

Ektopik böbrekler ve damarlarının konjenital varyasyonları

Ectopic kidneys and congenital variations of their vasculatures

Hanefi Bayaroğulları¹, Mehmet Murat Rıfaoğlu², Seckin Akküçük³, Ramazan Davran¹, Mürsel Davarcı², Onur Demirbaş²

¹ Mustafa Kemal University, Faculty of Medicine, Department of Radiology, Serinyol, Hatay, Turkey

² Mustafa Kemal University, Faculty of Medicine, Department of Urology, Serinyol, Hatay, Turkey

³ Mustafa Kemal University, Faculty of Medicine, Department of General Surgery, Serinyol, Hatay, Turkey

Özet

Amaç: Böbrek ektopilerinde damar anomalilerine sıklıkla rastlanılır. Multiple detector-row computerized tomography angiography (MDCTA), üriner sistem ve böbrek damarlarının değerlendirilmesinde günümüzde artan bir yere sahiptir. Bu çalışmamızda amacımız ektopik böbrekteki damar varyasyonlarını MDCTA kullanarak ortaya koymaktır.

Gereç ve Yöntem: Hastanemize 2009 ila 2011 arasında altı ektopik böbrek hastası araştırıldı. MDCTA, dört erişkin hastaya hipertansiyon nedeniyle, bir böbrek taşı olan erişkin hastaya nefrektomi öncesi, bir çocuk hastaya ise diafram hernisi operasyon öncesi yapıldı.

Bulgular: Altı hastanın 12 böbreğinin 20 renal arteri araştırıldı. İki vakanın birinin sol böbreğinde dört, diğerinde üç renal arter saptandı. Üçü ektopik biri normal olan dört böbrekte, bir ana bir de aksesuar arter saptandı. 17 ven inferior vena cava'ya, sol böbrekten çıkan bir ven sol common iliac vene, 2 sağ böbrek veni sol renal vene dökülüyorlardı.

Sonuç: Günümüzde MDCTA ektopik böbreğin damarlarını değerlendirilmesinde önem kazanmıştır.

Anahtar Kelimeler: Ektopik böbrek, kanlanma, MDCT, MDCTA

Abstract

Objectives: Renal vessel anomalies are more common in renal ectopia. Multiple detector-row computerized tomography angiography (MDCTA) has become increasingly important in the evaluation of the urinary system and the renal vasculature. The aim of this study is to demonstrate the variations of vascular supply of the ectopic kidneys by using MDCTA.

Material and methods: Six renal ectopic kidney cases were identified among the patients admitted to our hospital between 2009 and 2011. MDCTA was planned for four adult patients due to hypertension, for one adult patient who would undergo nephrectomy due to nephrolithiasis, and for one neonatal patient who would be operated on due to diaphragmatic hernia.

Results: 20 renal arteries were identified in 12 kidneys of six patients. There were four renal arteries in a left kidney and three renal arteries in another left kidneys of two cases. There were one main and one accessory arteries in four kidneys of which three were ectopic and one was normal. There were 17 veins connected to inferior vena cava, one vein of the left kidney was connected to left common iliac vein, and two veins of the right renal vein were connected to left renal vein.

Conclusion: Nowadays, MDCTA has gained significance in the evaluation of ectopic kidneys' vasculature.

Key Words: Ectopic kidney, vascular supply, MDCT, MDCTA

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Yazışma / Correspondence

Murat M. Rıfaoğlu, MD,
Mustafa Kemal University,
Faculty of Medicine,
Department of Urology,
31005, Hatay, Turkey
Tel: +90 326 229 10 00-3317
E-mail: muratrfai@yahoo.com

Introduction

An ectopic kidney is a congenital anomaly in which the kidney is located in an abnormal position within the body. Kidneys are susceptible to morphological variance, position, shape, size, rotation and especially vascularization. Kidneys are retroperitoneal, bean-shaped organs located in the lumbar diaphragmatic fossae. The urinary tract system anomalies account for %3 of all congenital system anomalies(1-3). The incidence of ectopic kidney is 1 in every 1.000 births. Multiple detector-row computerized tomography angiography (MDCTA) plays an increasingly important role in the evaluation of urinary system and renal vasculature. It is less invasive, easily available and applicable. MDCTA provides information about the morphological variety, position, shape, size, rotation of the kidney, and also about the vessel lumen, the vessel wall and the surrounding structures(4, 5).

