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Severe ureteropelvic junction obstruction with massive hydronephrosis causing compression of the inferior vena cava

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ABSTRACT

A 7-week-old girl presented with tachypnea and abdominal distension. Abdominal ultrasonography demonstrated a 16 cm cystic mass in the right abdomen. Contrast-enhanced T2-weighted magnetic resonance imaging revealed severe hydronephrosis of the right kidney with thinning of the renal parenchyma consistent with a massive ureteropelvic junction (UPJ) obstruction. A percutaneous nephrostomy tube was placed for decompression of her right kidney and 1270 ml of fluid was drained. The patient underwent an open pyeloplasty. Her follow-up renal ultrasonography demonstrated resolution of her severe hydronephrosis.

Introduction

Congenital ureteropelvic junction obstruction (UPJ) is a common etiology of hydronephrosis in neonates and pediatrics. The urinary tract dilation classification system defines high risk hydronephrosis as an anterior posterior renal pelvis diameter ≥15 mm. Massive dilation of the renal pelvis where the kidney occupies the hemiabdomen, crosses midline, or is at least five vertebral bodies in length was first described as "giant hydronephrosis" by Crooks et al., in 1979.[1] In children, this is defined as the kidney containing the equivalent of one day's urinary output.[2] Hydronephrosis presenting as a cystic abdominal mass is uncommon but has been previously reported. However, massive hydronephrosis causing compression of the inferior vena cava (IVC) in children is exceedingly rare with two cases reported in the literature.[3,4] Here, we report a 7-week old girl presenting with hypoxia and acute respiratory distress who was found to have a cystic abdominal mass on exam and ultimately diagnosed with a severe UPJ obstruction and massive hydronpehrosis causing IVC compression.

Case presentation

A 7-week-old girl presented with tachypnea and abdominal distension. Her medical history was remarkable for right antenatal and postnatal hydronephrosis. Voiding cystourethrography was negative for vesicoureteral reflux. Her abdomen was tense and grossly distended. The patient's lips appeared hypoxic when lying supine, which improved when rotated on her side, concerning for intermittent compression of the IVC. Abdominal ultrasonography demonstrated a 16 cm cystic mass in the right abdomen. Axial (Fig. 1A) coronal (Fig. 1B) views of contrast-enhanced T2-weighted magnetic resonance imaging (MRI) revealed severe hydronephrosis of the right kidney with thinning of the renal parenchyma consistent with a massive UPJ obstruction. A percutaneous nephrostomy tube was placed and 1270 ml of fluid was drained. A nuclear medicine renal scan demonstrated a split function of 29% and 71% in the right and left kidneys, respectively, and severely delayed drainage of the right kidney confirming the diagnosis of UPJ obstruction. The patient underwent an open pyeloplasty. Her follow-up renal ultrasonography and nuclear medicine renal scan demonstrated resolution of her severe hydronephrosis and improved renal function, respectively.

Discussion

Cystic abdominal masses in children are not uncommon. While many are detected with prenatal ultrasound, others may present with various symptoms and physical exam findings based on the size, location, and mass effect on adjacent organs. The differential diagnosis of cystic abdominal masses in children may range from ovarian cysts, renal cystic masses such as multicystic kidney disease or malignancy such as cystic nephroblastoma, as well as hydronephrosis, hepaticopancreatic cyst or pseudocyst. Ultrasound is the primary imaging modality of choice for initial evaluation of a cystic abdominal mass. MRI may subsequently be obtained for complete characterization. In this case, the patient's massive right-sided hydronephrosis was intermittently compressing the IVC posterior-laterally and responsible for her physical exam findings of

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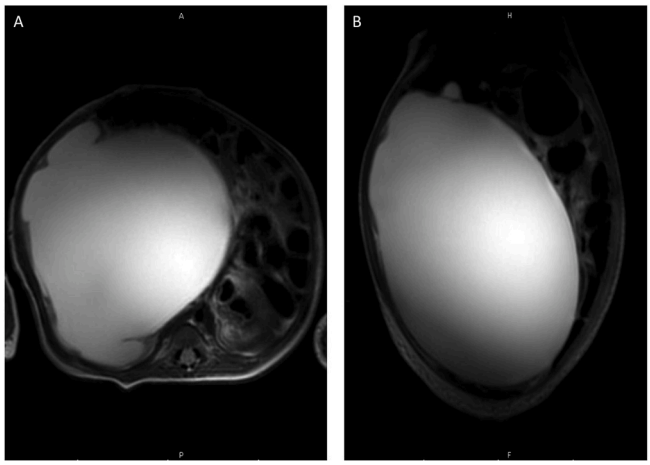

Fig. 1. Axial (A) and coronal (B) views of contrast-enhanced T2-weighted magnetic resonance imaging demonstrating severe enlargement of the right renal pelvis measuring 15.1× 1.1 × 1.3 cm, with marked thinning of the right kidney occupying almost the entire abdominal cavity and causing mass effect on the liver superiorly, bladder inferiorly, aorta and inferior vena cava laterally.

positional hypoxia. MRI enabled for excellent appreciation of renal parenchymal thickness, severe dilated renal pelvis, and its relation to surrounding visceral structures and major vessels.

One approach to acute management in this setting includes urgent surgical diversion with cutaneous pyelostomy creation. However, we opted for percutaneous nephrostomy tube drainage as this can be done quickly with local sedation, ultrasound guidance, rapid symptomatic relief, and without long-term disruption of the UPJ to affect a future urologic reconstruction. Indeed, in a prospective longitudinal study describing evaluation, management, and outcomes of 35 patients with massive hydronephrosis, all were managed with percutaneous nephrostomy tube drainage with the average immediate nephrostomy tube output 1.9 ± 0.4 L in children.[5] In this study, the children were on average 10 months in age in comparison to our 7-week old baby with 1.3L immediate drain output. Similarly, the patient's kidney had adequate renal function with split function determined to be 29% and was effectively management with an open pyeloplasty. It is not uncommon for long-standing severe obstruction to ultimately result in the loss of the renal unit and subsequently a non-functioning kidney, which was described in 31% of patients.

Accurate diagnosis of a UPJ obstruction with early decompression and ultimate reconstruction is paramount to prevent further renal impairment. Massive hydronephrsosis, if left undiagnosed, may result in long-term complications such as hypertension, forniceal rupture, and renal failure. A history of hydronephrosis in utero should raise suspicion for an UPJ obstruction. We describe our experience with MRI in evaluation of massive hydronephrosis and rapid nephrostomy tube decompression for symptomatic relief in the setting of vena caval compression prior to definitive reconstruction.

Conclusion

Severe UPJ obstruction may cause massive hydronephrosis with local extension and compression of the IVC. The differential diagnosis of a cystic abdominal mass in a neonate or child is broad but a history of hydronephrosis in utero should raise suspicion for an UPJ obstruction. While ultrasonography is an excellent initial imaging modality in children presenting with a flank or abdominal mass, MRI provides an adjunctive non-ionizing radiation diagnostic approach for complete characterization.

Declaration of competing interest

The authors declare that there is no conflict of interest regarding the publication of this paper.

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