CASE REPORT

Antenatal mild hydronephrosis with subsequent polyp of the upper ureter in a child presenting with recurrent Dietl's crisis

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SUMMARY

We report a 7-year-old boy who had antenatally diagnosed and postnatally confirmed asymptomatic right congenital hydronephrosis secondary to ureteropelvic junction obstruction with the anteroposterior diameter ranging from 7 to 15 mm on serial ultrasound scans till the age of 6 years. He then presented with recurrent attacks of Dietl's crisis almost every month in the seventh year of his life and eluded diagnosis for over a year at various national hospitals, and ended up with an international referral to us. Investigations of the acute attacks clinched the diagnosis of ureteral polyp causing intermittent crisis and he underwent segmental resection and reconstruction in the form of dismembered Anderson-Hynes pyeloplasty with good recovery. Our case revealed that prenatally detected hydronephrosis may worsen after spontaneous postnatal improvement and a polyp acting as a flip valve may produce intermittent hydronephrosis and symptoms later in life. The child should undergo urgent investigations during acute symptoms.

BACKGROUND

Upper ureteral polyp is an unusual cause of hydronephrosis in children and a rare benign neoplasm of mesodermal origin.^{1 2} The association of the two lesions—antenatally diagnosed moderate hydronephrosis improving on conservative management later getting complicated by upper ureteral polyp in the same patient is an extreme rarity. Only two similar cases have been reported in the literature.^{1 2} We present our initial experience with upper ureteral polyp in the scenario of a background antenatally diagnosed hydronephrosis leading to delay in diagnosis and referral to an international centre.

CASE PRESENTATION

A 7-year-old boy was diagnosed with right-sided congenital hydronephrosis during pregnancy and was followed up with serial periodic postnatal scans at every 6 months period. He was asymptomatic up to the age 6 years with the anteroposterior (AP) diameter of the right renal pelvis fluctuating between 7 and 15 mm and equal renal function with prompt drainage on MAG3 scans.

He developed episodic recurrent acute right loin pain and vomiting similar to Dietl's crisis which happened 10 times in the past 12 months.

He was seen by several professionals and had several admissions and investigations electively when the pain had settled down and did not show any gross abnormality other than the background right congenital fluctuating hydronephrosis.

His parents wanted further investigations of his recurrent problem. The boy was advised against swimming lessons as his problem was getting worse after swimming. Health professionals labelled the patient's mother to be a hypochondriac bordering on Munchausen syndrome by proxy and the patient was thought to have psychological problems of abdominal migraine or cyclical vomiting rather than any organic cause.

INVESTIGATIONS

The parents decided to get an international expert second opinion as the boy was clinically asymptomatic. Ultrasound scan showed right renal pelvis dilation measuring 5–6 mm only. Colour Doppler scan showed no evidence of any crossing lower polar vessels (figure 1A,B). MAG3 scan confirmed both kidneys to be functioning well and equally. After administration of furosemide, an evidence of slower drainage on the right side with dilation of upper right ureter and suggestion of an irregular filling defect partially obstructing the right upper ureter was evident (figure 2).

During the next episode, an urgent ultrasound scan demonstrated an increase in the AP diameter of the right renal pelvis to 31–38 mm from 7 mm with dilation of the right upper ureter and a faint filling defect in the lumen of the upper ureter. A provisional diagnosis of right upper ureteric polyp causing intermittent acute symptoms was made and the family was given further options of investigations and interventions in the form of cystourethroscopy, retrograde studies, ureteroscopic endoscopic excision, laparoscopic or open excision and reconstruction. Owing to the international referral and private nature of the treatment, the parents elected an open quick procedure.

DIFFERENTIAL DIAGNOSIS

Intermittent hydronephrosis secondary to an aberrant vessel to the lower pole, a calculus intermittently blocking a partially obstructed pelviureteric junction and an upper ureteral polyp on the background of an antenatal mild hydronephrosis complicating the presentation and interpretation of the finding during asymptomatic state were considered.

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Figure 1 (A and B) Ultrasound scan and colour Doppler when asymptomatic and during acute attack—note upper ureteral dilation with acoustic shadow.

Figure 2 MAG3 scans showing equal function and relatively slow drainage—note right upper ureteral dilation with filling defect.

TREATMENT

At the right anterolateral extraperitoneal exploration, the right upper ureter and pelvis were dilated and a polyp could be palpated just above the change of calibre from the dilated upper ureter to the collapsed lower ureter. A segmental resection of the right upper ureter containing the polyp with reconstruction using dismembered Anderson-Hynes pyeloplasty and JJ stent insertion was carried out uneventfully.

