Renal dysplasia with extrarenal calyces

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ABSTRACT

We describe a case of simple renal dysplasia with extrarenal calyces of the left kidney in a 2-year-old boy. Other anomalies also included pelviureteral junction obstruction and ectopic ureterocele on the same side, as well as grade V vesico-ureteral reflux on the opposite side.

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The presence of extrarenal calyces is a rare anomaly that was first described in 1925.[1] It is usually associated with other anomalies in the involved kidney as bifid kidney,[1] renal ectopia,[2] and horseshoe kidney.[3] We present a case of left simple renal dysplasia with extrarenal calyces that was present within a spectrum of other urological anomalies in a 2 year-old-boy.

Case Report. A 2-year-old boy presented in the Casualty Department in October 2004 with a distended tender abdomen following a car accident and blunt abdominal trauma. His parents gave a negative history for any medical problems since birth, his blood pressure at the time of presentation was remarkably high (138/106 mm Hg). Enhanced CT scan revealed right hydronephrosis (**Figure 1**). Both ureters appeared markedly dilated and tortuous along their entire length. A huge right retroperitoneal fluid collection was noticed on the right side pushing the bowel to the left side of the abdominal cavity. A 6 Fr. pigtail catheter was inserted percutaneously to drain the collection that proved to be an urinoma caused by leak from the injured right hydronephrotic kidney. The serum creatinine was 1.7 mg/dL at the time of presentation. It increased to 2.2 mg/dL 12 hours later. Bilateral percutaneous nephrostomies were performed. Bilateral antegrade nephrostograms confirmed the CT scan findings (**Figure 2**), and a voiding cystourethrogram revealed presence of grade V right vesico-ureteral reflux. The child was managed conservatively for 2 weeks. His blood pressure dropped to 102/72 mm Hg after Enalapril treatment, and his serum creatinine returned to normal values (0.4 mg/dL) after nephrostomy drainage. Cystoscopy showed a widely dilated right ureteric orifice. The bladder neck was considerably raised and obstructed by a huge ectopic left ureterocele. The orifice of the ureterocele could not be seen. The ureterocele was incised endoscopically. The left kidney was completely nonfunctioning by dimercaptosuccinic acid (DMSA) renal scan that was carried out after cystoscopy. The bladder was opened to excise the left ureterocele and to perform reconstruction of the refluxing right ureterovesical junction. As the right ureteric orifice appeared normal, the decision not to proceed with reconstruction of the right ureterovesical junction was made.

Left nephro-ureterectomy was completed. The left renal vessels were atretic, and the left kidney appeared small and atrophic. It had 3 separate extrarenal

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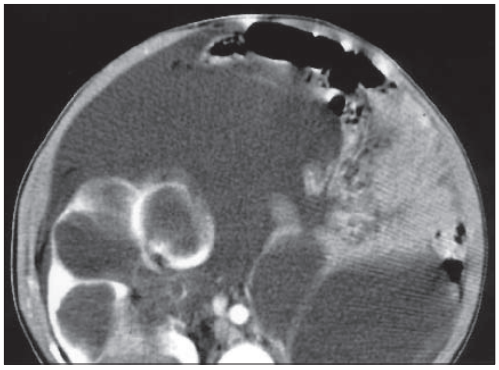

Figure 1 - Enhanced CT scan showing cystic dysplastic left kidney and advanced right hydronephrosis.

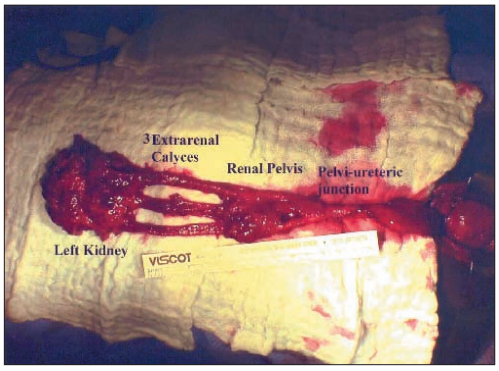

Figure 3 - Left nephro-ureterectomy specimen showing the atrophic left kidney, 3 extrarenal calyces, dilated renal pelvis, pelvi-ureteric junction obstruction and hydroureter.

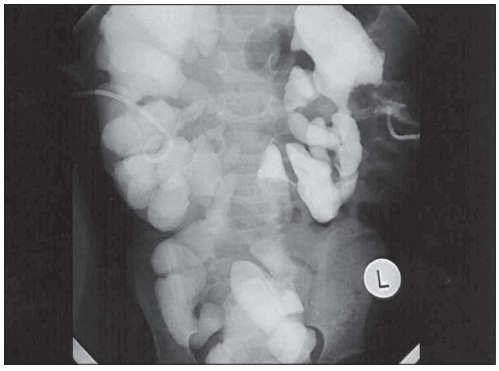

Figure 2 - Bilateral nephrostograms showing a distorted appearance of the calyces in the left kidney, as well as advanced right hydronephrosis and hydroureter.

