A 7-Year-Old Girl with Periodic Flank Discomfort

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A 7-year-old girl with a 12-month history of intermittent left flank pain was transferred to our institution from an outside hospital. Severe pain occurred about once per month and was associated with multiple episodes of nonbloody, nonbilious emesis. The pain did not radiate and was not alleviated or exacerbated by anything else. Symptoms lasted 2 to 3 days and then gradually resolved. The patient denied associated dysuria, hematuria, fever, or diarrhea. The patient had visited her pediatrician’s office several times, but diagnostic work-ups for urinary tract infection and constipation were negative. A recent renal ultrasound revealed severe left hydronephrosis. Due to recurrent pain, the patient was seen at an outside hospital, where a computed tomography (CT) scan was ordered (Figure 1).

Her most current painful episode started 5 days prior to admission. Initial physical examination revealed a well-developed and well-nourished child with normal vital signs, including normal blood pressure. Her anthropometric parameters were within normal limits for age. HEENT (head, eyes, ears, nose, and throat) examination, respiratory examination, and cardiovascular examination were all unremarkable. On abdominal examination there was left costo-vertebral angle tenderness but no palpable mass. Her neurologic assessment was normal. No rashes or skin lesions were present.

Figure 1. Abdominal computed tomography imaging of the (A) sagittal and the (B) cross-sectional revealing marked left hydronephrosis.

For diagnosis, see page 101

Editor’s note: Each month, this department features a discussion of an unusual diagnosis. A description and images are presented, followed by the diagnosis and an explanation of how the diagnosis was determined. As always, your comments are welcome via email at pedann@Healio.com.
Diagnosis:
Uretero-Pelvic Junction Obstruction

Her CT scan showed a marked left hydronephrosis, suggesting an uretero-pelvic junction obstruction (UPJO). At that time, referral to a pediatric urologist was made, but the patient’s current episode of pain and subsequent emergency department visit led to transfer to our institution for a prompt evaluation. In our center, a diuretic nuclear renography showed evidence of delayed excretion (Figure 2), confirming the diagnosis of left UPJO. The patient later had a robotic-assisted laparoscopic dismembered left pyeloplasty.

DISCUSSION

UPJO is a partial or total blockage of urine flow at the level where the renal pelvis converges with the ureter. Congenital unilateral UPJO is the most common prenatally acquired urological obstructive disease. Incidence of UPJO is about 1 in 400 live births screened by antenatal ultrasound. It occurs more often in boys than in girls, and the left kidney is affected more frequently than the right.

Classically, urological obstructions are classified as either intrinsic or extrinsic in cause. Congenital UPJO typically results from an intrinsic aperistaltic segment of the ureter. Currently, no well-defined pathogenesis of this embryologic anatomical malformation has been recognized. Although controversial, an abnormal distribution of Cajal’s uroepithelial interstitial cells has been proposed as an underlying mechanism. Altered uroepithelial expression of type 2 helper (Th2) cytokines (interleukin-5 and eotaxin) and growth factors (epidermal growth factor and transforming growth factor-beta) have also been associated with UPJO. Crossing “aberrant” vessels have been reported as an extrinsic cause of UPJO, but they represent only a minority of documented cases of UPJO.

Since the routine introduction of prenatal imaging, UPJO is usually

![Figure 2. A diuretic radionuclide renograph showing left kidney delay excretion, which suggests uretero-pelvic junction obstruction (red curve). Split renal function showing “fair” left renal uptake (left: 39.2% versus right: 60.8%).]
identified before any clinical manifestation appears. Older children may experience back or flank pain, which may be associated with nausea and vomiting.

This intermittent clinical picture of UPJO, first described by Josef Dietl in 1864, is usually self-limited. Symptom-free periods are variable in duration. Other symptoms might include hematuria and/or hypertension. It can also present in association with a urinary tract infection (eg, fever, dysuria, or incontinence).

Children with UPJO sometimes are misdiagnosed with cyclic vomiting syndrome. It is important to include UPJO in the differential diagnosis of this unusual entity to decrease renal morbidity.

Presently, the diagnosis of UPJO is generally suspected either by hydronephrosis on antenatal ultrasonography or by a flank mass on physical examination during infancy. Diuretic nuclear renography testing is ordinarily used for diagnosis of UPJO because it provides quantitative data concerning differential renal function and urinary flow (Figure 2). Radionuclide delayed excretion measurement correlates with the degree of obstruction and is particularly helpful in differentiating UPJO from severe hydronephrosis secondary to vesicoureteral reflux. In some cases, CT imaging can further help to characterize the anatomical lesion.

The overall goal in treating UPJO is to preserve renal function. Most experts recommend a restrained interventional approach. There are no randomized trials that provide evidence for the optimal management of congenital UPJO. The current management regimen is based on a small number of observational case reports. Common indications for surgical intervention include recurrent flank pain, progressive impairment of ipsilateral renal function, hypertension, history of pyelonephritis, or development of kidney stones. Bilateral UPJO is a rare cause of neonatal renal insufficiency and it represents a neonatal emergency. When urological intervention is indicated, the procedure of choice has historically been dismembered pyeloplasty. Nevertheless, minimally invasive surgery (eg, laparoscopic, balloon dilation, or robotic), even in younger children, has recently gained acceptance as an alternative approach. Prognosis is usually quite good. Some investigators have made attempts to identify urinary biomarkers in infants with UPJO to predict the need for surgical intervention in early stages.

REFERENCES