Unusual variant of inverted Y ureteral duplication with an ipsilateral seminal vesicle cyst and renal dysgenesis

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ABSTRACT

Inverted Y malformation is a rare variant of ureteral duplication with a marked female predominance. We describe a case of inverted Y ureteral duplication with concurrent ectopic ureteral insertion into a seminal vesicle cyst, a ureterocoele and renal dysgenesis, which occurred in a 29-year-old man with lower urinary tract symptoms, haematospermia and postcoital discomfort. To our knowledge, this is the first reported case with this constellation of urogenital abnormalities.

KEYWORDS

Ureteral duplication – Inverted Y – Seminal vesicle cyst – Renal dysgenesis

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Case history

A 29-year-old man presented with urgency, dysuria, haematospermia and postcoital discomfort of 2 months' duration with left abdominal pain. There was no history of fever or haematuria. His medical history included a left renal agenesis and treated bilateral cryptorchidism. Physical examination revealed a palpable, soft and tender intra-abdominal lump occupying the left lumbar region. Genitourinary examination was unremarkable. Laboratory tests were within normal limits. Abdominal ultrasonography demonstrated a left retroperitoneal cystic mass with a hypoechoic intravesical lesion. Computed tomography (CT) showed a cyst measuring 70mm × 66mm × 57mm, expanding from the left renal fossa to a large filling defect in the left bladder base (Fig 1). A ureterocoele with a hydronephrotic kidney was suspected. A cystic lesion of the ipsilateral seminal vesicle was also noted on CT.

On cystoscopy, there was severe elevation of the left hemitrigone with bladder neck obstruction, raising the suspicion of a ureterocoele. The ipsilateral ureteric orifice was not visualised. Magnetic resonance imaging (MRI) demonstrated a seminal vesicle cyst (SVC), which measured 50mm × 18mm × 15mm, and a ureter-like tube expanding from the lower ureter and opening into the cyst (Fig 2).

In order to investigate further, transrectal ultrasonography guided puncture of the ureterocoele was performed with aspiration of 260ml of chocolate coloured fluid and injection of contrast inside (Fig 3). Fluoroscopy and x-ray showed opacification of the left vas deferens (Fig 4). The aspirated chocolate coloured fluid contained spermatozoa.

A diagnosis of an inverted Y ureter with a stenotic ureterocoele and ureteral ectopia into a SVC was considered. The patient underwent an open midline suprapubic, extraperitoneal approach. The specimen was dissected extravesically and resected completely (Fig 5). Microscopically, the ureter-like tube was lined by urothelial epithelium. There were glomeruli and renal tubules with fetal characteristics in the wall of the cystic kidney, suggesting renal dysgenesis. During the six months following resection, he experienced neither haematospermia nor coital discomfort and denied any voiding difficulties.

Discussion

Inverted Y ureter (or caudal ureteral bifidity) is an uncommon variant of partial duplication in which two distal ureteral branches fuse proximally to become a single tube inserting into the renal pelvis.¹ Since the first inverted Y ureter was described in 1913, fewer than 40 cases have been reported in the literature, with a clear female predominance.¹ How this entity can come about remains unclear but it is thought to be secondary to two ureteric buds fusing prior to joining the metanephric blastema.^{1,2} In individuals with an inverted Y ureter, one of the two distal ureteral limbs commonly results in ectopia, a ureterocoele or atresia.^{1,5} This condition may present as pain, urolithiasis, haematuria, infection, incontinence, bladder outflow obstruction and azoospermia or it may be found incidentally.²

To our knowledge, this is the first case of an inverted Y ureter in a male patient in which one limb resulted in an

Figure 1 Sagittal, coronal and axial computed tomography of the abdomen showing the left retroperitoneal cyst expanding from the renal fossa to a large filling defect in the left bladder base. The right urinary tract was normal.