OUTCOME AND FOLLOW-UP

The excised specimen showed an intraluminal polypoidal/valvular tissue in the upper ureter with a small stalk inferiorly leading to the collapsed distal ureter (figure 3A). Histopathological examination of the excised tissue showed polypoid tissue composed of oedematous, congested and focally inflamed submucosa covered by mildly reactive transitional epithelium. A diagnosis of fibroepithelial polyp was made (figure 3B).

The patient had excellent recovery and he went home the following day. JJ stent was removed after 4 weeks. At 6 months follow-up, he is asymptomatic, well and thriving with normal ultrasound scan and MAG3 scans.

DISCUSSION

Antenatal hydronephrosis with an AP diameter ranging from 7 to 15 mm is of mild-to-moderate severity, remains asymptomatic and generally does not require repeated hospital admissions, investigations, referral to an international centre and an operative correction. Benign tumours of the ureter are extremely rare in children.

Most of the mild-to-moderate prenatal hydronephroses resolve spontaneously or improve with conservative treatment. However, there is a late recurrence of prenatal hydronephrosis with intermittent symptoms sometimes after improvement. Our case is a reminder of the fact that if mild-to-moderate prenatally diagnosed hydronephrosis which was improving earlier has recurred with intermittent severe symptoms later in life, lower polar vessels, secondary stone or an ureteric polyp should be

considered in the differential diagnosis and should be investigated during the acute phase to establish the diagnosis.

Epidemiologically, they are more commonly found in boys (80.6%); often arise in the proximal ureter and renal pelvis and especially more common on the left side (69.4%). Multiple and bilateral cases have been reported.^{2 3} Fibroepithelial polyps are benign mucosal projections that can be found throughout the urinary system.^{4 5} The majority of patients were male (89%) and had obstruction on the left side (78%) as found in one study.⁶ Only a minority (22%) of patients had a diagnosis of obstructing polyps suggested preoperatively, so it is very easy to miss the diagnosis unless high index of suspicion and lateral thinking during change in the clinical picture are given due importance.

It causes partial intermittent ureteral obstruction usually without loss of renal function, so the usual laboratory tests are unlikely to help in the diagnosis. It is associated with several episodes of intermittent lumbar pain and haematuria; ureteric colic with vomiting and urinary tract infection are the common presenting features due to intermittent hydronephrosis, which develops extensively in the upper part of ureters.⁶ The presentation is very similar to recurrent Dietl's crisis seen in patients with lower polar vessels causing intermittent congenital hydronephrosis.

Ultrasonography of the kidney may show mild-to-moderate hydronephrosis when the patient is asymptomatic and well, while this finding is markedly aggravated in association with acute attack of pain and other symptoms.⁷ Colour Doppler helps in ruling out lower polar vessels as the cause of Dietl's crisis. MAG3 scan shows partial obstructive pattern and in acute attack would show an obstructive curve. MR angiography helps to rule out extrinsic lower polar vessels and MR urography shows calibre change and the associated filling defects, and it is the gold standard as reconstruction of images gives a better three-dimensional picture as well. Intravenous urography, cystoscopy, retrograde pyelogram, ureteroscopy, frozen section and biopsy of the lesion can all help in the diagnosis.^{8 9}

A variety of treatments ranging from simple observation, excision of the lesion, endoscopic local resection or laser ablation to

Figure 3 Excised specimen showing polypoidal growth in the upper ureter and microphotographs showing histological picture of fibroepithelial polyp.

more extensive procedures such as resection of ureter and pelvoureteral junction have been reported. Ureterorenoscopy is currently the best method available for the identification and histological diagnosis of ureteral polyps.¹⁰ The preferred treatment is endoscopic or surgical local resection of the polyp with preservation of the renal unit and with or without segmental

resection of the ureter. The robotic, laparoscopic, open and endoscopic approaches all have been reported.⁵ Prognosis should be considered excellent if complete excision has been carried out.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

Learning points

- ▶ Our case revealed that prenatally detected hydronephrosis may worsen after spontaneous postnatal improvement and a polyp acting as a flip valve may produce intermittent hydronephrosis and symptoms later in life.
- ▶ Patients and parents are usually right and we need to reconsider the situation if there is recurrent presentation.
- ▶ Dietl's crisis is precipitated by over-hydration—our patient had a very important clue, that is, having acute attacks soon after swimming lessons implying that while in water, the skin absorbs extra water which creates extra water load and a sudden increase in the urine output, allowing the polyp to get impacted and covert partial obstruction to overt near complete one.
- ▶ We should evaluate each patient on the merit of their current presentation rather than a background label or diagnosis.
- ▶ Sudden change in the natural history of a disease should be considered a red flag and prompt attention and early diagnosis is necessary to avoid attendant morbidity.

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