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Case Report

Bilateral Supernumerary Kidneys in Conjunction with Horseshoe Anomaly

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ABSTRACT. Bilateral supernumerary kidney is a very rare urogenital anomaly, with four cases reported in the literature thus far. To the best of our knowledge, bilateral supernumerary kidney in conjunction with horseshoe fusion anomaly is not reported till date. Herein, we report a 63-year-old male patient with persistent lower abdominal pain. Complete radiological evaluation including ultrasound, computed tomography scan, excretory urography and retrograde pyelography were done and the diagnosis was established. The dilemma faced in the diagnosis and management of patients with supernumerary kidneys is discussed.

Introduction

Supernumerary kidney is a rare congenital anomaly, with about 100 cases reported in the literature.[1] Bilateral supernumerary kidney is an even rarer anomaly, with only four cases having been reported thus far.[1-4] Herein, we report a case of bilateral supernumerary kidney associated with horseshoe kidney, which is, to the best of our knowledge, being reported for the first time. In this report, an attempt is made to describe the highlights in the diagnosis and management of patients with supernumerary kidneys as well as a thorough review of the literature.

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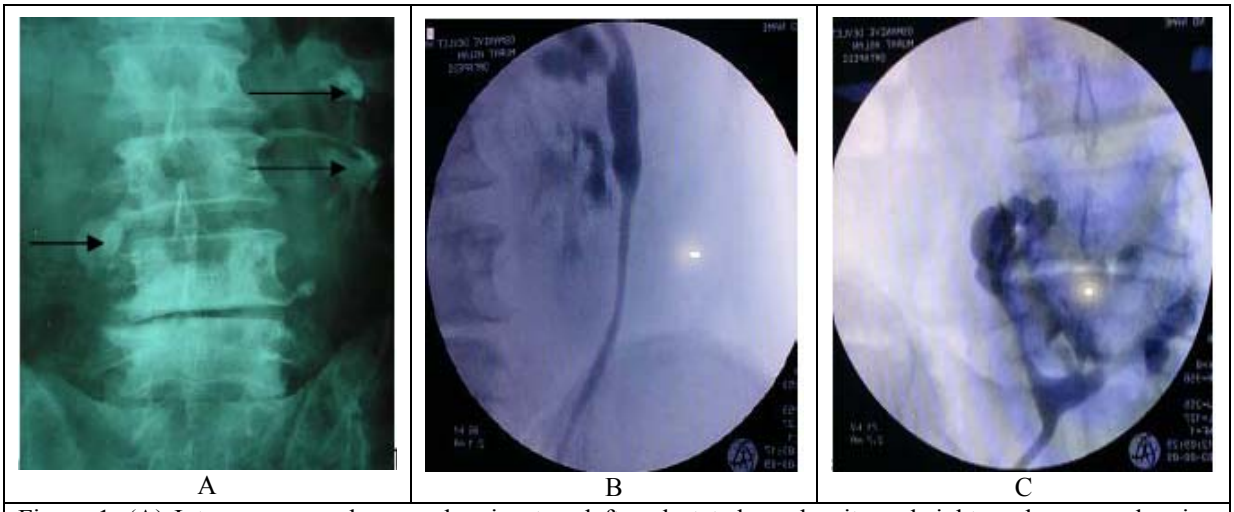
Case Report

A 63-year-old male patient presented with mild, persistent lower abdominal pain. Urine analysis and culture as well as serum blood values were within normal limits. Abdominal ultrasound aroused the suspicion of supernumerary kidney. Excretory urography (IVP) revealed two left malrotated renal units and right ectopic nephrogram with undefined boundaries in the lower-middle abdomen (Figure 1A). Retrograde pyelography (RGP) showed ectopic duplicate collecting systems, with one renal pelvis and one ureter on the right side and two separate pyelograms with one ureter on the left side (Figure 1B and C). Cystoscopy during RGP revealed no lower urinary tract abnormalities. Contrast-enhanced computerized tomography (CT) scan revealed a unified two left malrotated renal units with two separate collecting systems and one ureter. Similarly, on the right side, two unified ectopic kidneys with separate collecting systems and isthmus of horseshoe



