

Pediatrics

Benign Fibroepithelial Polyps: A Rare Cause of Ureteropelvic Junction Obstruction in Children



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ABSTRACT

FEPs are rare, benign mucosal growths that may cause urinary tract obstruction in both adults and children. We present the case of a ten year old Hispanic male with recurring urinary tract infections and hydronephrosis diagnosed with fibroepithelial polyps (FEPs). Despite multiple radiographic procedures, we were unable to accurately preoperatively diagnose FEPs. Here we demonstrate the difficulties in preoperative diagnosis and suggest that perhaps a combination of US and MRI in the setting of persistent urinary tract infections and flank pain may be the best approach for early diagnosis and conservative management, including less invasive treatment protocols.

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Introduction

Fibroepithelial polyps (FEPs) are benign tumors of mesodermal origin located in the posterior urethra, the ureteropelvic junction or upper ureter. They are responsible for 0.5% of UPJ obstructions requiring pyeloplasty.¹ Although, the exact etiology of FEPs is unknown, chronic irritation or infection, developmental or allergic factors, trauma, or congenital causes have been implicated.^{1,2} Male and left side preponderance is noted in children 6 weeks to 12 year old.¹ Patient's usually present with hematuria and/or intermittent flank pain from obstructive hydronephrosis.^{1–3} In rare cases, torsion of the polyp may cause severe ischemic pain.³ Given that FEP's are more common in adults, contrast studies to identify FEP's are often not conducted in children. Despite excellent prognosis, failure to visualize, even with Doppler renal ultrasounds, causes FEPs to frequently go unrecognized.^{3,4} Pre-operative diagnosis will allow for less invasive surgical management.⁴ In children, lack of recurrence for up to 15 years is noted with surgical excision.⁴ Here we

describe a case of a 10 year old male with a longstanding history of urinary tract infections found to have a FEP intra-operatively.

Case presentation

A 10 year old male presented with a 4 year history of fever, nausea, vomiting, pain in the right flank with occasional hematuria every 1–2 months and 4 episodes of renal colic. A single instance of a 2 mm right distal ureteral stone associated with severe right hydronephrosis was noted on ultrasound at an outside hospital with resolution a few days later. No stone was collected and the patient continued with the aforementioned symptoms.

Upon presentation, renal ultrasound indicated right hydro-ureteronephrosis in the proximal ureters and an intraluminal ureter lesion (Fig. 1A). An obstructing ureteral calculus was initially suspected, so abdominal/pelvic CT scan was done and demonstrated right hydroureter with evidence of lower pole crossing vessels. The patient was diagnosed with a crossing vessels mediated right UPJ obstruction. Laparoscopic mobilization of the lower pole crossing vessels was recommended but not pursued by the family due to their immigration status.

The patient followed up after 7 months with increased frequency of pain, hematuria, nausea, vomiting and fevers. CT angiogram once again suggested crossing vessels, possibly from the celiac or superior mesenteric arteries (Fig. 1B and C). A MAG3

Abbreviations: FEP, Fibroepithelial polyps; UPJ, ureteropelvic junction; MAG3 scan, Mercurioacetyltriglycine 3 scan.

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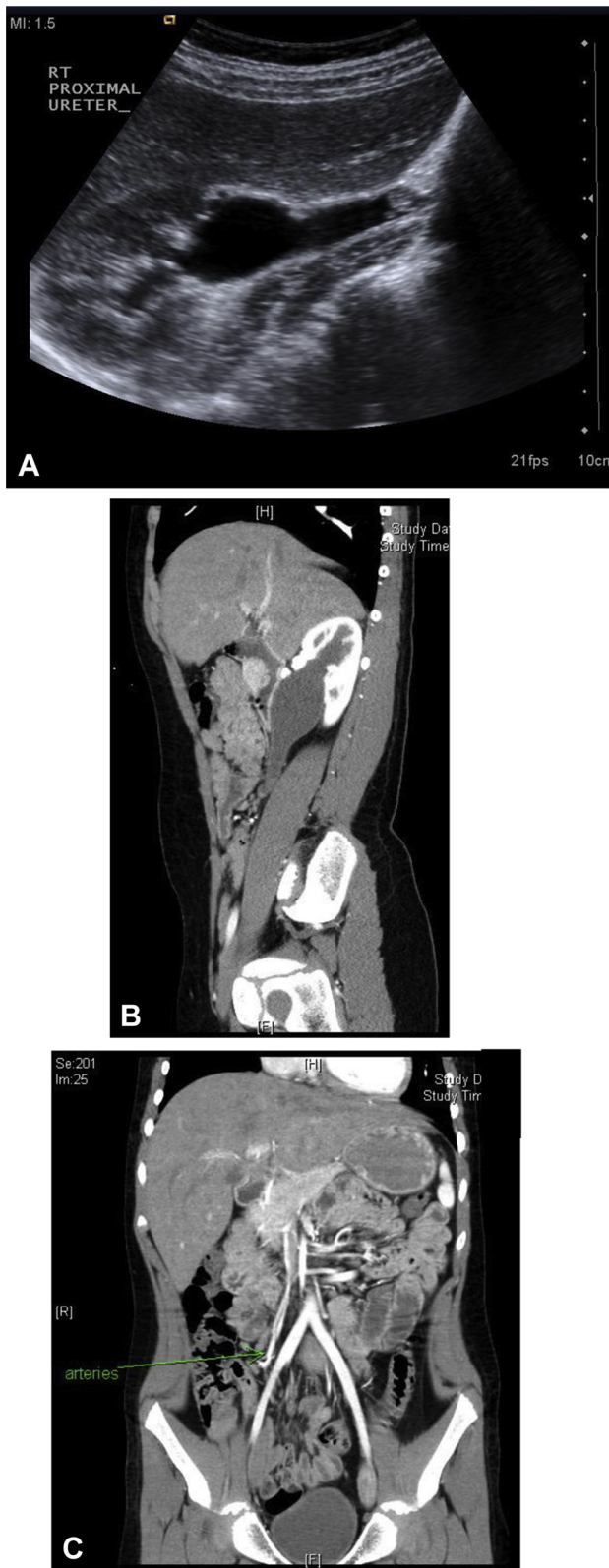


