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Case study

Four miniature kidneys: supernumerary kidney and multiple organ system anomalies

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Summary More than 350 years after Martius's first reported case in 1656, supernumerary kidney (SNK) continues to fascinate the world of medicine, generating new ideas in the domain of embryogenesis. Association of a normal kidney with a second or third ipsilateral smaller kidney is an extremely rare anomaly with only a total of 81 cases reported until today. We are reporting a case of SNK, clinically diagnosed as right hydronephrosis, associated with an ipsilateral ectopic ureter, a contralateral partially duplicated ureter, and a multiseptate gallbladder. Pathologic examination of the nephrectomy revealed 4 miniature kidneys, joining a dilated ureter through 4 separate conduits. Our patient is the first reported case of SNK with absent ipsilateral normal kidney, presence of more than 3 kidneys on 1 side, and associated anomaly in the gallbladder. This case represents a unique combination of rarities, suggesting insights in the domain of molecular embryology.
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1. Introduction

Supernumerary kidney (SNK) is an extremely rare anomaly. In 2009, the 81st reported case of SNK was published. This is 335 years after the first reported case. All of the cases consist of a normal kidney associated with a usually smaller, second, or even third kidney on the same side. This rare anomaly has been associated in few patients with a second anomaly of the urinary tract, such as renal artery stenosis, ureteral atresia, or ectopia. Most of the cases of SNK have originally been diagnosed as hydronephrosis/pyonephrosis, proving that SNK can be missed by the radiologist and the urologist, and for this reason, the incidence of SNK might be higher than reported. Therefore, raising awareness regarding SNK could lead to future proper diagnosis of this anomaly.

2. Materials and methods

2.1. Patient history

A 29-year-old man with history of hypertension presented to the urology clinic with chronic right back/flank pain, lower urinary tract symptoms of urgency and frequency, subjective gross hematuria, and a history of having a "swollen kidney" that may require an operation. There was no family history of kidney disease. Physical examination was unremarkable, although blood pressure was 191/110 mm Hg.

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Urinalysis showed 56 white blood cells per high-power field and 10 mg/dL of protein. Urine cytology revealed marked pyuria, necrosis, and abundant bacterial cocci in chains. Interestingly, urine culture was negative. The patient had normal overall renal function with a calculated creatinine clearance of 148 mL/min.

The patient underwent a computed tomography (CT) of the abdomen/pelvis with and without contrast, which showed him to have a severe right hydroureteronephrosis, with presumed atrophy of the renal parenchyma along with an ectopic right ureter with termination in the prostatic urethra. The left kidney was noted to have incidental findings of partial duplication of the left ureters. In addition, the patient's gallbladder was noted to be multicystic with areas of curvilinear calcifications. Lastly, the patient was noted to have numerous polyps in the stomach and small and large bowel.

Because of the bowel findings on CT, the patient was evaluated by gastroenterology and underwent a colonoscopy, esophagogastroduodenoscopy, and push enteroscopy. The only finding on these studies was a hyperplastic polyp in the rectum. A right upper quadrant ultrasound revealed a septate gallbladder, without wall thickening, pericholecystic fluid or stones, and a normal common bile duct. The patient underwent a laparoscopic nephrectomy. Cystoscopy was performed at the time of surgery and showed a small mucosal lesion at the bladder neck, possibly consistent with the ectopic implantation of the right ureter. The patient was also noted to have a right ureteral orifice at its orthotopic position, within the bladder. The left ureteral orifice was observed in its orthotopic position, draining clear fluid.

A laparoscopic simple nephrectomy was performed with partial ureterectomy. During the procedure, the adrenal gland was identified attached to the kidney and was removed with the kidney. The gross appearance of the kidney intraoperatively was consistent with a chronically hydronephrotic kidney. The ureter was also massively dilated. There were significant adhesions to surrounding structures including the vena cava, psoas muscle, and diaphragm, consistent with previous inflammation and/or infection. A diaphragmatic injury occurred during the procedure requiring primary closure and chest tube thoracostomy. Postoperatively, the patient had no significant change in overall renal function, and urinalysis revealed resolution of the pyuria. Blood pressure 2 weeks postoperatively was 161/98 mm Hg.

2.2. Macroscopic findings

Gross examination in the surgical pathology service revealed that the specimen designated as right kidney was a hollow and cystic mass weighing 232 g, measuring 15 × 12 × 4 cm, and covered with perinephric fat. The cystic area appeared to be a dilated ureter to which 4 small and miniature kidneys were connected through separate minuscule conduits (Fig. A). Kidneys 1, 2, and 3 were found and photographed at fresh state, and kidney 4 was found after fixation with formalin. These kidneys were covered by perinephric fat and separated from each other by a thin-walled cystic tissue, resembling a dilated ureter, and were situated at different locations at the periphery of the dilated ureter, which had a shiny and focally red mucosal surface. The outlet of the cystic mass was a segment of ureter measuring 4 cm in length and 1 cm in diameter, at the end of which a surgical clamp mark was evident. No stones were found. The sizes of the 4 kidneys (kidneys 1, 2, 3, and 4) were as follow: 4 × 3.5 × 1 cm (Fig. B), 3 × 2 × 0.5 cm (Fig. C), 2 × 1.5 × 0.5 cm (Fig. D), and 1.1 × 1 × 0.4 cm (Fig. E), respectively. On sectioning, each kidney had a normal-appearing capsule, the corticomedullary junction was well demarcated, and the cortex appeared to be thicker than expected, especially when compared with the size of the kidney and relative to the thickness of the medulla. In order of kidneys 1 to 4, the cortex measured 0.4, 0.4, 0.6, and 0.4 cm, respectively, and cortex/medulla ratio was 4:1, 5:1, 4:1, and 3:1, respectively. An adrenal gland, measuring 5.0 × 2.0 × 0.3 cm, was identified on the superior pole of the kidney 1. Further dissection of each kidney revealed a shiny pyelocalyceal system, attached to a miniature conduit measuring 1.8 cm in length and 0.2 cm in diameter, independently joining the cystic ureter.

2.3. Microscopic findings

Sections from 4 small kidneys showed well-developed cortical tissue overlying the relatively thin medullary parenchyma. In the cortical parenchyma, globally sclerotic glomeruli appeared to be restricted to the subcapsular area and represented less than 5% of total glomeruli. Few glomeruli showed a mild increase in size, but the glomeruli in general appeared mature and unremarkable with patent peripheral capillary loops (Fig. B, inset). There was no evidence of increase in mesangial matrix or cellularity, and segmental sclerosis was absent, except for 1 glomerulus adjacent to the area of thyroidization in kidney 2. By special stains (Periodic acid–Schiff and Jones silver stains), the glomerular basement membrane showed normal contour and thickness, with no wrinkling or other abnormality. Mild focal periglomerular fibrosis was evident in a minority of glomeruli. In each kidney, few dilated tubules close to the pyelocalyceal system were present, showing features of thyroidization including tubular dilatation and atrophy and presence of Periodic acid–Schiff–positive hyaline casts with scattered mononuclear inflammatory cells infiltrating between the atrophic tubules. Focal interstitial fibrosis occupying up to 5% of the total parenchymal surface area was present and was mainly located in the area of thyroidization. Remaining kidney parenchyma showed no evidence of inflammation or tubular atrophy. Arteries, especially the ones located close to the area of thyroidization, showed mild-to-moderate fibrous intimal thickening accompanied by reduplication of the elastic lamina. By Verhoeff's elastin stain, fibroelastosis was found in the arteries. No fibroplasia was noted. Some of the arterioles close to the area