FUSED PELVIC ('CAKE') KIDNEY: AN INCIDENTAL FINDING IN A 77-YEAR-OLD PATIENT

PELVİK FÜZYONE ('KEK') BÖBREK: 77 YAŞINDAKİ BİR HASTADA RASTLANTISAL BİR BULGU

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ABSTRACT

Fused pelvic ('cake') kidney is a rare congenital anomaly of the genitourinary system. When a fused pelvic kidney is diagnosed, further evaluation should be performed to assess the status of the kidneys and exclude other associated congenital anomalies. The early diagnosis of potential complications that may accompany this anomaly are important to prevent permanent renal impairment. Multidetector computed tomographic urography enables a comprehensive evaluation of patients with fused pelvic kidney in a single examination. In this report, we present the multidetector computed tomographic angiography and urography findings of a 77-year-old patient with a fused pelvic kidney, diagnosed incidentally. We also review the present literature and discuss the embrology, anatomic features and clinical significance of this rare anomaly with an emphasis on its multidetector computed tomography findings.

Key words: Fused pelvic kidney, cake kidney, complete renal fusion, multidetector computed tomography, urography

ÖZET

Pelvik füzyone ('kek') böbrek genitoüriner sistemin nadir bir anomalisidir. Pelvik füzyone böbrek tanısı konulduğunda, böbreklerin durumunu değerlendirmek ve eşlik eden diğer konjenital anomalileri dışlamak için daha ileri bir değerlendirme gerçekleştirilmelidir. Bu anomaliye eşlik edebilecek potansiyel komplikasyonların erken tanısı kalıcı renal hasarın önlenmesi açısından önemlidir. Çok kesitli bilgisayarlı tomografik ürografi, pelvik füzyone böbrekli hastaların tek bir incelemede geniş kapsamlı olarak değerlendirilmesine olanak sağlar. Bu raporda, rastlantısal olarak tanı konulan, pelvik füzyone böbrekli 77 yaşındaki bir hastanın çok kesitli bilgisayarlı tomografik anjiyografi ve ürografi bulguları sunulmaktadır. Ayrıca mevcut literatürü gözden geçirip, çok kesitli bilgisayarlı tomografi bulgularını vurgulayarak bu nadir anomalinin embriyoloji, anatomik özellikleri ve klinik önemini tartışılmaktadır.

Anahtar kelimeler: Pelvik füzyone böbrek, kek böbrek, komplet renal füzyon, çok kesitli bilgisayarlı tomografi, ürografi

INTRODUCTION

A fused pelvic kidney, also known as a 'cake kidney' or 'lump kidney' is defined as an anomaly, in which 'the entire renal substance is fused into one mass lying in the pelvis, and giving rise to two separate and distinct ureters which enter the bladder in a normal relationship' (1,3). This is an uncommon congenital anomaly of the genitourinary system and rarely reported in literature (1). The early diagnosis and recognition of potential complications that may accompany this anomaly are important to prevent permanent renal impairment. Today multidetector computed tomographic (MDCT) urography has become the imaging modality of choice in the evaluation of congenital anomalies of the urinary tract due to its ability to depict the urinary tract anatomy, including both the renal parenchyma and collecting structures and ureters (6). In this report, we present the MDCT angiography and urography findings of a 77-year-old patient with a fused pelvic kidney diagnosed incidentally. We also review the present literature and discuss the embrology, anatomic features and clinical significance of this rare anomaly with an emphasis on its MDCT findings.

CASE

A 77-year-old woman with a history of chronic obstructive pulmonary disease and congestive heart failure with one year duration, presented with epigastric pain and nausea. Laboratory examinations were unremarkable. An endoscopy showed findings of an antral gastritis. An ultrasound of abdomen showed both kidneys were fused into one mass, ectopically located in the pelvis. Renal MDCT angiography followed by urography were performed with 16-row multislice computed tomography (Light-speed Ultra, GE Medical System, Wisc., USA). Initially, unenhanced images were obtained. Subsequently, 100 ml of nonionic iodinated contrast agent (Iodixanol, Visipaque 320 mgI/ml, GE Healthcare, Milwaukee, Wisc.; USA) was injected through an 18 gauge cannula positioned in an antecubital vein at a flow rate of 4 ml/sec, and the scans were obtained in three phases: arterial, nephrographic and pyelographic phases with acquisition delays of 22 seconds, 100 seconds and 5 minutes, respectively. For three-dimensional image reconstruction, the raw MDCT data were processed on a separate workstation (Advanced Workstation 4.2, GE Medical System, Wisc., USA) with volume rendering, multiplanar reformatting and

Date received/Dergiye geldiği tarih:10.02.2009 - Dergiye kabul edildiği tarih: 30.09.2009

