A Congenital Renal Arteriovenous Malformation Mimicking Renal Cell Carcinoma: Case Report

Renal Hücreli Karsinoma Benzeyen Konjenital Bir Renal Arteriyovenöz Malformasyon

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ABSTRACT Renal arteriovenous malformation is a rare condition. It is usually asymptomatic. When symptomatic, it presents with hematuria, flank pain, a mass on renal imaging and abnormal radiologic findings. It can mimic renal cell carcinoma. The diagnosis and treatment of renal arteriovenous malformations are different from those of renal cell carcinoma. We described a renal arteriovenous malformation mimicking renal cell carcinoma. A 35-year old man was admitted to the hospital with a history of right flank pain. Computerized tomography scan of the patient revealed a right renal mass, enhancing on contrast imaging. The patient underwent right simple nephrectomy. Pathologic examination of the nephrectomy specimen revealed a congenital cystic arteriovenous malformation. We suggest that it is essential to make a differential diagnosis of any lesion mimicking renal cell carcinoma.

Key Words: Carcinoma, renal cell; diagnosis, differential; vascular malformations

ÖZET Renal arteriyovenöz malformasyon nadir bir durumdur. Genelde asemptomatiktir. Semptomatik olduğunda hematüri, böğür ağrısı, renal görüntülemede bir kitle görüntüsü ve anormal radyolojik bulgular ile ortaya çıkabilir. Renal hücreli karsinomu taklit edebilmektedir. Renal arteriyovenöz malformasyonların tanı ve tedavisi renal hücreli karsinomdan farklılık göstermektedir. Bu makalede, renal hücreli karsinomu taklit eden bir renal arteriyovenöz malformasyon olgusu sunulmuştur. Otuz beş yaşında bir erkek hasta, hastaneye sağ böğür ağrısı öyküsü ile başvurdu. Bilgisayarlı tomografi incelemesinde kontrast tutan bir sağ renal kitle gözlendi. Hastaya sağ basit nefrektomi uygulandı. Nefrektomi örneğinin patolojik incelemesi konjenital kistik arteriyovenöz malformasyon olarak değerlendirildi. Bu makalede renal hücreli karsinomu taklit eden herhangi bir lezyon için ayırıcı tanının gerekli olduğunu göstermeye çalıştık.

Anahtar Kelimeler: Karsinom, böbrek hücreli; tanı, ayırıcı; vasküler malformasyon

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Renal arteriovenous malformations are rare lesions consisting of abnormal shunts between the arterial and venous vascular systems. They may be acquired or congenital. Acquired renal arteriovenous malformations represent most of arteriovenous malformations, which usually result from trauma or percutaneous renal procedures (e.g. renal biopsy). Congenital renal arteriovenous malformations, which are also called congenital arteriovenous fistulas, usually present with hematuria and flank pain. Very large congenital lesions may present with hemodynamic changes such as hypertension, cardiomegaly and congestive heart failure.[1] Renal ar-

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teriovenous malformations are similar to renal cell carcinoma in terms of radiological and clinical findings. Therefore, they may be misdiagnosed as renal tumors.

There are only a few case series in the literature describing the outcome of congenital arteriovenous malformations presenting with hematuria, a renal mass on imaging and an unexpected renal process such as spontaneous rupture. In this report, we described a case of congenital cystic renal arteriovenous malformation mimicking renal cell carcinoma.

CASE REPORT

A 35-year old man was admitted to our clinic with a three-month history of right flank pain and pollakiuria. The patient had no history of trauma. His blood pressure, blood count, serum levels of urea and creatinin, and coagulation parameters were all within normal range. Uroflowmetric parameters analyzed for low urine flow were normal. Ultrasonography of urinary system demonstrated a right renal mass of approximately 4x5 cm in the renal sinus. Intravenous pyelography (Figure 1) and computerized tomography scan revealed a right renal mass. After intravenous contrast material injection, the mass in the renal sinus appeared as a hyperdense lesion (Figure 2). Considering the likelihood of renal cell carcinoma on computerized tomography imaging, the patient underwent open surgery. Intra-operatively, the right kidney was abnormally enlarged with a pulsatile aneurysmal dilatation of the renal hilus. Right simple nephrec-