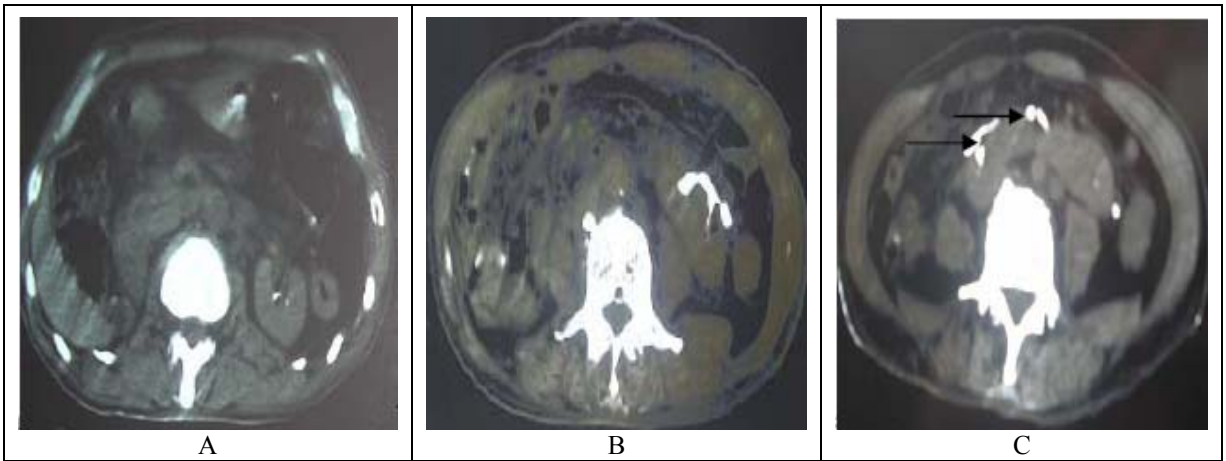
A B C

Figure 1. (A) Intravenous pyelogram showing two left malrotated renal units and right nephrogram showing undefined boundaries (indicated by arrows), (B) retrograde pyelogram showing double left renal collecting system with minimal hydronephrosis and (C) two ectopic collecting systems on the right side with one renal pelvis and one ureter.

anomaly (Figures 2A–C) were observed. After the diagnosis was confirmed and no associated anomalies were detected, the lower abdominal pain was attributed to minimal hydronephrosis and ectopic location of malrotated kidneys. Therefore, a conservative approach was decided with periodic follow-up including urine analysis and culture, blood analysis and abdominal ultrasound.

Discussion

Embryologically, supernumerary kidneys are formed by aberrant division of the nephrogenic cord into two metanephric blastemas with bifurcartion of one bud.[5] The supernumerary kidney may be either totally separate from the normal kidney or connected to it by loose areolar tissue.[5] Supernumerary kidneys have been reported to be associated with various congenital anomalies; ectopic ureteric opening,[6] horseshoe and pseudo-horseshoe kidney,[6] coarctation of the aorta,[7] ureteral and vaginal atresia,[2] complete duplication of the urethra and megaureter.[8] Because of the wide range of combined congenital anomalies and the relative



A B C

Figure 2. (A) Computerized tomography scan showing the upper left renal unit in its normal location, (B) malrotation of the left renal unit; (C) two right ectopic renal collecting systems (indicated by arrows), and the isthmus of horseshoe kidney anomaly between the lower medial two renal units on each side.

rarity of such cases, it is difficult to standardize a protocol for diagnosis and, thus, overdiagnosis with many unnecessary imaging tests is done. Therefore, diagnosis of patients with supernumerary kidneys represents a challenge. In the literature, majority of these cases were diagnosed using many kinds of radiological imaging techniques, including ultrasound, CT scan, IVP, magnetic resonance imaging (MRI), CT angiography and dimercapto succinic acid (DMSA) and diethylene triamine pentacetate (DTPA) scans. While some authors have reported that IVP, CT and ultrasound are adequate for the diagnosis of supernumerary kidneys,[9] other studies have included MRI, DTPA, DMSA and CT angiography in addition.[7,10] We believe that ultrasound, IVP and CT seem to be enough for the diagnosis in majority of the cases. However, sometimes, it may be difficult to recognize the supernumerary kidney on excretory urography alone because supernumerary kidneys are usually smaller and have reduced function. That is why, in our case, RGP was done in addition to IVP, CT and ultrasound; IVP did not show the ureters and the collecting systems clearly, especially on the right side (Figures 1A–C). RGP clearly visualizes the collecting systems, presence of hydronephrosis and duplication of the ureter or double orifice, anomalies that are usually associated with supernumerary kidney. RGP is simple and cheap, and can be performed under intra-urethral anesthesia without need for expensive or sophisticated equipment as in other radiological imaging techniques, and it offers a complete evaluation of the lower urinary tract.

The second challenge regarding supernumerary kidneys is the management of such cases. Because of the associated anomalies, different anatomical environment and non-systemized blood supply, surgical management of these patients is very difficult. Therefore, correct diagnosis is crucial before any surgical intervention. Although CT, IVP, ultrasound and/or RGP are enough to diagnose supernumerary kidneys and associated anomalies, if any, if surgical intervention such as stone removal or nephrectomy is planned, further radiological imaging should be performed to demonstrate the blood supply and the anatomical environment as well as the associated anomalies.

As in other congenital anomalies of the kidneys, these cases are usually susceptible for many complications such as urinary tract infection, stone formation, hydronephrosis, pyonephrosis, pyelonephritis and malignant changes. Therefore, regular follow-up is mandatory. In our case, although it was not expected that the patient would have significant complications in the future, due to his advanced age and the absence of associated ano-malies, a periodic follow-up was suggested.

We conclude that bilateral supernumerary kidneys are very rare congenital anomalies. Because of the wide range of associated anomalies and the scarcity of such cases, diagnosis and management remains difficult. CT scan, IVP, ultrasound and/or RGP are enough to make the correct diagnosis in majority of the cases. However, if surgical intervention is planned, further radiological imaging studies are recommended.

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