Figure 2 Axial and sagittal magnetic resonance imaging showing a ureter-like tube expanding from the left lower ureter and opening into an ipsilateral seminal vesicle cyst.

ureterocolic with the other limb ending in an ectopic insertion into the seminal vesicle. One previous report documented an inverted Y ureter with a distal limb that was inserted ectopically into the seminal vesicle; the other ureteral segment was normal.⁴ As with ectopic ureters in males,

Figure 3 Transrectal ultrasonography guided puncture of the ureterocolic with aspiration showing chocolate coloured fluid.

Figure 4 Fluoroscopy (A) and x-ray (B) of the pelvis after transrectal ultrasonography guided contrast media injection showing opacification of the vas deferens.

the ectopic branch of an inverted Y ureter always enters the urogenital system above the external sphincter and usually into the Wolffian structures, as in our patient.

Congenital SVCs are generally associated with ureteral abnormalities and renal agenesis/dysplasia owing to the common origin of the ureteral bud and seminal vesicle from the Wolffian duct system.⁵ However, neither ectopic insertion into a congenital SVC nor associated renal dysgenesis have been reported previously in cases of inverted Y ureters. In addition, our patient had experienced bilateral cryptorchidism. Our extensive review of the English literature revealed no other case with coexistence of all of these anomalies in the same patient. Aside from potential teratogenic and genetic factors,⁶ the coexistence of these several anomalies is difficult to explain.

An inverted Y ureter opening into a SVC may remain unnoticed until adulthood, as in this case, when seminal secretions reach the maximum level and are drained into the upper urinary tract.⁵ After years of sexual activity, both the ipsilateral ureterocolic and kidney in our patient filled with seminal fluid, causing lower urinary tract symptoms and a painful abdominal mass respectively. Haematospermia and postcoital discomfort were probably associated with reflux of chocolate coloured fluid from the ureter into the

Figure 5 Gross specimen of the left upper urinary tract showing inverted Y ureteral duplication with the ureterocoele and ectopic ureteral insertion into the seminal vesicle cyst.

SVC during sexual intercourse. As the ureterocoele was stenotic, he had experienced neither haematuria nor infection.

An inverted Y ureter can usually be diagnosed by intravenous urography, CT or cystoscopy with retrograde pyelography. However, neither urography nor pyelography were performed in this case. Instead, MRI was employed, confirming the suspicion of a stenotic ureterocoele, hydronephrotic cystic kidney and SVC. Invasive studies could include percutaneous or transrectal ultrasonography guided needle puncture of the cyst and diagnosis could be confirmed in mature patients by detecting spermatozoa.⁵ In addition, contrast media injection might indicate the anatomical relationship between the ureter and the cyst.⁵ As the SVC in our patient was small and the ureterocoele was large, it was easy to perform transrectal ultrasonography guided needle puncture of the ureterocoele with aspiration. This allowed accurate diagnosis as well as temporary relief of the lower urinary tract symptoms.

There are currently no guidelines on the surgical management of inverted Y ureters. Asymptomatic patients should be followed without intervention. In a symptomatic patient, treatment changes according to the anatomical variant and the associated anomalies. It can include endoscopic unroofing of ureterocoeles, laparoscopic excision of ectopic limbs, laparoscopic ureteroureterostomy for blind limbs and open nephroureterectomy.^{3,6} The present case, with a small SVC, could have been managed conservatively. Laparoscopic excision of the ectopic segment of the ureter with the SVC combined with endoscopic incision of the ureterocoele may be feasible, safe and effective but potential risk factors include infection and recurrence. Consequently, radical excision by nephroureterovesiculectomy is a good treatment option for this condition, especially in symptomatic patients with a dysgenetic kidney.

Conclusions

To our knowledge, this is the first reported case of inverted Y ureteral duplication with concurrent ectopic ureteral insertion into a SVC, a ureterocoele and dysgenetic kidney. Patients diagnosed with such an isolated urinary tract anomaly should be assessed for these related urogenital malformations. Conservative management can be used in selected cases. Nevertheless, radical *en bloc* excision is the standard treatment option for the variant found in our patient.

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