The aim of this study is to demonstrate the variations of vascular supply of the ectopic kidneys by using MDCTA.

Patients and Methods

Six renal ectopic kidney cases were identified among the patients admitted to our hospital between 2009 and 2011. The mean age of the patients was 33.83 (1-67 years). Of the patients three were male and three were female. Patient profile is summarized in Table 1. The symptoms in three patients were abdominal pain, hypertension and non-specific findings. The ectopic kidneys were diagnosed in these three patients with routine examination in ultrasonography (USG) and computerized tomography. The pelvic ectopic kidneys in two patients were diagnosed with lumbar magnetic resonance imaging (MRI) performed due to back pain complaints. One neonatal patient with respiratory distress underwent chest radiography. Congenital diaphragmatic hernia was diagnosed secondary to intestinal gas shadows in thorax.

MDCTA was planned for four adult patients due to hypertension, for one adult patient who would undergo nephrectomy due to nephrolithiasis, and for one neonatal patient who would be operated on due to diaphragmatic hernia.

For five patients, MDCTA was conducted with

Toshiba Aquilon 64 MDCT. The protocol for imaging processes was as follows; 100 mL of iodinated contrast was injected at 5 mL/sec, the scan was performed in a single breath hold for the area between the diaphragm and the pubic symphysis. The scanning protocol for abdominal angiography; a collimation of 64x0.3mm for each rotation; the primary data processing was achieved through 0.5 mm reconstructions, with an algorithm dedicated to angiographic studies. After completing the primary reconstruction, the images were transferred to the post-processing station for three-dimensional reconstructions. For this process, there are three methods. Most commonly used post-processing techniques are multi-planar (MPR) or curved planar reformation (CPR); the other two methods are maximum intensity projection (MIP) and volume rendering (VR) methods.

The scanning protocol for abdominal MDCT was applied for the newborn patient for the investigation of diaphragmatic hernia and is sufficient for evaluation of renal vessels. The scanning protocol for abdominal angiography wasn't applied to avoid the harmful effects of radiation and nephrotoxicity of contrast material.

Results

12 kidneys in six patients were evaluated in the study. In three patients, bilateral ectopic kidneys (Fig. 1a-d) and in three patients, unilateral ectopic kidneys were observed (Fig. 2a-e). Morphological features and vasculatures of nine ectopic and three normal kidneys were evaluated with MDCT. Five kidneys were located in the pelvic region, three kidneys were between normal localization and pelvis, and one kidney was located in the right thorax (Fig. 3a-f). Five of the ectopic kidneys were in the right and four were in the left side. Morphologically, hypoplastic changes in two ectopic kidneys and atrophic changes secondary to nephrolithiasis in one ectopic kidney were observed (Fig. 4a-d). Patient profiles and ectopic kidneys are summarized in Table 1.

The length, diameter, origin and level of the renal arteries and the level of the renal vein connected to inferior vena cava (IVC), and other venous system vascular structures are detailed in Table 2.

Discussion

Kidneys locate retroperitoneal space on both sides of vertebral column and lie against the psoas muscles,

Table 1: Properties of patients, ectopic kidneys and vascular structures.

Case No	Age	Gender	Ectopic kidney			Localization of kidneys and sizes		The number of renal artery		The number of renal vein	
			Right	Left	Fus	Right	Left	Right	Left	Right	Left
1	30	Female	-	+	-	T12-L3 108x59x45	L4-S1* 87x56x54	1	1	1	3
2	38	Male	+	+	-	L4-S1* 83x54x36	L2-S1 158x78x53	1	4	2	3
3	1	Female	+	-	-	T5-10 54x30x28	T12-L4 65x30x24	1	3	1	1
4	67	Female	+	-	-	L4-S1* 74x52x36	T12-L3 92x51x46	1	1	1	2
5	41	Male	+	+	-	L3-L5 110x57x38	L4-S2* 118x71x42	2	2	1	3
6	26	Male	+	+	-	L4-S1* 83x70x44	L1-L4 95x62x49	2	1	1	1
		M	F								
Total	6	33.8	3	3	5	4	-	8	12	7	13

Abbreviations and signs: fus; fused, T; thoracic, L; lumbar, *; pelvic ectopic kidney, M; male, F; female.

