of asthma, which is a chronic pulmonary disease characterized by reversible airflow obstruction. Dyspnea, wheezing, cough, and chest tightness are com-

mon symptoms. Pulmonary infections, congestive heart failure, bronchiectasis, upper airway obstructions, and other airway diseases such as chronic obstructive pulmonary disease (COPD), pulmonary embolism, and gastroesophageal reflux disease should be considered in the differential diagnosis. Bronchoconstriction, the main feature of asthma, can also be encountered in pulmonary embolism. Bronchospasm was seen after experimental thromboembolism in dog studies.^[9] In particular, wheezing and dyspnea could interfere with the diagnosis of pulmonary embolism in asthmatic patients. Recently, studies have shown that asthmatic patients have activated coagulation in the airway.^[10] Inflammation affects procoagulant and fibrinolytic activities, and there is endothelial dysfunction in the airway circulation in asthmatic patients, which is a primary factor in the pathogenesis of pulmonary embolism. Studies have shown that asthmatic patients have local vascular inflammation and increased risk for pulmonary embolism.^[10]

In conclusion, we recommend that patients with recurrent PTE or deep vein thrombosis should be evaluated for IVC anomalies. The presence of a dilated azygos vein may be an indicator of interrupted IVC; therefore, additional images should be obtained to identify associated vascular anomalies. Interrupted IVC or asthma can be a risk factor for recurrent PTE. MSCT and CTA imaging are the most helpful techniques in the evaluation of vascular abnormalities. When complex congenital vascular anatomy is suspected, CTA is currently an excellent choice as the first-line diagnostic imaging modality.

Conflict-of-interest issues regarding the authorship or article: None declared.

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Key words: Asthma; azygos vein/radiography; diagnosis, differential; pulmonary embolism; venae cavae abnormalities.

Anahtar sözcükler: Astım; azigos ven/radyografi; tanı, ayırıcı; pulmoner emboli; vena kava anormallığı.