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CASE REPORT

Ureteral triplication: A rare anomaly with a variety of presentations

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Abstract Ureteral triplication remains a very rare congenital malformation of the urinary tract with a wide spectrum of presentation. The sporadic nature of this condition and its association with other anomalies makes evidence-based management difficult. We report two cases of triplication in association with the VACTERL syndrome, one developing pelvi-ureteric junction obstruction and the other vesico-ureteric reflux.

Introduction

Ureteric triplication, whilst rare compared to duplication, shares a similar female predominance [1]. Although first reported in 1927 by Perrin, it was classified into four types by Smith [2] in his review of 11 cases in 1946, with over 100 cases subsequently reported [1—3]. Triplication may occur in isolation, in conjunction with other urological anomalies, as part of the VACTERL syndrome. Or inherited as a component of an autosomal dominant condition with amastia [1,3,4].

Pelvi-ureteric junction (PUJ) obstruction remains a common cause of hydronephrosis in the paediatric population. Generally, the upper pole moieties do not form true renal pelvises but often obstruct in association with ectopia and/or uretrocele. In contrast, obstruction at the PUJ has been reported only rarely [5].

We present two cases of ureteric triplication associated with the VACTERL syndrome: one with an upper pole PUJ obstruction and another in conjunction with a complex ipsilateral lower limb anomaly.

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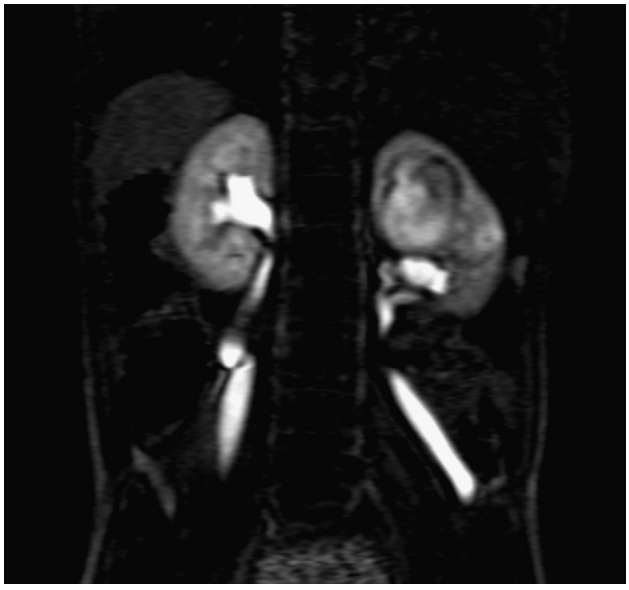

Figure 1 MRU revealing dilated, hydronephrotic upper pole moiety of left kidney.