Figure 1. US & CT images of FEP: (A) Sonographic image demonstrating the FEP. (B) Sagittal CT angiogram demonstrating right pelvic fullness with proximal ureteral dilatation at the level of L5. (C) Coronal CT angiogram demonstrating crossing vessels from the celiac and SMA noted at this level. Coronal image through the renal pelvis shows mild hydronephrosis with dilatation of the renal pelvis and proximal ureter down to the non-shadowing irregular ureteral filling defect (FEP) that measures 0.54 cm in diameter and is indicated by the arrow. The right kidney measures $9.50 \times 3.60 \times 3.17$ cm.

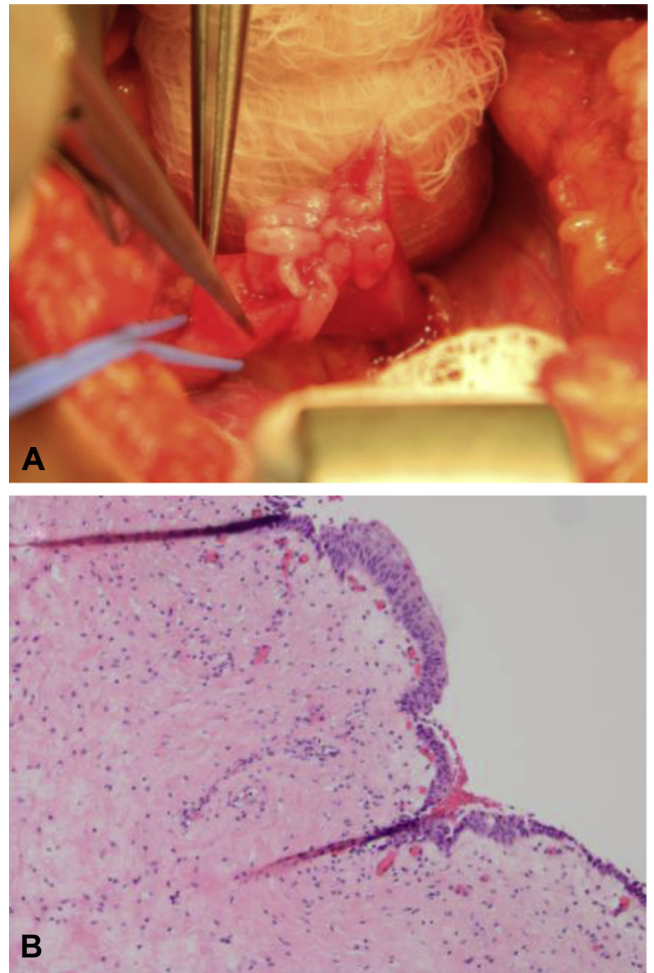


Figure 2. Gross and Microscopic specimens of the FEP: (A): Intra-operative image of smooth, white surfaced irregular shaped polyp with frond like tissue, 2.5×2.0 cm, involving the lumen of the right proximal ureter. (B) High power view of section from the tissue shows a polypoid structure lined by benign transitional epithelium. The underlying stroma is striking with marked edema and splaying of space between the underlying muscle. No areas of mineralization or polarizable foreign material are noted within the polyp.

nuclear renal scan with lasix demonstrated 51% of total right kidney function. An obstructive drainage pattern with the T1/2 Lasix drainage time never being reached was noted for the right kidney. A right pyeloplasty with possible transposing vessels was scheduled.

