Medusa's Curls: Ureteropelvic Junction Obstruction Secondary to Multiple Long Intraluminal Polyps

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Fibroepithelial polyps represent a rare cause of intrinsic ureteropelvic junction obstruction in the pediatric population, accounting for less than 5% of cases. Herein, we present this peculiar case of a 10-year-old boy with multiple large intraluminal FEPs resembling Medusa's hair and the challenges associated with its subsequent treatment plan. UROLOGY 130: 138–141, 2019. © 2019 Elsevier Inc.

Fibroepithelial polyps (FEPs) are benign lesions of mesodermal origin that can cause intrinsic ureteropelvic junction obstruction (UPJO) in children, accounting for less than 5% of cases. FEPs are overwhelmingly more common in males (92%), multiple in appearance, and seen unilaterally in 67% of the situations.[1] In addition to their rare nature, FEPs may pose a significant diagnostic challenge in children, as they cannot be definitively detected preoperatively by ultrasonography (US). The fact that FEPs may appear in any part of the ureter, extending from its proximal to distal end, makes their complete excision challenging. Herein, we present this unusual case of a 10-year-old boy with multiple large intraluminal FEPs involving the proximal and mid ureteral segments resembling the Medusa's hair mythology goddess, and the subsequent treatment plan.

CASE REPORT

A 10-year-old male presented to the ER with a 5-day history of left flank pain and hematuria. US revealed Society of Fetal Urology (SFU) grade 2 hydronephrosis (HN) with a renal pelvis anteroposterior diameter (APD) of 10 mm and mild dilation of the left proximal ureter (5 mm), with no evidence of stones. All symptoms spontaneously resolved with analgesic treatment, and a 2-month follow-up appointment was scheduled in the pediatric urology clinic.

Follow-up renal bladder ultrasound (US) at our outpatient clinic demonstrated left-sided SFU grade II HN with APD of 15 mm. His history revealed no further episodes of flank pain, hematuria, or urinary tract infections. Two weeks later, the child presented again to the ER with left-sided flank pain, nausea and vomiting, consistent with acute Dietl's crisis. US in the acute setting demonstrated significant worsening of left sided HN with an APD of 32 mm (Fig. 1A). An urgent renal scan revealed symmetric differential renal function with a prolonged diuretic $T^1/_2$ on the left side of 31 minutes, suggestive of UPJO (Fig. 1B). Due to his intermittent symptomatic presentations and renogram findings, the patient was taken to the operating room for a left laparoscopic dismembered stented pyeloplasty **(Attached video)**. Intra-operatively, multiple long intraluminal FEPs were seen at the UPJ and extending into the proximal ureter for 3 to 4 cm (Fig. 2A, B). Due to the long ureteral segment affected by the FEPs, renal descensus and extensive distal mobilization of the ureter were performed to allow for a tension-free anastomosis between the spatulated ureter and renal pelvis. The patient had a smooth postoperative recovery being discharged after 3 days.

Four weeks postoperatively, at the time of stent removal, a retrograde pyelogram was performed, revealing a filling defect in the transition of the proximal to mid ureter suggestive of residual FEPs. Flexible ureteroscopy identified a few small polyps and a large pedunculated one with a thin stalk in the mid ureter, which were easily excised with holmium laser (Fig. 3A-C). The postoperative course was uneventful. The previous stent was exchanged and removed a month later without any complications. Follow-up US revealed substantial improvement of the left side HN and no ureteral dilatation (Fig. 3D). The child no longer complained of abdominal pain postsurgery.

COMMENT

UPJO may be caused by intrinsic or extrinsic factors. FEPs represent a rare, intrinsic etiology of UPJO in children.[1-4] In the senior author's personal experience of 241 consecutive pediatric pyeloplasties, 58 cases have presented with

