International Journal of Pediatrics and Adolescent Medicine 9 (2022) 179–181

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Contents lists available at ScienceDirect

International Journal of Pediatrics and Adolescent Medicine

journal homepage: http://www.elsevier.com/locate/ijpam

Acute abdominal pain localized in right iliac fossa: Not always acute appendicitis. A case of giant hydronephrosis in an 8-year-old boy and literature overview

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ARTICLE INFO

Article history:
Received 4 October 2021
Accepted 8 December 2021
Available online 21 December 2021

Keywords:
Giant hydronephrosis
Abdominal pain
Children and diagnosis management

ABSTRACT

Abdominal pain is a frequent complaint in children, leading them to seek medical attention. It can have several causes, though acute appendicitis is the most feared diagnosis when pain is localized in the right iliac fossa. We report a case of an 8-year-old boy with the complaint of acute abdominal pain, initially referred by his family doctor to a radiologist for an abdominal ultrasound (US) for suspected acute appendicitis. A fortuitous diagnosis of giant hydronephrosis (GH) was made upon admission, which showed the palpation of a huge poorly delineated abdominal mass that was probably missed at the previous examination by the general physician (GP). Uroscan confirmed the diagnosis of GH secondary to obstruction at the ureteropelvic junction. A renal MAG3 (mercaptuacetyltriglycine) scan showed revealed differential renal function (15%) on the right side, normal on the left side. Robot-assisted right pyeloplasty with the transposition of right lower polar vessels via *trans*-peritoneal laparoscopy was performed, and JJ probe left in-situ for a month. The boy is doing well and is under active follow-up. GH is rare; its diagnosis requires both meticulous examination and a high index of suspicion. Its management is uncodified but in children, pyeloplasty is preferred to nephrectomy.

1. Introduction

Abdominal pain is a common complaint in children. Although with medical causes in a large majority of cases, surgical etiologies are the most feared due to potential complications. When the pain is localized in the right iliac fossa, diagnosis of acute appendicitis is the most thought of choice, requiring prompt investigation and case management. Depending on the age of the patient, however, several other differential diagnoses need to be ruled out. Among these, constipation, pyelonephritis, streptococcal pharyngitis, cholecystitis, Henoch-Schonlein purpura, mesenteric adenitis (viral or bacterial), inflammatory bowel disease, cecal diverticulitis, urolithiasis, acute appendicitis, acute pancreatitis, incarcerated inguinal hernia, abdominal masses, trauma, etc. are the common diagnoses [1–4].

Giant hydronephrosis (GH) is rare, in developed countries, because of the large diffusion of medical imaging and better obstetrical surveillance and antenatal diagnosis. GH may nevertheless present later in life by diffuse abdominal pain, progressive abdominal distension with or without associated bouts of fever. Several cases have been reported in neonates, children, and even adults [5–10].

We report a novel case of GH diagnosed fortuitously in an 8-year-old boy who was initially investigated by his family doctor for acute abdominal pain located in the right iliac fossa. We aim to remind clinicians of the necessity for a meticulous clinical examination and the need for a high index of suspicion while caring for children complaining of abdominal pain.

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Peer review under responsibility of King Faisal Specialist Hospital & Research Centre (General Organization), Saudi Arabia.