Operative report

The retroperitoneum was approached via a right flank incision and the right renal pelvis and ureters were exposed. The right UPJ had severe angulation. A tagging suture was placed at the distal renal pelvis/upper ureter and a transverse ureter incision revealed an irregularly shaped 2.5×2.0 cm polyp circumferentially involving the lumen of the ureter (Fig. 2A). The polyp had irregular frond like tissue filling the lumen of the ureter. Pathology confirmed a polypoid FEP lined by benign transitional epithelium with marked edema of the underlying stroma (Fig. 2B). Surgery proceeded with right ureteroureterostomy and stent placement.

Postoperative course

The patient was discharged on day 3 with double J stent removal 1 month postoperatively. The 3 month postoperative MAG 3

nuclear renal scan and renal ultrasound indicated mild left and right hydronephrosis. No obstruction was found. He continues to do well.

Conclusion

Diagnosis of FEPs presents a unique challenge due to their variable size, shape and the inability to visualize the UPJ area with echography, urography, or retrograde pyelography.³ Delayed surgical intervention may adversely affect renal function, causes pain, fever, nausea, vomiting and persistent urinary tract infections. In this case, traditional imaging modalities of renal Doppler ultrasound, CT scan and MAG 3 nuclear renal scan with Lasix failed to preoperative diagnose FEP.

Historically, FEPs are diagnosed with ultrasound, CT scans, nuclear medicine renal scans, and retrograde urograms.¹ Although not usually noted, FEPs on ultrasound may present as solid vascular formations with no posterior acoustic shadowing with or without mild hydronephrosis.¹ CT scans may also reveal hydronephrosis with a soft tissue obstruction within the ureter. In our patient, neither an US or CT scan was successful in an FEP diagnosis. Excretory urograms are generally more successful in diagnosing FEP's, where they appear as long, smooth ureteral filling defects, associated with varying degrees of hydronephrosis (if any).³ Unfortunately, their position may change between urograms resulting in the misdiagnosis of a non-opaque ureteral calculus, blood clot, or obstruction due to inflammatory process or tumor.² Further, the close proximity of FEPs to the upper ureter renders visualization difficult with misdiagnosis for more common obstructions such as congenital narrowing of the ureter or crossing lower pole vessels.¹

Retrograde pyelograms are currently also recommended for FEPs given they provide better visualization of the proximal and distal ureters.⁵ However, retrograde pyelography is invasive, may require anesthesia and exposes the patient to radiation. Further, as noted in our case, retrograde pyelogram may still fail to diagnose an FEP.

An often underutilized modality in children is the less invasive, non-radiating, magnetic resonance urography (MRU). MRU's include a heavily T2 weighted sectional images in orthogonal planes, which allows clear distinction between fluid (bright signal) and tissue (dark signal).⁴ Further, MRU's may also be considered given they combine the benefits of a retrograde pyelogram along with MRU. Given FEP are non-malignant in children, in contrast to adults, endoscopic or laparoscopic resection can be performed.² Segmental resection of the ureter or pelvis, open dismembered pyeloplasty, or nephroureterectomy should only be considered, if malignancy is suspected.³

Currently, there is no consensus on the best imaging modality for FEP diagnosis. An MRU may be used given it provides similar results as retrograde pyelograms while being less invasive and

avoiding radiation. Although US are not ideal for FEP visualization, it is the appropriate first step in the work up of renal colic and urinary tract infection. Upon successful preoperative diagnosis, a pediatric patient can undergo laparoscopic or endoscopic FEP resection. Based on this case, we believe a combination of US and MRU in a setting of persistent urinary tract infections and flank pain may allow for preoperative diagnosis of FEP allowing for less invasive surgical management of children.

Conflict of interest

The authors have no conflict of interest to report.

Consent

Written informed consent was obtained from the patient for publication of this Case report and any accompanying images. A copy of the written consent is available for review by the Editor of this journal.

Authors' contributions

Dr. Anitha Ezekiel was essential in data extraction, interpretation, compilation and drafting of the manuscript. Dr. Vaidehi Agrawal was essential in data interpretation, compilation, drafting and final approval of the manuscript. Dr. Elena Romero is a radiologist who was essential in data interpretation, compilation, drafting and final approval of the manuscript. Dr. Leon I. Smith-Harrison was the primary urologist who encountered this unique case and provided valuable oversight to the production of this case report. He was essential in drafting and final approvals of the final version as presented above.

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