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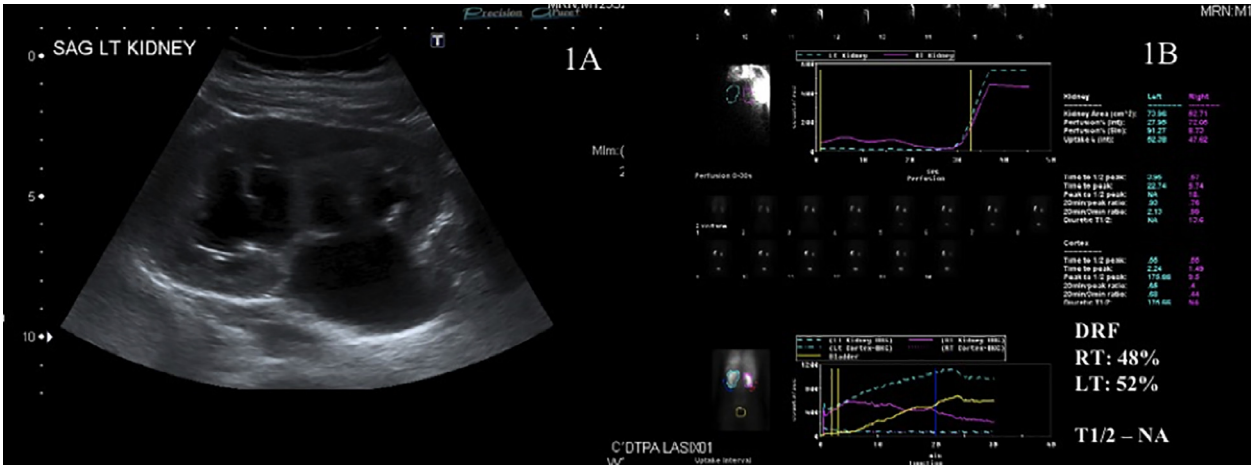

Figure 1. (**A**) US demonstrated significant worsening of left sided HN with an APD of 32 mm, (**B**) renal scan suggestive of UPJO. APD, anteroposterior diameter; HN, hydronephrosis; UPJO, ureteropelvic junction obstruction; US, ultrasonography. (Color version available online.)

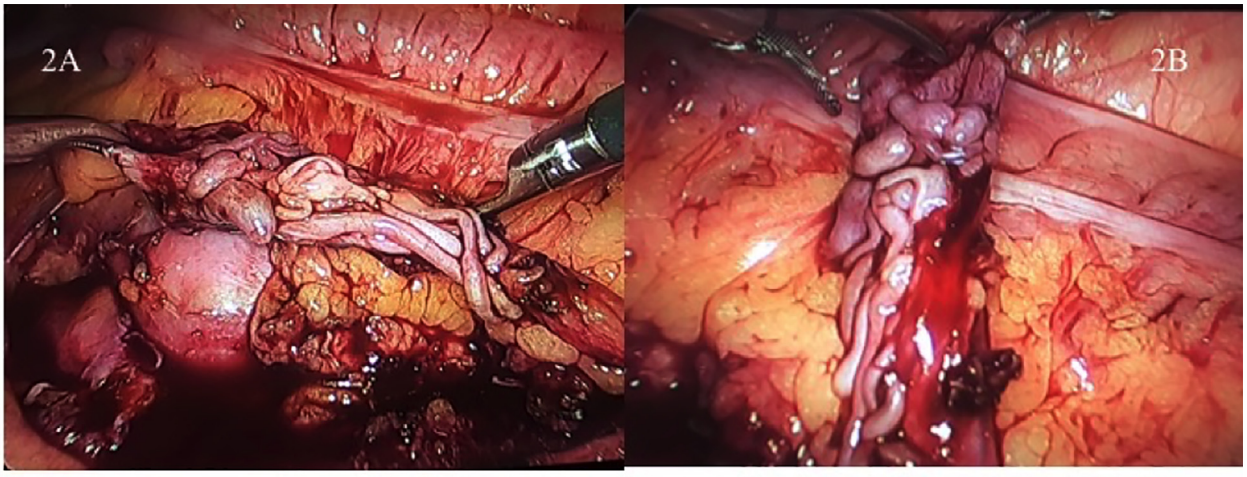

Figure 2. (**A**, **B**) Intra-operatively, multiple long intraluminal FEPs seen at the UPJ and extending into the proximal ureter for 3 to 4 cm. FEPs, fibroepithelial polyps. (Color version available online.)

