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Case Report | Olgu Sunumu

POLYCYSTIC HORSESHOE KIDNEY WITH RUPTURED STANFORD TYPE B AORTIC DISSECTION

POLİKİSTİK ATNALI BÖBREK İLE BİRLİKTE GÖRÜLEN RÜPTÜRE STANFORD TİP B AORTİK DİSEKSİYON

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ABSTRACT

60-year-old male patient presented to the emergency clinic complaining of nausea, vomiting and abdominal pain with no remarkable medical history except hypertension and active smoking. The computed tomography (CT) revealed a Stanford B-type aortic dissection starting from the thoracic aorta to the iliac arteries and polycystic horseshoe kidneys with multiple cysts in the liver. Since polycystic horseshoe kidney and Stanford type B dissection of the thoracic aorta is rare, and no study showing coexistence in the literature was found. This is the first case report that reveals the association of these clinical findings and CT images in literature.

Keywords: Polycystic kidney, horseshoe, aortic dissection

ÖZ

At nalı böbrek genellikle asemptomatiktir; ancak enfeksiyon, üreter taşları ve malignite gibi nedenlerle semptomatik hale gelebilir. Literatürde, yetişkin polikistik böbrek hastalığı ile ilişkili eşzamanlı durumlar arasında karaciğerde yaygın kistik lezyonlar, kolon divertikülü, kapak kalp hastalığı ve serebral arter anevrizması yer alır. Hipertansiyon ve aktif sigara içimi dışında kayda değer bir tıbbi öyküsü olmayan, 60 yaşında erkek hasta, bulantı, kusma ve karın ağrısı şikayetleri ile acil polikliniğe başvurdu. Yapılan Bilgisayarlı tomografi (BT) incelemede; torasik aorttan iliak arterlere uzanım gösteren Standford tip B diseksiyon ve eşlik eden ve polikistik at nalı böbrek görüldü. Literatürde polikistik at nalı böbreği ve torasik aortun Stanford tip B diseksiyonunu bir arada var olduğunu gösteren herhangi bir çalışmaya rastlanmamıştır. Bu klinik bulgular eşliğinde BT görüntülerini ortaya koyan ilk olgu sunumudur.

Anahtar Kelimeler: Polikistik böbrek, atnalı, aortik diseksiyon

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Introduction

Polycystic horseshoe kidney (PHsK) is a combination of two distinct renal disorders, horseshoe kidney and autosomal-dominant polycystic kidney disease (ADPKD) that is a hereditary disorder due to mutations in the PKD1 gene.[1] Polycystic horseshoe kidney is very rare, with an incidence ranging from 1 in 134,000 to 1 in 8,000,000 cases.[2] Horseshoe kidney is usually asymptomatic. However, it may become symptomatic due to causes such as infection, ureteral stones, and malignancy.[3] In the literature, concurrent conditions associated with adult polycystic kidney disease include common cystic lesions in the liver, colonic diverticula, valvular heart disease and cerebral artery aneurysm.[4]

Case Report

60-year-old male patient presented to the emergency clinic complaining of nausea, vomiting, and abdominal pain radiating to the back. His medical history was unremarkable except for hypertension and active smoking. The patient was using Irbesartan as an anti-hypertensive therapy. There was no other medication. There were no unusual features in laboratory blood and, urine tests, and there were normal findings on electrocardiogram (ECG). CT revealed a dissection starting from the thoracic aorta to the iliac arteries. (Figure 1 and 2) There was a rupture that restricts itself in the isthmic region in the pattern to the descending aorta. (Figure 3 and 4) The imaging also showed polycystic horseshoe kidneys with multiple cysts in the liver. The right ureter contained urolithiasis of about 12 mm in diameter, but hydronephrosis had not developed.

CT the celiac truncus, superior mesenteric artery (SMA), and inferior mesenteric artery (IMA) were coming out of the true lumen. Thus, the patient had multiple renal arteries. One of these renal arteries originated from the native iliac artery true lumen, the other from the false lumen of the abdominal aorta, and the others from the true lumen of the abdominal aorta.

The final diagnosis was Stanford B-type aortic dissection with concurrent polycystic horseshoe kidney and polycystic liver disease.

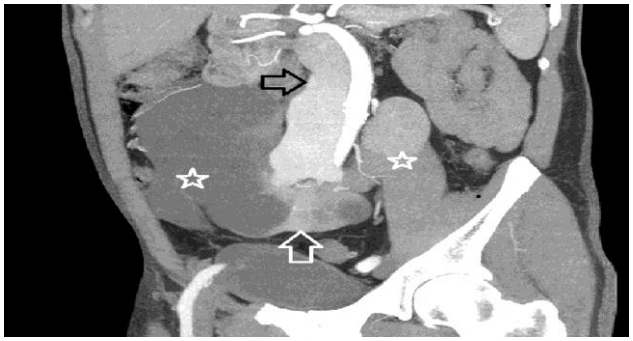

Figure 1. Axial contrast-enhanced CT images showing a horseshoe configuration of kidneys (white stars on right and left sides and white arrow of horseshoe kidney,) with ruptured aorta aneurysm (black arrow)