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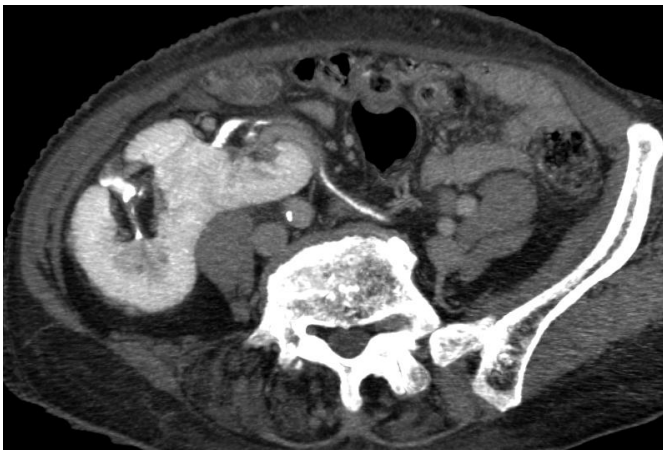
maximum intensity projection. MDCT revealed a fused pelvic kidney located in the right of the pelvis (Figure 1). It was drained by two separate ureters which enter the bladder in a normal relationship. The left ureter crossed the midline to enter the left side of the bladder (Figure 2). There were no stone and dilatation in the pelvicalyceal systems and both ureters. The fused pelvic kidney had two arteries, originating from common iliac arteries (Figure 3). One of them originated from the right common iliac artery and directly entered the hilum supplying the right portion of the fused kidney. The other originated from the left common iliac artery and divided into two branches before entering the hilum and supplying the majority of the fused renal mass. There was a calcified plaque at the origin of the left renal artery. A renal vein arose at each hilum and united to form a single trunk which opened into right common iliac vein. The patient was investigated in order to exclude concomitant anomalies. No additional anomalies were detected. In view of the normal renal function and the asymptomatic clinical course, no further intervention was considered necessary. The patient is currently being followed-up.



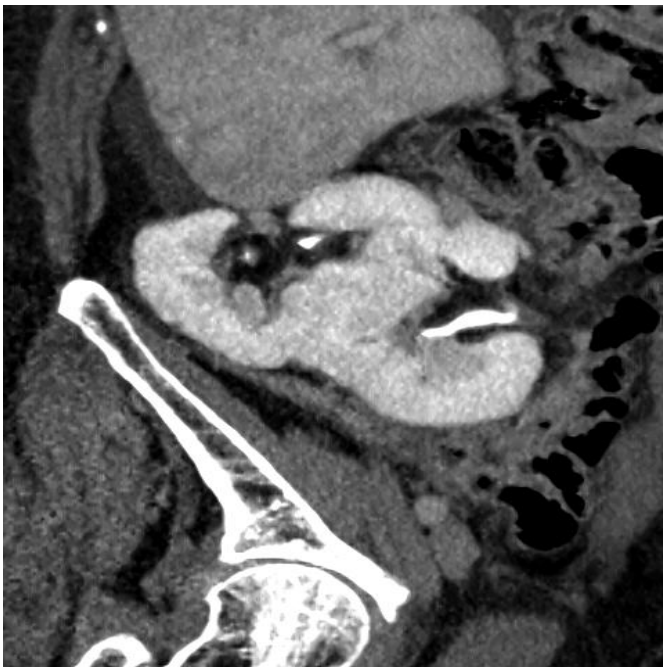


Figure 1. Oblique axial (A) and coronal (B) maximum intensity projection multidetector computed tomography images show fused pelvic kidney located in the right pelvis

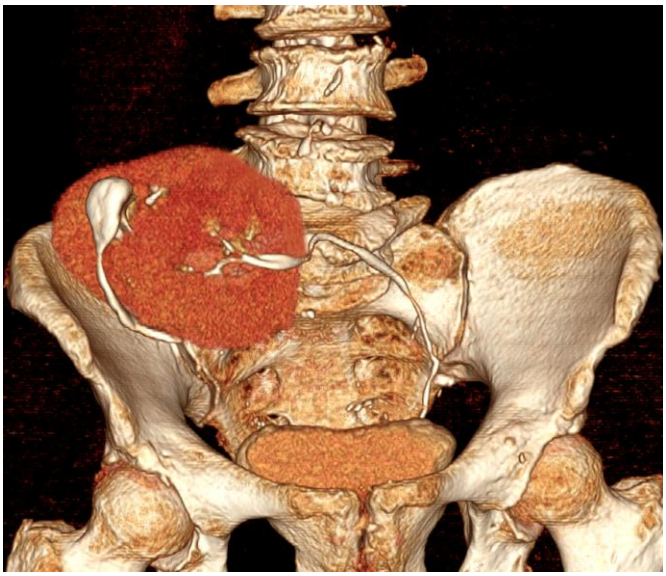


Figure 2. Coronal volume rendering multidetector computed tomography image in the pyelographic phase shows fused pelvic kidney that is located in the right pelvis and drained by two separate ureters. The left ureter crosses the midline to enter the left side of the bladder