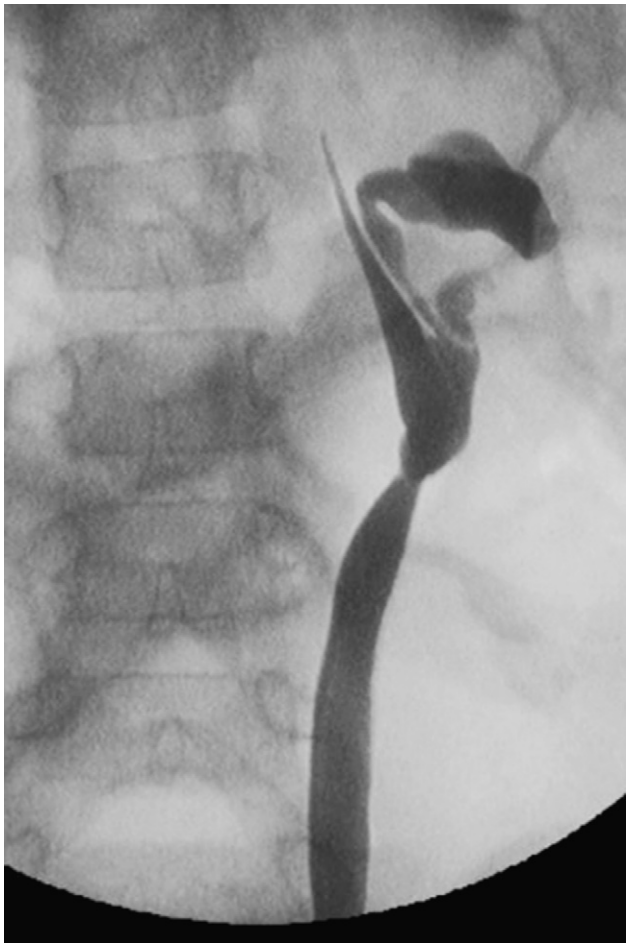

Figure 2 RPG indicating Smith type III left ureteral triplication. Note tapered appearance of upper pole moiety, with an absence of filling, as a result of the PUJ obstruction.

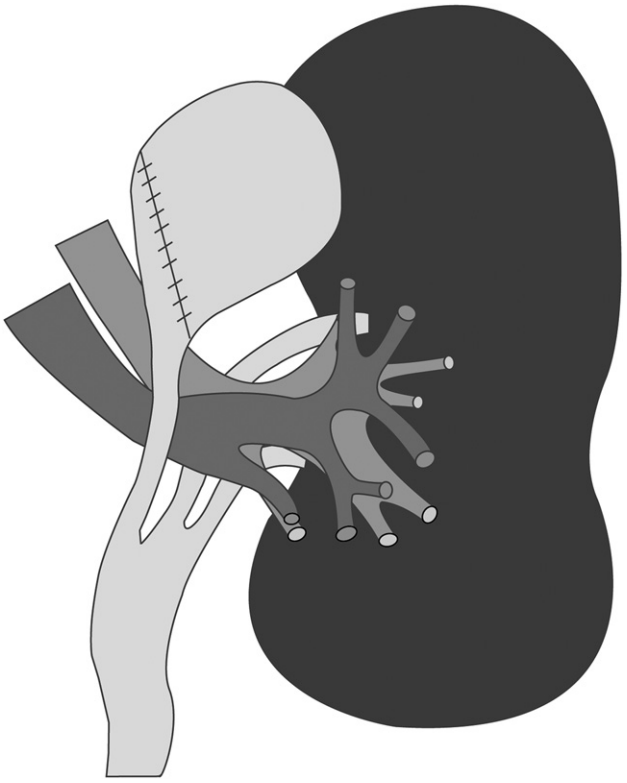

Figure 3 Diagram illustrating anatomical configuration following upper pole pyeloplasty.

Case report

Case 1

A term male infant was diagnosed with the VACTERL syndrome with an anorectal malformation (ARM), a ventricular septal defect and left-sided hydronephrosis initially diagnosed by abdominal ultrasound. A micturating cystourethrogram (MCUG) revealed bilateral VUR for which he was commenced on prophylactic antibiotics. The ARM was initially treated with a colostomy and subsequent anorectoplasty at 3 months of age and the ventricular septal defect repaired at 7 months of age.

Although the patient remained infection free during regular follow up, he was noted to have a duplex left kidney (not seen on initial ultrasound) and increasing hydronephrosis of the upper pole on subsequent ultrasound between 2 and 3 years of age. Intravenous pyelography revealed a probable bifid left renal system with obstruction of the upper pole component.

To delineate the anomalous anatomy, MR urography (MRU) was obtained (Fig. 1), revealing a hydronephrotic upper pole moiety. Retrograde pyelography (RPG) confirmed that the ureter underwent fusiform dilatation at the level of the lower pole of the kidney and then trifurcated (Fig. 2, Smith type III). At subsequent open operation the findings were confirmed, with the attenuated upper ureter found to attach in a non-dependent fashion to a grossly dilated renal pelvis in a typical PUJ configuration. A dismembered pyeloplasty was performed, via a standard approach, with the anastomosis anterior to the renal pedicle (Fig. 3). Performing a dismembered