https://doi.org/10.1016/j.ijpam.2021.12.001

1.1. Case presentation

An 8-year-old male child was referred to us by a radiologist from a local private clinic for right-sided renal distension discovered fortuitously during an abdominal ultrasound (US) ordered by the family doctor for acute abdominal pain localized in the right iliac fossa associated with vomiting. The boy's past medical history was positive for adenoidectomy with *trans*-tympanic aerators placement, two episodes of bronchiolitis (December 2011, January 2012), hemangioma treated with beta-blockers, pneumonia (2012), and asthma. His perinatal history was unremarkable. He had never presented with urinary tract infection nor had he previously been diagnosed with any renal dilatation. Upon admission in our department, the boy was afebrile, weighted 27.9 Kg, had a height of 128 cm, body mass index of 17.02 Kg/m^2. Heart and respiratory rates were normal for age, oxygen saturation was 98% at room air, blood pressure 125/93 mm Hg. He was oliguric (300 ml urines in 24h) (differential renal function-DFG was 97ml/min/1.73m^2 according to Schwartz formula). He spontaneously complained of mild abdominal pain. Clinical examination revealed a mildly tender abdomen with a huge uncharacterizable mass covering the right hypochondrium, right lumbar region, right iliac fossa, extending beyond the abdominal midline. Abdominal US (Fig. 1) revealed massive pyelo-calyceal dilatation in favor of ureteropelvic junction stenosis. Urines strip test was negative for blood, nitrites, and nitrate; urine culture remained sterile; blood urea was 5.5 mmol/L; plasma creatinine 54 μmol/L; C-reactive protein was 24.2 mg/L (N < 5 mg/l); complete blood count and plasma electrolytes were within normal limits. A uroscan was performed the following day; it shows (Fig. 2A & 2B) significant right renal dilatation with enlarged pelvis measured at 58 mm, associated with the almost destruction of the parenchyma. Absence of vascular abnormalities, the normal contralateral kidney were observed. We diagnosed him with GH secondary to ureteropelvic junction stenosis. The renal MAG3 scintigraphy showed asymmetrical renal function, with normal left kidney function and only 15% renal function with a completely flat curve on the right side. Therefore, a choice was made to preserve his right kidney, and a right pyeloplasty with transposition of right lower polar vessels by robot-assisted *trans*-peritoneal laparoscopy was performed with a JJ probe left in-situ for a month (Dr. A. Victor). The boy is currently doing well and is under close follow-up.

2. Discussion

Hydronephrosis results from dilatation of the pelvicalyceal system secondary to obstruction and stasis of urinary flow, mainly at the ureteropelvic junction. In general pediatric medicine, both physiological and pathological hydronephrosis are more common, representing 50%–87% of the urinary tract anomalies diagnosed antenatally [11].

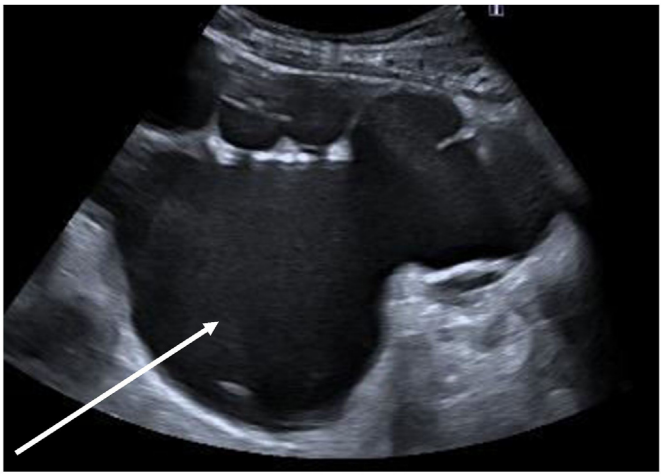

Fig. 1. The abdominal US showing massive right renal dilatation.

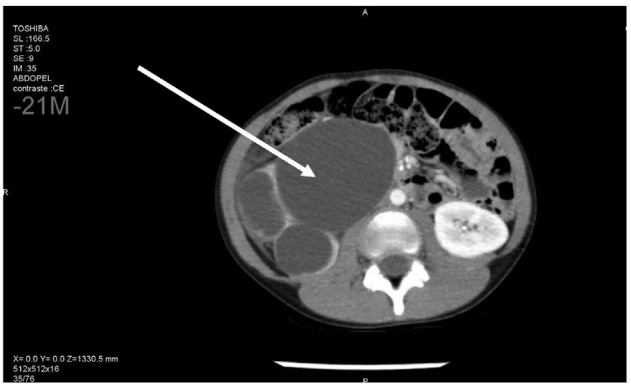

Fig. 2a. Uroscan arterial images showing giant right renal hydronephrosis. A- Transverse section.

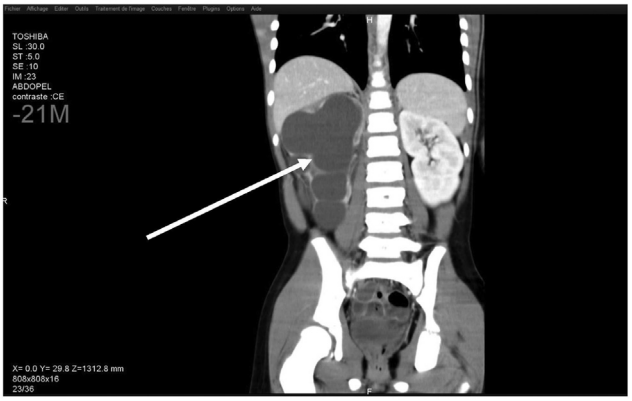

Fig. 2b. Uroscan arterial images showing giant right renal hydronephrosis. B- Coronal section.