but when a kidney fails to ascend in normal position, it is called renal ectopia (6-8). The exact mechanism is unknown. Teratogenic agents, genetic factors, chromosomal anomalies, medicines ingested during pregnancy, disorders in the fusion mechanism of the ureteric bud and the metanephrogenic blastema may be among the reasons, but many idiopathic cases have been reported (9). Normally, the upper pole of the right kidney extends to T11-12 intercostal spaces, while the upper pole of the left kidney extends to T11 vertebra. Lower pole of the right kidney is 2.5-3 cm above the iliac crest, but the lower pole of left kidney is 4-5 cm above the iliac crest, at the level of vertebrae L2-3. Kidneys are susceptible to morphological variance, position, shape, size, rotation and especially vascularization (10, 11). In our study, hypoplastic changes were observed in two ectopic kidneys and atrophic changes in one ectopic kidney. Abnormalities of shape and axis were observed in all ectopic kidneys.

Ectopic kidneys can be observed in the pelvis, abdomen and rarely in thorax. Ectopia can be unilateral or bilateral, with or without fusion. Simple renal ectopia refers to a kidney that is located on the usual side but in an abnormal position. Crossed renal ectopia refers to a kidney that has crossed from left to right or vice-versa. The crossed kidney is mostly fused with the other kidney,

which is called crossed fused ectopia (12). Thoracic kidney is a rare developmental abnormality. It can be either due to delayed closure of the diaphragm (pleura-peritoneal membrane defect) or accelerated ascent of kidneys before the normal diaphragm leaflets closure. The kidney usually lies in the posterior-lateral aspect of the diaphragm, in the foramen of Bochdalek (13, 14). In our study, five kidneys were in the pelvic region, three kidneys were between the normal localization and pelvis, and one kidney was in the right thorax.

The incidence is approximately 1:1000 for solitary pelvic kidney, 1:1300-1:7500 for crossed fused renal ectopia and 1:20000 for ectopic thoracic kidney. The incidence of thoracic kidney is <5% in all ectopic kidneys (2, 3). Generally, an ectopic kidney is smaller, irregular in shape, has variable rotations and is vascularized by multiple arteries with various levels of origin (15).

The urinary tract system anomalies comprises %3 of all congenital system anomalies. In the literature, ectopic kidneys are more frequent in males and on the left side (2, 3). However, in present study, the ratio of men and women was equal, and ectopic kidneys were mostly on the right side (%55.5).

An ectopic kidney is usually asymptomatic and detected incidentally during the physical examination of the patient for an unrelated complaint. The functional

Table 2: Properties of vascular structures in ectopic and normal kidneys

Case No	Renal Vessel	Length (Mm)	Diameter (Mm)	The origin and level of renal artery	The level of the renal vein communicated with VCI and other venous system
1	RRA1	48.2	5.4	L1	
	LRA1	10.4	3.2	L4	
	LRA2	82	3	L4 (from left CIA)	
	RRV1	31.7	6.2		T12
	LRV1	50	8		L4 (The posterior of the left CIA)
	LRV2	39	6.6		L4 (The anterior of the right CIA)
	LRV3	62	7.85		L4 (The posterior of the right CIA)
2	RRA1	77.6	3.5	L4 (AB)	
	LRA1	42	5.3	L2	
	LRA2	70.5	4.7	L3	
	LRA3	34	4.2	L3	
	LRA4	80.8	3.6	S1 (from left CIA)	
	RRV1	67.17	6.25		L3-4 (opens to LRV2)
	RRV2	104	4.94		L4 (opens to LRV2)
	LRV1	118.3	9.41		L2
	LRV2	91.28	11.73		L3
LRV3				S1 (opens to CIV)	
3	RRA1	59	3	L1	
	LRA1	12	1.5	L1	
	LRA2	10	1.5	L1	
	LRA3	17	1.6	L2-3	
	RRV	34	8.2		T12
	LRV	43	11.73		L2 (Retroaortic)
4	RRA1	29	3.6	L3-4 (AB)	
	LRA1	50.4	4.2	L1	
	RRV1	54	4.4		L3
	LRV1	73.9	8		L1
	LRV2	106.4	6.2		L3 (Retroaortic)
5	RRA1	46.6	4.8	L3	
	RRA2	70.6	4.8	L4 (AB)	
	LRA1	40	6.2	L3	
	LRA2	60	2.9	L5 (Left CIA)	
	RRV1	67.8	10.5		L2-3
	LRV1	140.10	9.89		L2
	LRV2	84.60	5.24		L3 (Retroaortic)
	LRV3	90.60	5.2		S1 (Left CIV)
6	RRA1	35	4.6	L3-4	
	RRA2	31	3.5	L5 (AB)	
	LRA1	53.55	5.4	L2	
	RRV1	75.1	12		L3
	LRV1	79.6	9.68		L2

