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Jacobs Journal of Nephrology and Urology

Image Case Report

Congenital Ureteropelvic Junction Obstruction

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Received: 06-15-2015

Accepted: 07-24-2015

Published: 07-30-2015

Introduction

Ureteropelvic junction (UPJ) obstruction is defined as an obstruction of the flow of urine from the renal pelvis to the proximal ureter. The condition is frequently encountered by both adult and pediatric urologists. The reported incidence of ureteropelvic junction (UPJ) obstruction is 1 in 500 live births and is more common in males [1]. Congenital abnormalities may be observed in both adults and children, but adults may also present with UPJ obstruction following previous surgery or other disorders that can cause inflammation of the upper urinary tract. Crossing vessels compressing or distorting the UPJ may be the sole cause of ureteral outflow obstruction. More commonly, however, they coexist with other causes of UPJ obstruction [2].

The critical decision to be made in dealing with suspected UPJ obstruction is whether the radiologic findings correlate with the physiologic picture. In other words, severely dilated hydronephrotic kidneys may, in fact, drain well when studied appropriately. Defining the exact anatomy and function of these kidneys is crucial when evaluating and treating these patients.

Case Report

A 20-year-old male presented to the emergency department complaining of intermittent flank pain for two months. He denied associated symptoms or modifying factors. Physical examination revealed costovertebral tenderness. An ultrasound (Figure 1), and subsequently a computed tomography (CT) scan of the abdomen (Figure 2) were performed. The ultrasound showed normal echogenicity of the kidneys bilaterally. The left kidney measured 18.6 x 10.4 x 10.1 cm with severe left-sided hydroureteronephrosis containing echogenic debris. Vascular flow was present demonstrated bilaterally. The CT scan also showed severe left hydronephrosis with extensive cortical thinning and without hydroureter.

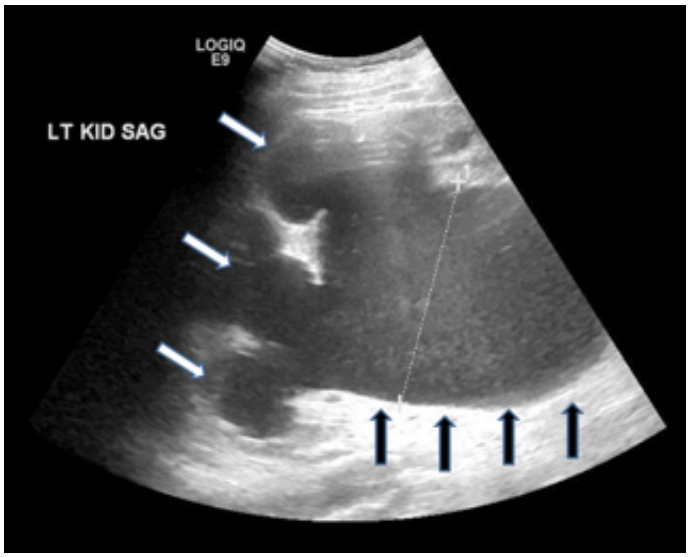

Figure 1. Ultrasound of the kidney demonstrating the "bear claw" appearance of the kidney. White arrows indicate the renal calyces and black arrows identify the renal pelvis.

No radiopaque calcification was seen and the urinary bladder was unremarkable. There was no evidence of mass or other intra-abdominal pathology. Laboratory examination revealed normal kidney function with a blood urea nitrogen (BUN) of 12 mg/dl, creatinine of 0.96 mg/dl and glomerular filtration rate (GFR) of 100. The patient was seen by urology and diagnosed with a congenital ureteropelvic junction (UPJ) obstruction. A percutaneous nephrostomy tube was

successfully placed by interventional radiology as a temporizing measure. His renal function remained normal, however the pain persisted. A CT urogram showed stable left renal cortical scarring and atrophy with improvement in the left hydronephrosis. A nuclear medicine renal scan with Lasix was performed to assess flow and function. Results showed poor perfusion and poor uptake by the left kidney with the left kidney accounting for 35% of the total renal function. He subsequently underwent a robotic assisted laparoscopic pyeloplasty where the obstruction was identified as a stricture at the dependent portion of the renal pelvis. This segment was excised. The patient had an uneventful recovery and was discharged on hospital day #7.

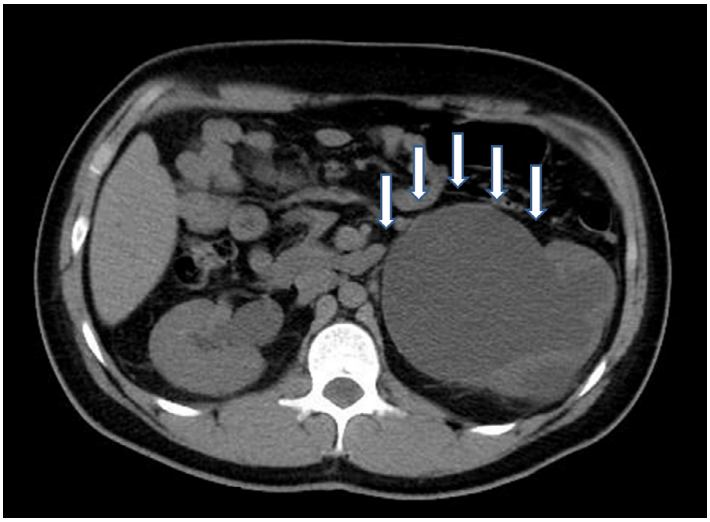

Figure 2. Computed tomography of the abdomen and pelvis identifying hydronephrosis of the left kidney (arrows)."

Discussion

Congenital ureteropelvic junction (UPJ) obstruction

Congenital UPJ obstruction is defined as an obstruction of urine flow from the renal pelvis to the proximal ureter. Reports of bilateral involvement range from 10-40% [3]. Congenital UPJ obstruction is usually caused by intrinsic stenosis of the proximal ureter, less commonly by extrinsic compression of the UPJ by an aberrant or accessory renal artery or arterial branch, and rarely from intraluminal folds [4]. Clinically, a palpable abdominal mass, urinary tract infections, hematuria or failure to thrive may be present. Older children and young adults may report nonspecific abdominal or flank pain and nausea or vomiting could be present. These symptoms may be exacerbated during brisk diuresis.

In suspected cases, initial evaluation is via ultrasound to assess for hydronephrosis. Subsequently, voiding cystourethrogram can evaluate for the presence of vesicoureteral reflux, and diuretic renography can assess for obstruction. CT scan may be performed, but is not the ideal study in children due to radiation risk. The differential function is important in determining the need for intervention. Functionally significant obstruction is often diagnosed with diuretic renal scanning. Dynamic contrast-enhanced magnetic resonance urography (MRU) has also recently been used in assessing UPJ obstruction. The advantages of MRU are the lack of radiation exposure along with information regarding renal anatomy and function with a single study.

Asymptomatic individuals may be observed through serial ultrasounds and renal function monitoring. Symptomatic cases or those involving renal function deterioration typically require dismembered pyeloplasty. This procedure consists of excision of the ureteropelvic junction and part of the pelvis and reattachment of the normal end of the ureter to the remaining pelvis.

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