Most of these cases are silent, but reports of GH in neonates have also been reported in the literature [5,11].

GH is rare, with a little >500 cases published [5]. Sterling (1939), initially described GH as containing >1000 ml of fluid, with a mass filling the abdomen [12]. This definition was revised by Crooks et al. who suggested that GH was a renal mass that occupies half of the abdominal cavity, meets or crosses the midline, and extends vertically to five vertebrae at least [13]. GH is more often unilateral but may also be bilateral. Huge GH with urine volumes as high as 24–80 l have been reported in adults. In children, these rare cases are reported essentially from developing countries where antenatal and postnatal diagnosis and management are often delayed [8,14,15]. The differential diagnoses to consider should include acute appendicitis, as initially thought of by the family doctor in our case, nephrolithiasis, abdominal cysts, retroperitoneal hematoma, hepatomegaly, splenomegaly, ascites, etc. [16] In children, the principal cause of GH is the obstruction at the pyelo-ureteral junction, present in all patients in the case series by Kaura et al. [5] GH may also result from renal pelvic stones and congenital malformations [17,18].

2.1. Clinical presentation

GH presenting symptoms are diverse and can be misleading in the absence of a meticulous clinical examination, as illustrated in

the current case report. A case of an adult patient thought initially to have a renal tumor has also been reported [19]. GH can present with abdominal pain, progressive abdominal distension, obstruction of the gastrointestinal tract or that of the contralateral urinary system, recurrent urinary tract infection, or hematuria. A case of respiratory embarrassment with compression of inferior vena cava has also been reported in an infant [20].

Circumstances under which GH may be diagnosed are variable and age-dependent: newborn infants with an antenatal history of hydronephrosis may present with GH as a complication in their early life [5,21]. In a study of 35 patients (19 adults, 16 children), Kaura et al. [5] reported flank pain (100%) and abdominal lump (63%) as the most common presenting symptoms in adults, whereas children presented more often with an abdominal lump (100%), followed by the flank pain (87.5%) and fever (75%). In our patient, the diagnosis was likely missed by the family doctor as the child had no notable past medical history, was afebrile, and complained only of abdominal pain. Therefore, he was referred initially for abdominal ultrasound ordered for suspected acute appendicitis. In some rare cases, GH has presented with intestinal obstruction, respiratory distress, hypertension, pedal edema, obstructive jaundice, and contralateral ureteropelvic junction obstruction [16]. Giant hydronephrosis tends to affect, preferentially, the male gender and the right kidney in 83% and 66%, respectively [5,22]. Abdominal ultrasound and uroscan are the first imaging modalities to confirm the diagnosis, whereas renal scintigraphy allows the assessment of the residual renal function and guides the appropriate management choice.

2.2. Management

Due to the lack of international consensus on its management, GH patient care is tailored to each individual, and it relatively differs between children and adults. The timing of the diagnosis also likely influences the management option. A case diagnosed early enough and before renal tissue destruction has more chances of being treated conservatively. Overall, the extant literature suggests that children with GH should be managed conservatively. There is, however, some discrepancy between authors as to which lower DRF should guide the choice between nephrectomy and pyeloplasty. In a report of 12 patients with reduced DRF (<10%), all had improvement soon after a preliminary nephrostomy preceding pyeloplasty [23].

Several other reports favor this conservative approach in children with GH, even in those with DRF <10% [24–27]. However, Kaura et al. reported that patients with DRF <15% (eight adults, three children) underwent nephrectomy, with pyeloplasty having been performed in only one adult with DRF <15%, reasons for this are unclear [6]. Our patient with a DRF of 15% benefited from robot-assisted transperitoneal laparoscopy which presents several advantages such as a minimally invasive technique, smaller incisions, shorter length of hospital stay, etc. with long-term outcomes that were comparable to open procedures [28,29], he is doing well and his follow-up is underway.

3. Conclusion

We conclude that GH is rare and its presenting symptoms are extremely variable. The diagnosis which can be tricky requires both meticulous clinical examination and a high index of suspicion. Its management remains uncodified in literature, but pyeloplasty is preferred to nephrectomy in children, especially when differential renal function is >10% in unilateral cases.

Declaration of interests

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

Funding statement

No funding received.

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