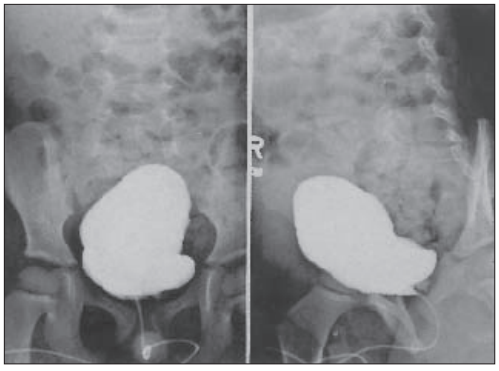

Figure 4 - Voiding cystourethrogram showing good bladder capacity and no evidence of right vesico-ureteric reflux.

calyces; each was approximately 6 cm in length. The pelviureteral junction was narrow and was associated with a markedly dilated renal pelvis (**Figure 3**). The ureter was markedly dilated and tortuous, and ended in a huge ureterocele. Histopathological examination of the removed kidney revealed abortive as well as well-formed tubules and glomeruli of various sizes, surrounded by foci of cellular mesenchyme, fibrosis, smooth muscle, adipose tissue and thick walled blood vessels with mononuclear inflammatory cellular infiltration. The renal calyces, renal pelvis and ureter were all lined by atrophic urothelium with a hypertrophied fibromuscular wall infiltrated by mononuclear inflammatory cells.

Follow up of the patient 10 months after operation showed a very healthy young boy. The renal function remained normal. His blood pressure was 110/65 mm Hg without antihypertensives, and voiding cystourethrogram revealed no evidence of reflux on the right side (**Figure 4**).

Discussion. The presence of the calyces and renal pelvis outside the renal parenchyma is a rare anomaly known as extrarenal calyces. In 1995, Kosinski and Oszukowski[4] observed extrarenal calyces in 3 out of 300 autopsy kidneys. The 1% incidence in their autopsy material is by far a lot more than the number of cases reported in the literature. The total number of cases reported so far was only 20.[1-8] Most cases of extrarenal calyces are associated with other anomalies in the affected kidney. In our case, extrarenal calyces occurred in a dysplastic kidney with pelviureteral junction obstruction and an ectopic left ureterocele. Vesico-ureteral reflux on the right side disappeared spontaneously after left nephro-ureterectomy and removal of the left ectopic ureterocele. Right reflux has most probably resulted from the significant infravesical obstruction that was caused by the left ureterocele.

During exploration of our case, the 3 major calyces were abnormally long. They were all grouped within a single sheath, and were initially mistaken with the proximal ureter. Despite the rare incidence of extrarenal calyces, it is important to bear this anomaly in mind when operating on a kidney with distorted calyceal appearance in preoperative imaging studies.

This would safeguard against inadvertent injury of the calyces when operating on a well functioning kidney.[8] In our case, renal dysplasia caused renal hypertension that was reversible after nephrectomy. Abnormalities of the lower urinary tract as ureteroceles, ectopic ureters or vesico-ureteral reflux are common in children with renal dysplasia.[9] Extrarenal calyces were also present in our case.

References

1. Eisendrath DN. Report of case of hydronephrosis in a kidney with extrarenal calyces. ***J Urol*** 1925; 13: 51-58.
2. Malament M, Shwartz B, Nagamatsu GR. Extrarenal calyces: their relationship to renal disease. ***Am J Roentgenol*** 1961; 86: 823-829.
3. Mohanty C, Ray B, Samaratunga U, Singh G. Horseshoe kidney with extrarenal calyces. A case report. ***J Anat Soc India*** 2002; 51: 57-58.
4. Kosinski H, Oszukowski P. Calices renales majores extrarenales. ***Folia Morphol (Warsz)*** 1995; 54: 181-186.
5. Veseen LL. Extrarenal calices. ***J Urol*** 1933; 30: 487-490.
6. Dunn GP, Gibson TE. Extrarenal calyces. ***Urol Cutaneous Rev*** 1951; 55: 259-263.
7. Rao DVN, Sharma SK, Rao NS, Bapna BC. Extrarenal calyces with complications. A case report. ***Aust NZJ Surg*** 1972; 42: 178-179.
8. Garg P, Godara R, Karwasra RK. Extrarenal calyces: a rarity. ***Urol Int*** 2003; 71: 331-332.
9. Karmazyn B, Zerin JM. Lower urinary tract abnormalities in children with multicystic dysplastic kidney. ***Radiology*** 1997; 203: 223-226.