Malrotated right supernumerary kidney: Case report of a rare anomaly

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Introduction

Supernumerary Kidney is an extremely rare congenital anomaly of the urinary system with less than 100 cases being reported in the literature. The first case of this anomaly was described, more than 350 years ago, by Martius in 1656. It affects both gender equally and due to its rarity, the true incidence cannot be estimated. Herein, we report a case of 18-year-old lady who presented with vague intermittent abdominal pain and found to have malrotated right supernumerary kidney with pelviureteric junction obstruction. She preferred to have regular visits evaluating for possible complications.

Case presentation

Our patient is an 18-year-old lady who presented with a vague and intermittent abdominal pain. She had multiple hospital visits for this problem for the last few years. She does not have any comorbidities and she was not taking any medications. Her physical examination was remarkable for a mass palpable in the right lower quadrant. Routine laboratory investigations were within normal limits.

Ultrasoundography of the abdomen revealed the presence of a right para-aortic mass. She underwent further imaging evaluation with enhanced computed tomography which demonstrated the presence of two right kidneys beside normal left kidney (Fig.1 and Fig.2). The right kidneys were not fused and each has distinct collecting system and vascular supply. The superiorly-located kidney measuring 8.6 by 3.2 cm with a renal artery arising from the aorta at the level of inferior endplate of L1. The larger inferiorly-located kidney measured 8.0 by 5.5 cm and had marked rotational anomaly where the renal pelvis facing anterolaterally. This accessory kidney receives dual blood supply from aorta. One is seen arising from the anterior aspect of the aorta at the level of L4 and runs anteriorly and inferiorly to pass anterior to the pelviureteric junction, where it forms a crisscross with ileocolic branch from the superior mesenteric vessels. The second artery is arising from the right lateral aspect of the aorta at the level of inferior endplate of L4, it runs posterior to the kidney where it passes through a renal parenchymal defect at midportion of the kidney to enter to the renal sinus. The accessory kidney has also dual venous drainage to inferior vena cava with the anterior vein having a retroaortic course. There was a single separate ureter draining this kidney and is inserting anteroposterior-lateral to the insertion site of right ureter. There is obstruction at the pelviureteric junction noted with moderate dilatation of the renal pelvis, this is secondary to crisscross vessels anterior to the junction.

These findings were explained to the patient and provided with the possible management options. Given the minimal symptoms, she chose to have regular follow up visits to be evaluated for possible complications with ultrasonography and laboratory tests.

Discussion

Supernumerary Kidney is an accessory organ with a distinct collective system, arterial supply and venous drainage with a well-defined capsule. It may be either total isolated from the ipsilateral normal kidney or attached to it by a fibrous or parenchymal tissue. It may lie cranial or more commonly caudal to the ipsilateral kidney. The supernumerary kidney is usually present on the left side with right supernumerary kidney, as our case, is less frequent. The size of the
supernumerary kidney is usually smaller compared to the ipsilateral kidney; however, it may vary in size and in our case the supernumerary kidney was larger than the native kidney. The ureter of supernumerary kidney is usually a bifid ureter or less commonly be a separate ureter. The supernumerary kidney should be distinguished from a duplex kidney which is a more common condition, which may not have a separate blood supply. The arterial supply for supernumerary kidney usually originate from the aorta and its venous drainage is via the inferior vena cava. However, sophisticated anatomical variations exist necessitating a thorough evaluation prior to surgical procedures to prevent damage to the supernumerary kidney.

Various associated congenital anomalies have been reported in supernumerary kidney. These include genitourinary anomalies such as horseshoe kidney, ureteral atresia, vaginal atresia, ectopic ureter implantation and duplication of urethra, in addition to gastrointestinal anomalies (e.g. imperforate anus), cardiac anomalies (e.g. ventricular septal defects and coarctation of aorta) and central nervous system malformations (e.g. meningomyelocele). In our case, no such associations were noted.

Although the embryological basis for the development of an accessory kidney has not been fully understood, it is believed that it develops as a result of abnormal division of the nephrogenic cord into two metanephric blastemas at fifth to seventh week of gestation.

This anomaly is often asymptomatic and it usually remains undiagnosed until the fourth decade of life. The most common presenting symptoms are abdominal discomfort, fever and palpable mass. Several pathologic conditions may affect the supernumerary kidney including hydronephrosis, pyelonephritis, renal calculi and ureteropelvic junction obstruction, as in our case, along with benign and malignant renal neoplasms. In cases with ectopic insertion of ureter, supernumerary kidney may present as urinary incontinence. Such pathologies are more likely to occur in cases of supernumerary kidney associated with other anomalies.

The diagnosis is established based on imaging which may include ultrasonography, computed tomography (CT), magnetic resonance imaging (MRI) which are also useful in finding associated anomalies and for evaluation of the anatomy and planning surgical interventions.

The management of patients with supernumerary kidney depends...
on the symptoms and the function of the kidney. For patients with asymptomatic supernumerary kidney, regular follow up is advised. However, nephrectomy may be advised for symptomatic patients or with non-functional kidney.

**Conclusion**

Supernumerary Kidney is a very rare congenital anomaly especially the right-sided one. Although it is asymptomatic in most cases, it can present with vague abdominal pain as in our case.

**Source of support**

None.

**Conflicting interest**

None.

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