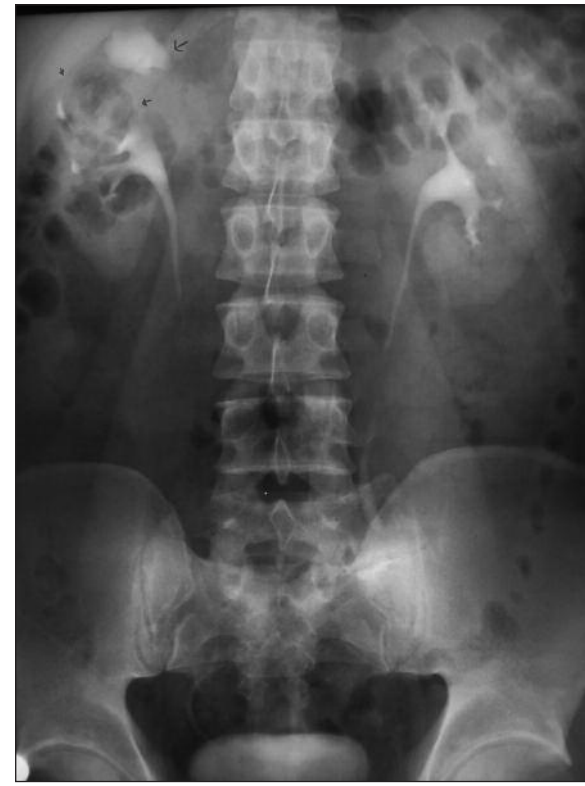

FIGURE 1: After intravenous contrast material injection, a pyelogram phase of 15 minutes demonstrates a filling defect within the right renal pelvis and upper pole caliceal deformation and a renal mass.

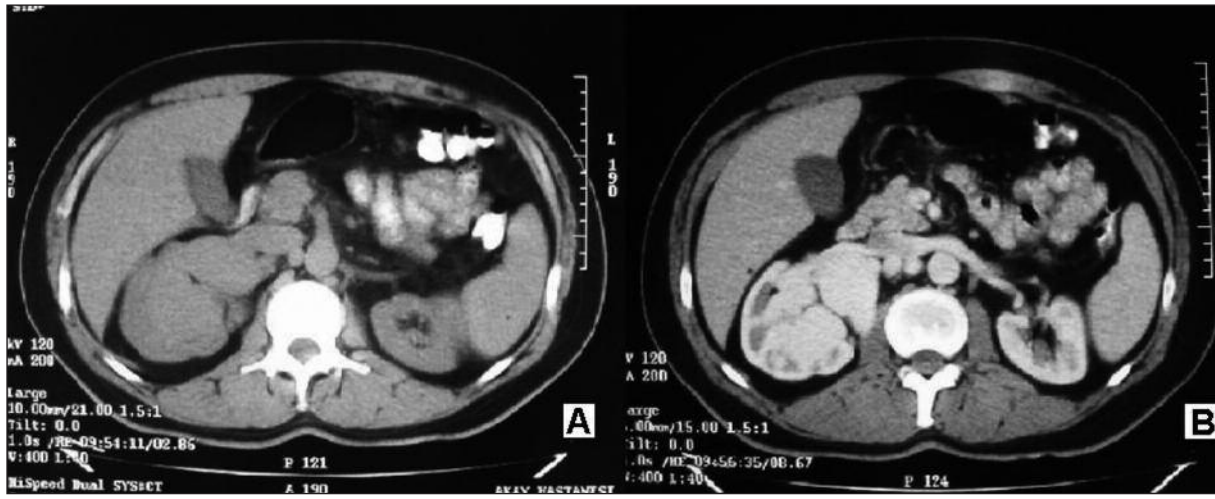

FIGURE 2: Axial computerized tomography scan. Before intravenous contrast material injection, the renal mass appears hypodense in the right renal hilus **(A).** After intravenous iodinated contrast material injection, the renal mass appears as a hyperdense, enhancing lesion with an indentation on the right renal vein **(B)**.

tomy was performed because of the indentation by the aneurysm to the right kidney and inferior vena cava, and the risk of rupture of the aneurysm. Pathologic examination of the nephrectomy specimen revealed a congenital cystic arteriovenous malformation (Figure 3). Six months after the operation, the patient had no problem.

DISCUSSION

The presenting symptoms of renal arteriovenous malformation that are characterized by multiple communications between the main or segmental renal arteries and veins are usually gross hematuria and hypertension. There are two classifications for arteriovenous malformations. The first classification is acquired or congenital and the second classification is acquired, congenital or idiopathic. Congenital renal arteriovenous malformations account for up to 27% of all cases, and are of two distinct types, classical and aneurysmal.[2] Classical congenital renal arteriovenous malformations are more common and appear as intrarenal malformations, presenting with gross hematuria in more than 70% of cases. Aneurysmal congenital renal arteriovenous malformations present more commonly with hypertension and high-output cardiac failure and can mimic renal pathologies such as parapelvic cyst and renal cell carcinoma.[2-4] However, acquired renal arteriovenous malformations account for up to 80% of cases and may develop following trauma, surgery, renal biopsy, malignancy, and inflammation. Our case presented to the hospital with flank pain and an unusual renal mass in radiological imaging and clinical examination. Our patient had an arteriovenous malformation, which caused neither hematuria nor hypertension and he had no history of trauma.

