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Case Report

Investigating the Abnormal Presentation of Ureteropelvic Junction Obstruction in Adolescence: A Case Report

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ABSTRACT

Background and Aim: Ureteropelvic junction obstruction (UPJO) is a common cause of hydronephrosis and is typically diagnosed antenatally. About 70% of these cases will self-resolve; however, symptomatic children may present with abdominal pain (i.e. Dietl crisis), vomiting, rash, or fever.

Case Presentation: A 14-year-old male patient presented with cyclical vomiting every two months for the past two years. The patient's mother also described an eight-year history of headaches and intermittent abdominal pain. A renal magnetic resonance urography scan revealed decreased right kidney function and delayed cortex to ureter transit time. A pyeloplasty and stent placement were performed to correct the obstructing vessel.

Conclusion: Clinical guidelines for abdominal pain and cyclical vomiting earlier in the patients' healthcare may have led to an appropriate workup and treatment years before. We recommend physicians consider UPJO as a differential diagnosis in adolescent patients with cyclical vomiting and abdominal pain and consult the North American Society for Pediatric Gastroenterology, Hepatology, and Nutrition (NASPGHN) guidelines to better guide the diagnosis.

Keywords: Hydronephrosis, Cyclical vomiting, UPJO, Dietl crisis

Introduction

Ureteropelvic junction obstruction (UPJO) is a relatively common cause of hydronephrosis, occurring in 0.1% of newborns and more often in males than females [1-3]. Since the advent of routine prenatal ultrasonography in the 1970s, UPJO is almost exclusively diagnosed antenatally [4, 5], and up to 70% of these cases will self-resolve or are monitored and surgically corrected as early as possible [5]. In symptomatic cases, children will present with abdominal pain (i.e. Dietl crisis), vomiting, rash, or fever [6-8]. If undiagnosed and untreated, UPJO can increase the risk of urinary tract infections and lead to the development of nephrolithiasis, hydronephrosis, thinning of the renal parenchyma, and chronic kidney disease [5].

If presenting with typical symptoms, such as abdominal or flank pain, the first investigative study typically leading to a diagnosis of UPJO is ultrasonography. In chronic cases, this will often reveal hydronephrosis. From here, magnetic resonance urography (MAG3) can be used to diagnose UPJO, assess renal function, and determine the cause of the obstruction and secondary hydronephrosis [9, 10].

However, UPJO may not initially present with flank pain making it more challenging to diagnose. In 2002, Schulte-Bockholt et al. reported on four pediatric cases of UPJO and secondary hydronephrosis initially presenting as cyclical vomiting syndrome [8]. Because many conditions can cause vomiting in pediatric patients, when vomiting is the chief complaint, establishing a diagnosis of UPJO can be difficult. However, recurrent vomiting does often indicate an underlying pathology requiring further work-up. According to North American Society for Pediatric Gastroenterology, Hepatology and Nutrition guidelines (NASPGHN), the initial workup of cyclic vomiting syndrome should include an evaluation of electrolytes, blood urea nitrogen, creatinine, glucose, and an upper-gastrointestinal imaging series with an ultrasound conducted at any time for patients with abdominal pain [1].

These work-up processes can frequently lead to unexpected findings and potential underlying diagnoses, such as UPJO; therefore, physicians must maintain broad differentials to capture possible diagnostic zebras. In this case report, we describe a case of a 14-year-old male with an eight-year history of headaches, cyclical vomiting, and abdominal pain later diagnosed as UPJO.

The article provides a comprehensive overview of UPJO as a significant cause of hydronephrosis in pediatric patients. It emphasizes the importance of routine prenatal ultrasonography in the early diagnosis of UPJO, outlines typical symptoms, discusses diagnostic challenges, and describes the diagnostic process, culminating in a case study of a 14-year-old male.

Case Presentation

The patient, a 14-year-old male, presented to the National University of Natural Medicine Health Center in January 2023 complaining of migraines with photophobia and recurrent vomiting occurring every 2 months for the past two years. These episodes typically lasted for 7-10 h with vomiting once per hour. The patient's mother described an 8-year history of headaches and intermittent abdominal pain. The patient's past medical history also included asthma, recurrent erythematous rashes over the chest and left shoulder, and temperature sensitivity since 6 years old.