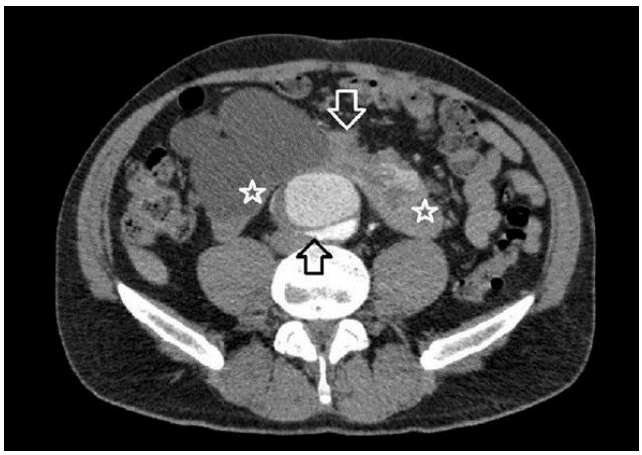

Figure 2. Axial contrast-enhanced CT images showing a horseshoe configuration of kidneys (white stars on right and left sides and white arrow of horseshoe kidney,) with ruptured aorta aneurysm (black arrow)

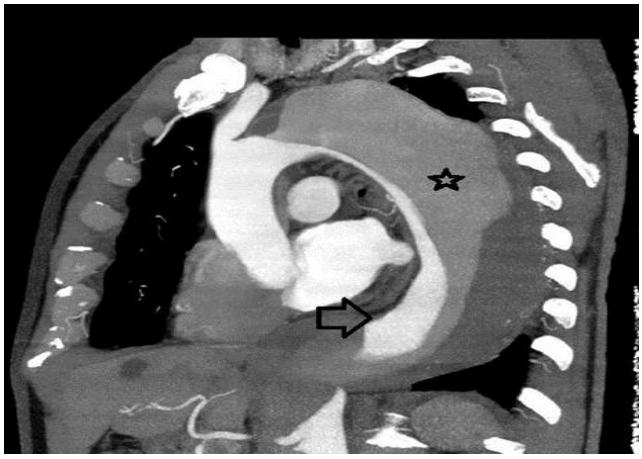

Figure 3. Sagital contrast-enhanced CT images showing Stanford B-type aortic dissection (black star) that restricts itself in the isthmic region in the pattern to the descending aorta (black arrow indicates the real lumen of the thoracic aorta) with parenchymal cyst in the liver

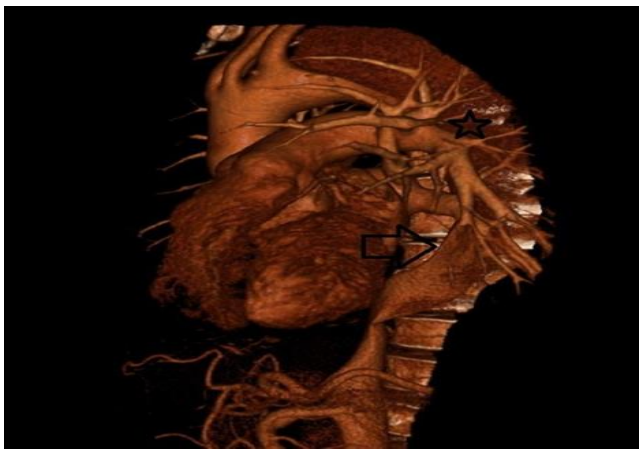

Figure 4. 3D-CT image showing Stanford B-type aortic dissection (black star) that restricts itself in the isthmic region in the pattern to the descending aorta (black arrow indicates the real lumen of the thoracic aorta)

Discussion

Polycystic horseshoe kidney is a combination of two distinct renal disorders. One of them is the horseshoe kidney which is a renal fusion anomaly during embryogenesis. The other one is an autosomal-dominant polycystic kidney disease (ADPKD) that is a hereditary disorder due to mutations in the PKD1 gene, localized on the short arm of chromosome 16 and PKD2 gene localized to chromosome 4.[5] Horseshoe kidney is the most common form of all renal fusion anomalies with an incidence of 1 in 400. Polycystic horseshoe kidney is an infrequent occurrence with incidence ranges of 1 in 134,000 to 1 in 8,000,000 cases, and only a few cases reported in the literature.[2]

The coexistence of polycystic renal disease and horseshoe kidney disease lowers the age of renal failure. Isolated ADPKD is the third most common cause of end-stage kidney disease.[1] In the literature, concurrent conditions associated with adult polycystic kidney disease include common cystic lesions in the liver, colonic diverticula, valvular heart disease, and cerebral artery aneurysm.[4] According to the study of Adhisivam et al., ventricular septal defect (VSD (60%) was the most common cardiovascular malformation followed by an atrial septal defect (ASD (25%,), and pulmonary stenosis PS (25%). Tetralogy of Fallot (TOF) was observed in (10% of) children who had urinary tract malformation. These authors suggested that this association was due to the common embryonic origin of both the urinary tract and heart, the mesoderm.[6]

Pardo et al.[7] highlighted an association of polycystic kidney disease with an increased incidence of cardiovascular abnormalities, in particular of mitral and tricuspid valve prolapse, and multivalvular incompetence. Pardo et al.[7] suggested that autosomal dominant polycystic kidney disease is a disorder of connective tissue. In our case, there was a Stanford Type-B dissection in the aorta with the polycystic horseshoe kidney. Thus, it is also our opinion that some connective tissue abnormality has occurred. However, the true incidence of concurrent aortoiliac occlusive disease remains unknown. Guvendi et al.[8] reported a horseshoe kidney with a concurrent aortoiliac occlusive disease using CT angiography

Since polycystic horseshoe kidney and Stanford type B dissection of the thoracal aorta is rare and no study showing coexistence in the literature was found. This is the first case report that reveals the association of these clinical findings and CT images in literature.

Compliance with Ethical Standards

Consent was obtained from the patient.

Conflict of Interest

The authors report no conflicts of interest.

Author Contribution

Authors contributed equally to this work.

Financial Disclosure

None

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