Abbreviations: RRA; right renal artery, RRV; right renal vein, LRA; left renal artery, LRV; left renal vein, CIA; common iliac artery, CIV; common iliac vein, AB; aortic bifurcation.

Figure Legends:

Figure 1: A 41 years old male patient with MDCT-A evaluation, (a). Coronal volume rendering imaging, (b). Maximum Intensity Projection (MIP) imaging shows bilateral ectopic kidneys and two renal arteries of right ectopic kidneys (short red arrows), two renal arteries of left ectopic kidneys (tall red arrows), (c, d). Oblique Multiplanar Reformatting imaging, the vein of the right kidneys (short blue arrow), the vein of left kidney (tall blue arrow).

Figure 2: A 46 years old female patient with MDCT-A evaluation, (a, b). Coronal and sagittal volume rendering imaging show the right ectopic kidney, the artery of right ectopic kidney (short red arrow) and the artery of the left normal kidney (tall red arrow), (c). Sagittal Multiplanar Reformatting imaging shows the vein of right ectopic kidney (short blue arrow) and the retroaortic accessory vein of the left normal kidney (tall blue arrow), (d). Coronal Multiplanar Reformatting imaging shows the artery and vein of the right kidney (short red and blue arrows), (e).

Axial Multiplanar Reformatting imaging shows the main vein of the left kidney (*tall blue arrow*)

Figure 3: The MDCT examination of the 1-month-old girl child, (a). Axial MDCT imaging shows right ectopic thoracic kidney secondary to congenital diaphragmatic hernia and bowel ans in right thorax (Bochdalec hernia), (b, c). Coronal and sagittal Multiplanar Reformatting imaging show the artery of the ectopic right thoracic kidney (*thick red arrow*), the vein (*thick blue arrow*) and inferior vena cava (*white arrow*), (d). Axial imaging show the retroaortic vein of the left normal kidney (*thin blue arrow*), (e). Coronal Multiplanar Reformatting imaging shows three arteries of the left kidney (*thin red arrows*), the retroaortic renal vein of the left kidney (*thin blue arrow*)

ectopic kidney may go undetected all life long. In some cases of ectopic kidney, individuals can have such urinary problems as urine blockage, urinary tract infections or urinary stones (8).

During ascension, each kidney is vascularized by neighboring vessels, initially from internal and external iliac vessels and directly from aorta after the 8th gestational week (12). There is a strong correlation between renal ascension and vascularization. As the kidneys ascend up, they derive their blood supply from the segmental mesonephric arteries. The mesonephric arteries may fail to regress and persist as supernumerary vessels. There is a good correlation between the kidney ascension and the level of the origin of the renal arteries. Any anomaly in the renal artery development can stop the ascension and cause ectopia or rotational anomalies. Anomalies in renal vessels are more common in the renal ectopia, and the presence of multiple renal arteries is the most frequent variant(16).

A wide variety of diagnostic modalities such as ultrasonography, intravenous urography, duplex and color Doppler USG, MR angiography, intravenous digital subtraction angiography (DSA) and MDCTA are used in the evaluation of ectopic kidney and its vasculature.

For the evaluation of renal vasculatures, conventional angiography or DSA used in the past. However, it has an invasive nature associated with complications. DSA is not suitable for screening. Nowadays the method of choice is MDCTA. MDCTA has become increasingly important in the evaluation of the urinary system and the renal vasculature. It is less invasive, easily available and applicable. However, exposure to ionizing radiation and the use of iodinated contrast material, which has

and the artery of the ectopic right thoracic kidney (*thick red arrow*), (f). Coronal MDCT lung window image shows better the herniated intestinal segment

Figure 4: A 38 years old male patient with MDCT-A evaluation, (a). Coronal volume rendering imaging shows the bilateral ectopic, right atrophic kidney (secondary to nephrolithiasis) and renal stones in the right renal pelvis (*white arrow*), the artery of right ectopic kidney (*short red arrow*) and the arteries of the left normal kidney (*tall red arrows*), (b). Coronal MIP, Coronal Multiplanar Reformatting and Coronal MIP images shows the veins of right ectopic kidney (*short blue arrow*) and the veins of the left kidney (*tall blue arrows*), two renal veins of right kidney open accessory left renal vein (c, d).

nephrotoxic potential, are the main drawbacks. As well as the information about angiography features, MDCT provides information about the morphological variety, position, shape, size and rotation of the kidney. Furthermore, it offers information not only about the vessel lumen and the vessel wall but also the surrounding structures (4, 5, 17-19).