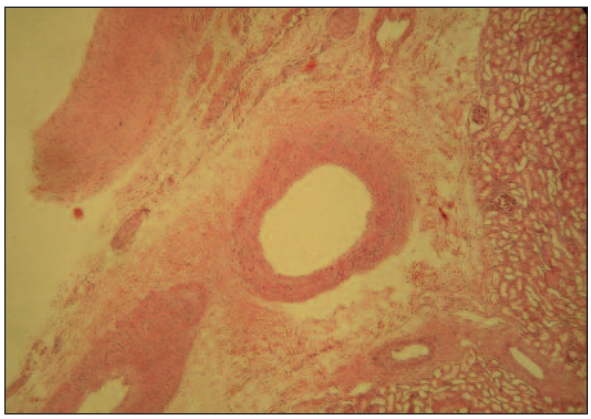

FIGURE 3: Pathologic examination of right simple nephrectomy specimen, demostrating intact renal parenchyma on the right. Proliferating, thick-walled, wide and dilated vessels appear adjacent to the parenchyma (Hematoxilen-Eosine, 50x).
(See for colored form http://tipbilimleri.turkiyeklinikleri.com/)

The radiologic appearance of the renal arteriovenous malformation, especially on renal angiographic examination, reveals several conditions such as an artery communicating directly with one or more veins and an aneurysmal dilatation of renal artery or vein. Although our patient did not undergo renal anjiography, it was considered congenital arteriovenous malformation dilated in an aneurysmal way. As it was previously described, renal arteriovenous malformations may mimick parapelvic cysts, hydronephrosis or benign and malignant renal masses. Radiological findings in our patient resembled renal cell carcinoma. Therefore, it was treated like a right renal mass.

Renal arteriovenous malformations mimick renal cell carcinomas in terms of radiologic findings and may clinically present as renal mass lesions. A filling defect and caliceal deformation may appear in intravenous pyelograpy. A hyperdense and contrast-enhancing renal mass may be determined in computerized tomography and magnetic resonance imaging.[5] Doppler ultrasonography, dynamic computerized tomography and magnetic resonance imaging are the most effective methods for the differential diagnosis especially in the perioperative period.[5] Computerized tomography and intravenous pyelography assisted the diagnosis of renal arteriovenous malformation mimicking renal cell carcinoma in our case. In cases with renal masses in computerized tomography scans or other radiologic methods, renal arteriovenous malformation should be considered in the differential diagnosis.

Treatment of renal arteriovenous malformations consists of several procedures such as open surgery, laparoscopic approach and catheter-directed embolizations.[6,7] The new treatment procedures for renal arteriovenous malformations include endoscopic methods and angioemboliza-

tion procedures. While open surgery methods are not commonly used, the current methods of choice are endovascular techniques and renal-preserving surgery.[8] In our patient, as the arteriovenous malformation could not be diagnosed in the preoperative period, we planned open surgery for renal cell carcinoma and intraoperatively, we decided to perform simple right nephrectomy. Actually, arteriovenous malformation was misdiagnosed in the presented case because colored Doppler ultrasonography was not used for the differential diagnosis of the renal mass. In fact, nephrectomy is not a standard choice for the treatment of renal arteriovenous malformation. However, we performed nephrectomy because the preoperative diagnosis was renal cell carcinoma. If renal Doppler ultrasonography had been used in the preoperative period, the patients would not have undergone open surgery and nephrectomy.

In conclusion, congenital arteriovenous malformations are rare, usually asymptomatic and benign lesions. The diagnosis of these lesions may be difficult because of their similarity to renal cell carcinoma and benign renal lesions such as hydronephrosis and parapelvic cyst. When there is a suspicion of renal arteriovenous malformation in the computerized tomography scan, renal color duplex ultrasonography and dynamic computerized tomography should be used for differential diagnosis. If the suspicion of renal cell carcinoma or renal mass persists, renal arterial angiography should not be avoided because the treatment of arteriovenous malformations does not require surgical procedures such as nephrectomy.

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