At the initial presentation, there was suspicion of cyclical vomiting syndrome, therefore a complete laboratory workup with imaging was done in adherence with the NASPGHAN guidelines [1] for cyclical vomiting in children: Complete blood count, comprehensive metabolic panel, gamma-glutamyl transferase, uric acid, lactate dehydrogenase, ferritin, amylase, lipase, complete urinalysis, an abdominal ultrasound, and an upper gastrointestinal series. At a lifetime prevalence of 1.9%, cyclical vomiting syndrome is one of the most common causes of recurrent episodes of vomiting in children [11], and involves at least five vomiting attacks over any interval or 3 attacks over 6 months with a return to baseline health between attacks. Due to this patient's presentation, cyclical vomiting syndrome needed to be evaluated and ruled out.

The patient's initial laboratory workup revealed elevated uric acid, potassium, and creatinine with proteinuria found on urinalysis (8.6 mg/dL, 5.2 meq/L, and 1.27 mg/dL, respectively); estimated glomerular filtration rate was also decreased at this time (53 mL/min), and the patient's elevated blood urea nitrogen level indicated azotemia (20 mg/dL) (Table 1). Subsequent abdominal ultrasound in March 2023 revealed a notable right-sided dilation of the right renal collecting system and severe thinning of the overlying parenchymal mantle. The appearance suggested severe chronic hydronephrosis with renal atrophic changes and the possible presence of vesicoureteral reflux. The upper gastrointestinal series revealed no abnormal findings. Immediately, the patient was referred to pediatric nephrology at Oregon Health

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and Science University Doernbecher Children's Hospital for a renal MAG3 scan.

In June 2023, the patient's MAG3 revealed the differential function of the right kidney to be 27% and cortex to ureter transit time of 18 min while the left kidney maintained a healthy differential function of 73% and an 8 min cortex to ureter transit time. Hydronephrosis and parenchymal thinning were noted in the right kidney due to an inferior pole crossing artery, a finding present in roughly 50% of symptomatic UPJO cases requiring surgical correction [12]. Follow-up laboratory testing at this time revealed hyperparathyroidism (Table 1) likely due to chronic kidney disease and the patient was started on calcitriol (0.25 mcg 3×/week).

In September 2023, a pyeloplasty was performed to correct the obstructing vessel, and a stent was placed at this time. There were no surgical complications. A follow-up cystoscopy was performed in October 2023 to remove the ureteral stent and a follow-up ultrasound in November 2023 revealed decreased hydronephrosis as compared to pre-operative findings: 24 mm pre-surgical pelvis diameter vs 6 mm post-surgical pelvis diameter (Figure 1).

Following the patient's pyeloplasty, intermittent follow-up visits and ultrasounds will be necessary to ensure functional improvement of the kidneys and the absence of proteinuria and hypertension, complications occurring in roughly 15% of pyeloplasty cases [13].

Treatment and clinical outcomes

Perspective of the patient and the patient's guardian

A few months following the pyeloplasty, the patient noted markedly reduced abdominal pain, headache frequency, and episodes of recurrent vomiting.

"I have been doing rather good since the surgery and the lack of kidney pain and cyclic vomiting has been incredible. These factors having been eliminated have allowed me to start doing more physical activities and I have noticed a significant increase in my physical ability after the surgery and after taking the caltriciol for ~6 months. Overall, I have noticed a significant decline in my chronic headaches, vomiting/nausea episodes, and kidney pain in my right side."

The patient described that the decrease in symptoms, following years of unrelenting symptomatology, has felt freeing and relieving after having spent much of his youth compromised by a now-revealed urological condition that contributed to systemic symptoms.

In addition, the patient expressed a high level of motivation to return to school as soon as possible.

The patient's mother was the main caretaker during the many years of unexplained abdominal pain and concomitant symptoms. Throughout the months following thorough workup, and eventual diagnosis, she recounted the challenges faced as a family in navigating the healthcare system and seeking answers to the perplexing symptoms her son exhibited. The patient's mother also highlighted the emotional roller coaster she experienced throughout her son's care, from the initial uncertainty and anxiety surrounding the diagnosis to the eventual relief experienced after the surgical intervention. Her narrative brings to light the transformative impact of the surgery on her son's health, offering a testament to the significance of medical interventions in improving the quality of life for individuals grappling with UPJO. Her perspective serves as a valuable addition, providing a human touch to the clinical understanding of the patient's journey. In reflection on the healthcare journey after reading this manuscript, the patient's mother told us "I really hope this helps others somewhere along the line access (or provide) the right care needed in a much timelier fashion. If there is any blessing to what a mess of an experience this had been, it would be that alone."