Renovascular hypertension, renal transplant recipient and donor evaluation, direct renal trauma, arteriovenous communications, renal artery aneurysm, renal parenchymal or vascular calcifications and renal manifestations of a systemic disease constitute the main clinical applications of MDCT and MDCTA. Diagnostic accuracy of renal MDCT and angiography depends on the quality of initial raw data obtained during the study. The raw data were collected and sent to work station for post-processing. Most commonly used post-processing techniques are MPR or CPR, MIP and VR. MIP images can produce angiography-like images and excellent overview of the vascular anatomy and the vessel lumen, wall and calcification. MPR and CPR images are useful in the evaluation of the arterial luminal diameter and the arterial stenosis (20-22).

In most individuals, each kidney is supplied by one renal artery arising from abdominal aorta, but in approximately 30% of individuals more than one artery can be present. Renal arteries are usually 4-5cm in length and 5-6mm in diameter and they typically arise from the aorta at the level of superior margin of L2 vertebral body, slightly inferior the origin of the superior mesenteric artery. The right renal artery orifice is located on the anterolateral wall of the aorta and the left renal artery orifice is located in more lateral location. The right

renal artery demonstrates a long downward course and the left renal artery a short horizontal course according to the anatomical conditions of the kidneys. Anatomic variations of the renal artery are common in general populations.

The renal lobar veins converse to form the main renal vein at the renal hilum and it lies anterior to the renal artery. The left renal vein is three times longer than right renal vein and its length is 6-10cm. It normally courses between the superior mesenteric artery and aorta before draining into the medial aspect of IVC. The left renal vein may demonstrate anatomic variations like as retro-aortic, circum-aortic and partially duplicated vein. It receives several tributaries (The left adrenal vein, the left gonadal vein and lumbar vein) before draining into the IVC. The right renal vein averages 2-4cm in length and joins the lateral aspect of the IVC. Each vein may include multiple veins (23-25).

In our study, 20 renal arteries were identified in 12 kidneys in six patients. The highest number of arteries was observed in the left kidney of case 2. The artery arising from the highest point of the aorta was at the level of L1. The artery arising from the lowest point was from the left common iliac artery at the level of S1.

The right renal vein is shorter than the left. It is usually 2.5 cm long and does not receive any tributary. The left renal vein is three times longer (7.5 cm) than the right. It crosses anterior to the aorta to open into the lateral aspect of IVC. It may receive 2 tributaries; left suprarenal and left gonadal veins (26). The venous variants are; a- the presence of supernumerary veins with an incidence of 15-30%, b- the circum-aortic renal vein and retro-aortic renal vein with an incidence of 3% (27, 28). Supernumerary renal veins are much more common on the right side than the left. Since variation influences the technical feasibility of the operation, morphology is specifically significant for transplant surgeons.

In the present study, 18 renal veins were identified in 12 kidneys of six patients. There were three renal veins in the left kidneys of case 1 and case 5 (one main and 2 accessory). There were one main and one accessory veins in four kidneys of which three were ectopic and one was normal. There were 16 veins connected to IVC, one vein of the left kidney of case 5 was connected to left common

iliac vein, and two veins of the right renal vein (kidney) of case 2 were connected to left renal vein.

Conclusions

Nowadays, MDCT and MDCTA have gained significance in the evaluation of the urinary system and the renal vasculature. They are less invasive, easily available and applicable. MDCT supplies information about the morphological variety, position, shape, size and rotation of the kidney. Furthermore, it supplies information not only about the vessel lumen and vessel wall but also about the surrounding structures. We believe that MDCT and MDCTA are beneficial for screening the ectopic kidney and its vasculatures.

Conflicts of interest

The authors declared no conflicts of interest.

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