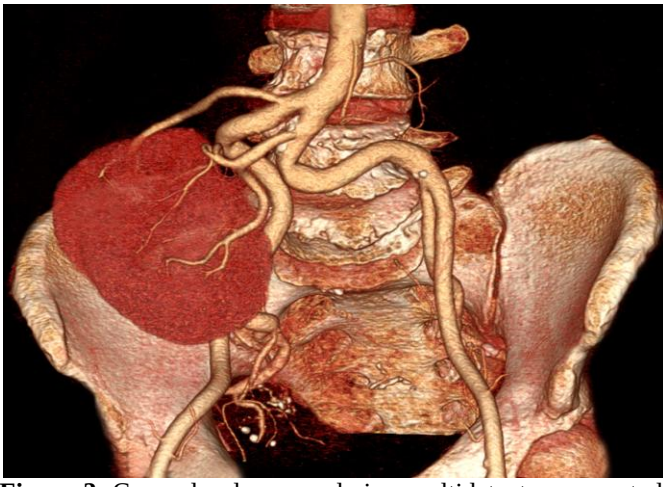


Figure 3. Coronal volume rendering multidetector computed tomography image in the arterial phase shows the fused pelvic kidney supplied by two arteries. One arises from right common iliac artery and another from left common iliac artery. There is a calcified plaque in the origin of the latter

DISCUSSION

The fused pelvic kidney occurs at an early stage in the embryological development. In the embryo, the two masses of metanephrogenic tissue lie within the pelvis. Each developing kidney reaches its normal position in the lumbar region following complicated movements involving ascent, lateral migration, axial deflection and internal rotation. The nephrogenic blastomas are squeezed together by the umbilical arteries at the beginning of the cranial migration of the ureteral buds which may cause their fusion (7). According to an alternative explanation, this anomaly could result from the growth of the ureteric buds into a common metanephric blastema (2). Completely fused kidneys such as the cake kidney are usually prevented from ascending to their normal position and remain in an ectopic pelvic position. The fused pelvic kidney may be located to the right or midline in the

presacral area. The fusion of the renal parenchyma prevents normal medial rotation to a varying degree, and therefore malrotation is a usual accompaniment, with the renal pelvis typically directed anteriorly. With rare exceptions, such kidneys possess two ureters, both of which enter the bladder in the normal regions of the trigone and characteristically do not cross (4).

The vascular supply of the fused pelvic kidney is consistent with its arrested migration. In the developing kidney the vascular supply is re-established progressively as it migrates cranially to reach its definitive position. During ascent, the normal kidney receives its blood supply first from the middle sacral artery, then the iliac arteries, and finally from the aorta. If the migration is arrested, the temporary blood supply will become permanent. Thereby, fused pelvic kidney derives its blood supply from the aorta near the bifurcation or from the common iliac arteries. Venous drainage is usually into the distal inferior vena cava or the common iliac veins. This anomalous blood supply is at increased risk for vascular compromise due to pelvic trauma, vascular disease, pregnancy or other space-occupying lesions (3).

The fused pelvic kidney may remain entirely asymptomatic and is diagnosed as an incidental finding during radiological examinations. Rarely, it may cause local pain by dragging on the renal vessels due to the weight of the organ (3). The abnormal position of the kidneys, pelvis and ureters can lead to poor outflow, urine stasis and a predisposition to hydronephrosis, stone formation, and infection, just as in other forms of a fused kidney. The ureters are also short and have a tangential course, which increases the risk of stone formation and ureteric obstruction (5). In addition, it is pointed out that the maldevelopment of collecting tubules may be the factor promoting cyst formation and that the enlargement of renal cysts may ultimately bring about renal failure (3). Because the fused pelvic kidneys have an aberrant blood supply from the distal aorta or common iliac arteries, where atherosclerosis is common, renal artery stenosis and hypertension may develop. The majority of reported cases with fused pelvic kidney had concominant anomalies in other organs including Fallot tetralogy, aortic coarctation, ventricular septal defects, abnormal testicular descent, vaginal absence, sacral agenesis, caudal regression syndrome, spina bifida or anal abnormalities (7).

To date, the diagnosis of fused pelvic kidney usually has been made by ultrasonography and excretory urography. Ultrasound is often the initial procedure performed in the work-up of the patients. Recently, MDCT urography has gradually evolved as an accepted method for comprehensive evaluation of the urinary tract. MDCT scanners offer shorter image acquisition time, narrower collimation, improved temporal and spatial resolutions and near isotropic data acquisition, which is advantageous for high resolution two or three-dimensional reformatted images compared with spiral computed tomography. Congenital anomalies of the urinary tract can be visualized better with MDCT urography rather than with excretory urography, because MDCT enables to depict the urinary tract anatomy, including both the renal parenchyma and collecting structures and ureters (8). MDCT urography is helpful to screen for the presence of stones, hydronephrosis or masses. It also provides additional information about the vascular supply of the fused kidneys (6). Thereby, MDCT urography has the potential to provide a comprehensive evaluation of patients with renal fusion anomalies in a single examination. The disadvantages of this method include potential for allergic reactions to iodinated contrast material and exposure to ionizing radiation.

In conclusion, when a fused pelvic kidney is diagnosed, further laboratory and imaging evaluation should be performed to evaluate the status of the kidneys and exclude other associated congenital anomalies. The diagnosis of fused pelvic kidney is not necessarily associated with a poor diagnosis. It requires long-term follow-up of renal function and early detection of possible future complications. MDCT enables a comprehensive evaluation of patients with fused pelvic kidney in a single examination.

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