Overall, the patient experienced significant benefits from the surgery, and their mother, the patient's primary caretaker, expressed relief about concern for her son and the transformational impact the treatment had on their lives.

Discussion

The Dietl crisis (i.e. severe abdominal pain pathognomonic for UPJO and hydronephrosis) is an alarming presentation that should be met with immediate diagnostic imaging [14]. However, when presenting with an alternative chief complaint, the correct workup may be delayed. Hydronephrosis has previously been reported to mimic cyclical vomiting syndrome [8], but this case was particularly abnormal as it is rare that adolescents present with undiagnosed and untreated UPJO. Although the relationship between vomiting, migraines [15], and chronic kidney disease/hydronephrosis has been previously described in the literature, the relationships between these are poorly established enough that they are

Table 1. Laboratory results summary

Lab (Reference Range)	1/17/2023	5/26/2023	6/15/2023	09/07/2023	11/22/2023	01/05/2024
Uric acid (4.4-7.6 mg/dL)	8.6 (H)					
CMP - potassium (3.4-5.0 meq/L)	5.2 (H)	4.6	4.8	4.3		4.9
Phosphorus (2.4-4.7 mg/dL)	5.4 (H)	5.4 (H)	5 (H)	4.9 (H)		4.3
CMP - creatinine (0.40-1.05 mg/dL)	1.27 (H)	1.2 (H)	1.3 (H)	1.36 (H)		1.4 (H)
CMP - BUN (6-20 mg/dL)	20	23 (H)	25 (H)	22 (H)		18
Globulin level (2.3-3.5 g/dL)			2.2 (L)			2 (L)
eGFR (bedside Schwartz)	53					
Protein concentration urine (<20 mg/dL)			296 (H)	82 (H) (6/19/23)		306 (H)
Creatinine concentration urine (mg/dL)			148.1	85.6 (6/19/23)		96.41
Protein/creatinine ratio urine (<0.10 mg/mg)			2 (H)	0.96 (H) (6/19/23)		3.179 (H)
Cystatin C (0.5-1.2 mg/L)			1.6 (H)			
UA, complete (protein – negative)	3+ (H)					
UA, dip - protein (neg-trace mg/dL)			≥300(H)	≥300 (H)	100 (H)	200 (H)
UA, dip - blood (negative)				Trace-intact	Negative	Negative
Urine culture (no growth)	No growth			No growth after 48 h	No growth after 24 h	
Parathyroid hormone, intact (12-88 pg/mL)			219 (H)	81		57.3
Calcium (8.5-10.3 mg/dL)						9.7
Vitamin B-12 (232-1,245 pg/mL)						279
Folate (>4.5 ng/mL)						13.3
Lipase (11-82 U/L)	23					
Amylase (29-103 U/L)	81					
% Iron saturation (20-55%)			35	27		32.3
Ferritin (14-80 ng/mL)	54.5		14	14		20.6
Iron (50-170 ug/dL)			152	129		151.16
Total iron binding capacity (250-400 ug/dL)			440 (H)	472 (H)		468 (H)
Transferrin (200-400 mg/dL)			314	337		334.25
Magnesium, plasma (1.6-2.6 mg/dL)			1.7			
CBC – white blood cells (4.90-15.5 K/cu mm)	6.6		4.81 (L)	5.94		4.4 (L)
Hemoglobin (13.0-16.0 g/dL)	13.7		13	13.7		14.2
CBC – neutrophil (2.80-11.1 K/cu mm)	n/a		2.49 (L)	3.4		n/a
CBC – monocyte (0.30-1.30 K/cu mm)	0.4		0.28 (L)	0.41		n/a

Abbreviations: UA: Urinalysis; CBC: Complete blood count; CMP: Complete metabolic panel; BUN: Blood urea nitrogen; eGFR: Estimated glomerular filtration rate; N/a: Not applicable H: High; L: Low.