flank pain, of which 8 (14%) were found to have FEPs and 50 (86%) were secondary to crossing vessels. Diagnosing FEPs preoperatively may be challenging, as they cannot be detected using standard diagnostic tools such as US. When suspecting of FEPs, computerized tomography scan or magnetic resonance imaging may help with the diagnosis, but the respective issues with radiation exposure and general anesthesia (depending on the child's age) limit their routine applicability in the pediatric population.[1] Therefore, retrograde pyelogram can be used at the start of the procedure to confirm obstruction and better delineate the anatomy of the UPJ but may not always confirm FEPs. An underutilized less invasive alternatively is non-radiating, magnetic resonance urography (MRU) which may provide similar results as retrograde pyeograms while removing radiation exposure. Currently, there is no consensus on the best imaging modality for FEP diagnosis.[5]

FEPs usually present as the sole cause for UPJO in children. In rare occasions though, they can be found simultaneously with a crossing vessel. [6] Such circumstances pose a challenge for treatment, as lower pole crossing vessels are deemed to be the sole etiology of the obstruction. As a result, some authors have advocated for the Hellstrom angiopexy or vascular hitch procedure as a simpler alternative to dismembered pyeloplasty in such cases.[7] However, persistent obstruction after angiopexy requiring redo dismembered pyeloplasty probably occurs because of failure to recognize an intrinsic factor such as a FEP at the time of the initial procedure, as described by Carter Ramirez et al.[6] In all cases of crossing vessels, meticulous inspection of the UPJ with clinical suspicion for potentially concomitant intrinsic obstruction is advisable. Our patient presented with intermittent Dietl's crisis and a delayed urinary drainage on renal scan, suggestive of UPJO due to lower pole