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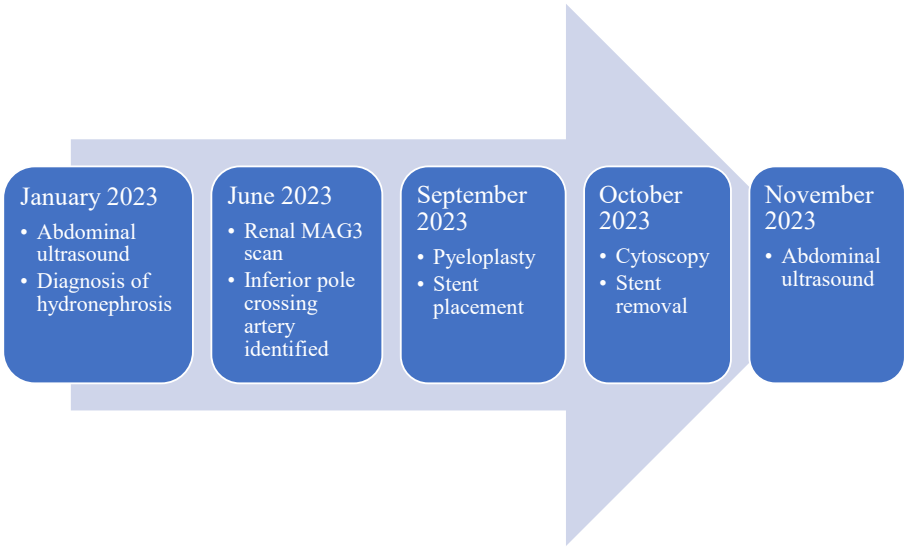

Figure 1. Timeline of medically appropriate imaging and associated findings

MAG3: Magnetic resonance urography.

seldom or never mentioned on medical databases, such as UpToDate as of this publication [16, 17].

It is common for physicians to shy away from severe or complex diagnoses for fear of being wrong, but this phenomenon, known as "zebra retreat", prolongs the time for patients with complex symptom pictures to receive accurate diagnoses and associated treatments [18, 19]. To combat zebra retreat, physicians should practice generating a broad list of differential diagnoses before limiting their clinical decision-making to common conditions simply based on prevalence. In addition, clinicians who stay up-to-date on medical literature relevant to their patients will be less likely to miss uncommon or newly described medical phenomena. This case is a reminder that the possibility of diagnostic zebras, should not preclude physicians from performing an appropriate and thorough work-up. In this case, the use of clinical guidelines, NASPGHN in particular, meant to provide clinicians with appropriate strategies for work-up led to a diagnosis and associated treatment that resulted in effective care for the patient and resolution of symptoms. However, as this was an atypical presentation of UPJO, it may be beneficial for this, or similar, symptom clusters to appear in other pediatric clinical guidelines.

Conclusion

The use of clinical guidelines for abdominal pain and cyclical vomiting earlier in the patients' healthcare process could have led to an appropriate workup and treatment many years before the patient's presentation to our clinic in 2023. We recommend that doctors consider UPJO and hydronephrosis as differential diagnoses in pediatric patients with recurrent vomiting and/or abdominal pain and consult the NASPGHN guidelines to guide diagnosis. Future research should explore the prevalence and mechanisms for atypical presentations of UPJO for the adolescent population to aid expedient treatment.

Ethical Considerations

Compliance with ethical guidelines

There were no ethical considerations to be considered in this research.

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Authors' contributions

All authors equally contribute to preparing all parts of the research.

Conflict of interest

The authors declared no conflict of interest.

References

[1] Li BU, Lefevre F, Chelimsky GG, Boles RG, Nelson SP, Lewis DW, et al. North American Society for Pediatric Gastroenterology, Hepatology, and Nutrition consensus statement on the diagnosis and management of cyclic vomiting syndrome. J Pediatr Gastroenterol Nutr. 2008; 47(3):379-93. [DOI:10.1097/MPG.0b013e318173ed39] [PMID]

[2] Duong HP, Piepsz A, Collier F, Khelif K, Christophe C, Cassart M, et al. Predicting the clinical outcome of antenatally detected unilateral pelviureteric junction stenosis. Urology. 2013; 82(3):691-6. [DOI:10.1016/j.urology.2013.03.041] [PMID]

[3] Chang CP, McDill BW, Neilson JR, Joist HE, Epstein JA, Crabtree GR, et al. Calcineurin is required in urinary tract mesenchyme for the development of the pyeloureteral peristaltic machinery. J Clin Invest. 2004; 113(7):1051-8. [DOI:10.1172/JCI200420049] [PMID]

[4] Bilge I. Symptomatology and clinic of hydronephrosis associated with uretero pelvic junction anomalies. Front Pediatr. 2020; 8:520. [DOI:10.3389/fped.2020.00520] [PMID]

[5] Passoni NM, Peters CA. Managing ureteropelvic junction obstruction in the young infant. Front Pediatr. 2020; 8:242. [DOI:10.3389/fped.2020.00242] [PMID]

[6] Burhop J, Clingenpeel JM, Poirier MP. Emergency department diagnosis of dietl crisis in a 7-year-old girl with abdominal pain. Pediatr Emerg Care. 2016; 32(6):386-7. [DOI:10.1097/PEC.0000000000000339] [PMID]

[7] Maddileti V, Gazula S, Dantala P, Srinivas M. Dietl's crisis: An under appreciated clinical entity in the paediatric population. BMJ Case Rep. 2021; 14(8):e244943. [DOI:10.1136/bcr-2021-244943] [PMID]

[8] Schulte-Bockholt A, Kugathasan S, Mesrobian HGO, Werlin SL. Ureteropelvic junction obstruction: An overlooked cause of cyclic vomiting. Am J Gastroenterol. 2002; 97(4):1043-45. [DOI:10.1111/j.1572-0241.2002.05626.x] [PMID]

[9] Wood LN, Souders CE, Freedman AL. Is a reassuring MAG-3 Diuretic renal scan really reassuring? Curr Urol. 2015; 8(4):178-82. [DOI:10.1159/000365713] [PMID]

[10] Ucar AK, Kurugoglu S. Urinary ultrasound and other imaging for Ureteropelvic Junction Type Hydronephrosis (UPJHN). Front Pediatr. 2020; 8:546. [DOI:10.3389/fped.2020.00546] [PMID]

[11] Raucci U, Borrelli O, Di Nardo G, Tambucci R, Pavone P, Salvatore S, et al. Cyclic vomiting syndrome in children. Front Neurol. 2020; 11:583425. [DOI:10.3389/fneur.2020.583425] [PMID]

[12] Al Aaraj MS, Badreldin AM. Ureteropelvic junction obstruction. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2024. [Link]

[13] Oktar T, Selvi I, Dönmez Mİ, Alan Y, Değirmenci E, Ziylan O. What to expect on the long-term follow-up of pediatric pyeloplasty: Critical time intervals and risk factors. J Pediatr Surg. 2024; 59(6):1170-6. [DOI:10.1016/j.jpedsurg.2023.11.021] [PMID]

[14] Lahoud RM, Esker WD, Thurston SA, Kohanski M, Elder JS, Lim R. Dietl crisis: Presentation and imaging findings in a 7-year-old boy. Radiol Case Rep. 2020; 16(3):555-9. [DOI:10.1016/j.radcr.2020.12.048] [PMID]

[15] Hené IZ, Goldschmeding R, Hené RJ. Headache and a syndrome of bilateral hydronephrosis. Nephrol Dial Transplant. 2002; 17(8):1527-9. [DOI:10.1093/ndt/17.8.1527] [PMID]

[16] Warady BA, Weidemann DK. Chronic kidney disease in children: Definition, epidemiology, etiology, and course. UpToDate. 2023. [Link]

[17] Desilets DJ. Cyclic vomiting syndrome. UpToDate. 2023. [Link]

[18] Miller CS. Skin-deep diagnosis: Affective bias and zebra retreat complicating the diagnosis of systemic sclerosis. Am J Med Sci. 2013; 345(1):53-56. [DOI:10.1097/MAJ.0b013e3182684aab] [PMID]

[19] Onder O, Yarasir Y, Azizova A, Durhan G, Onur MR, Ariyurek OM. Errors, discrepancies and underlying bias in radiology with case examples: A pictorial review. Insights Imaging. 2021; 12(1):51. [DOI:10.1186/s13244-021-00986-8] [PMID]