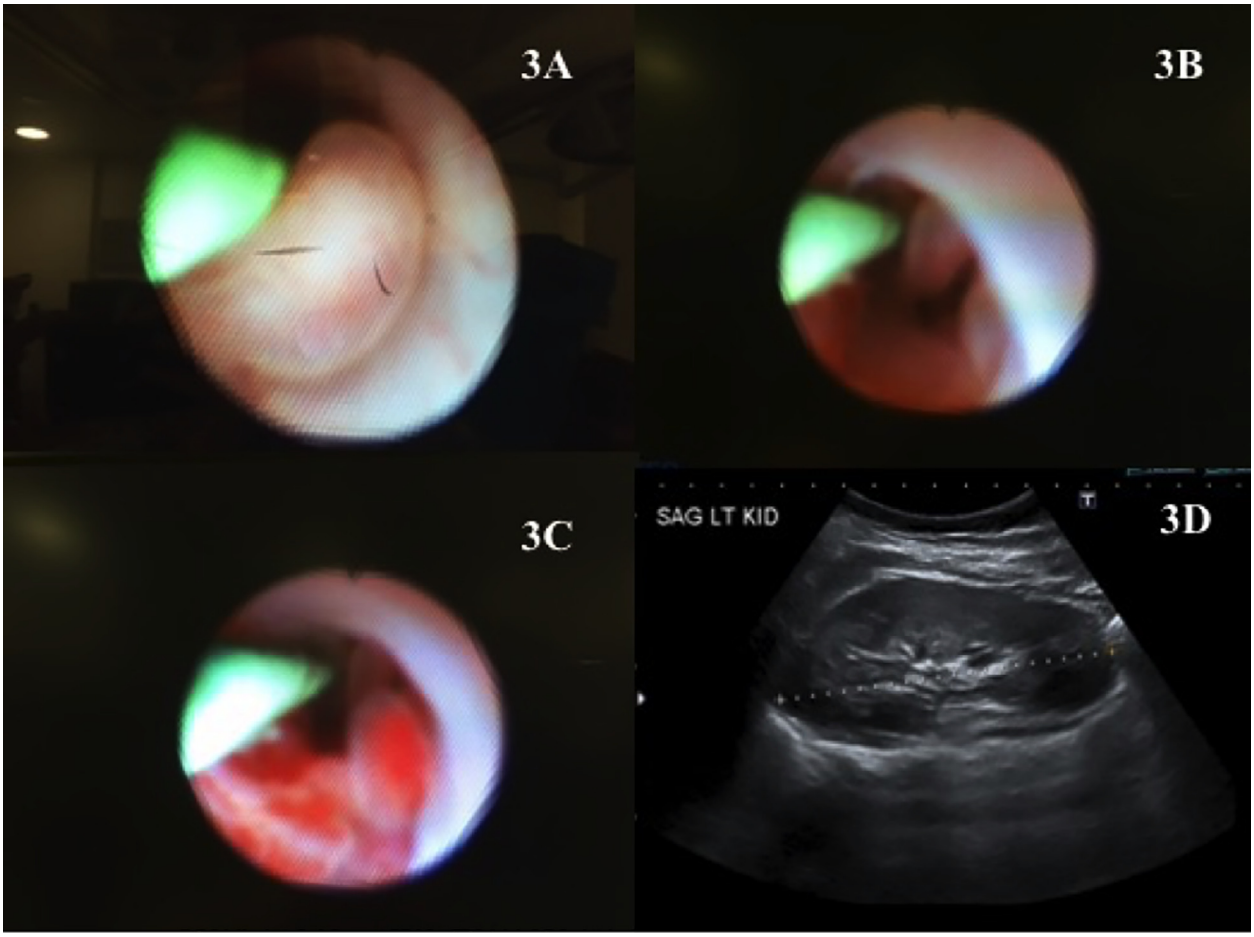

Figure 3. (**A-C**) Flexible ureteroscopy showing a few small polyps and a large pedunculated one with a thin stalk in the mid ureter. (**D**) Follow-up US showing substantial improvement of the left side HN and no ureteral dilatation. (Color version available online.)

crossing vessels. However, to our surprise, at the time of the repair, no crossing vessels were identified. To rely only on ureteral peristalsis intraoperatively to rule out intrinsic obstruction and confirm a patent UPJ may have its flaws, as seen in the supplementary video (Appendix 1). The key factor in diagnosing the intraluminal FEPs was the decision to perform a dismembered pyeloplasty, as no extrinsic source of ureteral narrowing was identified.

The extensive spatulation of the ureter revealed the large burden of polyps, extending from the UPJ into the proximal ureter for 3-4 cm. Special attention should be paid when attempting dismembered pyeloplasty in cases of FEPs, as they may extend through a long segment of ureter, requiring complete excision of that part, creating tension at the ureteropelvic anastomosis. In the present case, renal descensus and extensive distal mobilization of the ureter allowed for a tension-free suture line. At the time of stent removal, flexible ureteroscopy was key in identifying more polyps in the mid ureter that had been missed intra-operatively. Ablation of those polyps with holmium laser was convenient and very successful, as shown by others.[4] Although endoscopic ablation with holmium laser has been used as first line therapy for FEPs in the past,[2] this approach may not accomplish complete resection of those lesions, as the base of some large polyps cannot be completely excised by this method.[8-10]

This report represents a unique case given the size, the number and location of the FEPs, as their elongated and pedunculated appearance resembled the Greek mythology Medusa's hair. Our minimally invasive approach combining initial laparoscopic dismembered pyeloplasty that ruled out crossing vessels and confirmed the intrinsic nature of the UPJO, with subsequent laser ureteroscopy was key to completely remove all FEPs and relieve the obstruction. Based on that, we recommend laparoscopic/robotic-assisted pyeloplasty as the first line treatment for FEPs in children, especially in those with multiple and large polyps, in whom ureteroscopy alone may not be feasible or too difficult to remove all the lesions.[11]

CONCLUSION

Due to the diagnostic challenges posed by FEPs, older children with UPJO presenting with intermittent

abdominal or flank pain should not be assumed to have crossing vessels as the etiology of their obstruction, thus warranting clinicians to be cautious and rule out rare intrinsic causes for UPJO, such as FEPs. In the setting of numerous FEPs involving a long ureteral segment, Anderson-Hynes dismembered pyeloplasty with subsequent ureteroscopy and laser ablation of the remaining polyps at the time of stent removal (if necessary) is a safe, effective and minimally invasive approach to address this challenging condition.

SUPPLEMENTARY MATERIALS

Supplementary material associated with this article can be found in the online version at https://doi.org/10.1016/j.urology.2019.04.028.

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