An exceptional cause of macroscopic haematuria: haemorrhagic ureterocele

Clinical History:

Young man with known systemic lupus erythematosus on long-term corticosteroid therapy plus plaquenil, currently suffering from lumbar pain, dysuria and macroscopic haematuria. Physically found afebrile, with left flank and pelvic tenderness but no peritonism.

No significant abnormalities of routine laboratory tests including acute phase reactants and renal function.

Imaging Findings:

Urgent ultrasound (Fig.1) showed a demarcated 5-cm echogenic mass at the left posterolateral aspect of the urinary bladder, with upstream hydronephrosis. Cystoscopy confirmed bloody urine and a large intravesical roundish mass with mildly inflamed overlying mucosa, initially interpreted as submucosal tumour.

When planning further investigation, a CT (Fig.2) obtained three years earlier was found in the Picture Archiving and Communication system (PACS), which diagnosed a thin-walled left intravesical ureterocele with a stone at the ureterovesical junction, ipsilateral mild hydronephrosis, and a 6-mm calyceal calculus. These findings did not lead to treatment.

Comparison with the previous study proved very helpful to elucidate the current CT-urography appearance (Fig.3): the left ureterocele was increased in size with thickened hyperattenuating (50-60 Hounsfield Units) walls and containing a calculus, interpreted as haemorrhagic complication. Ipsilateral hydronephrosis was confirmed, with a second stone at the ureterovesical junction and migrated previous calyceal stone.

Elective ureterocele resection was planned after medical therapy.

Discussion:

Corresponding to a submucosal saccular dilatation of the intramural ureter which prolapses into the urinary bladder, ureterocele is a developmental abnormality resulting from congenital weakness of the lower ureteral wall and narrowed distal meatus. Compared to heterotopic ureteroceles with ectopic ureteral orifice and duplicated renal collecting system which are typical of the paediatric age group, single-system “adult” orthotopic ureteroceles (AOUs) with a normal insertion into the bladder trigone are rarely associated with renal dysplasia, obstruction, reflux and abnormal renal function, and are often incidentally detected in young adults (mean age 31 years; range 20-49 years). The AOU wall consists of three layers, namely ureteral epithelium and bladder urothelium separated by
connective tissue and muscle. In normal conditions, cystoscopy visualizes the AOU with smooth walls covered by normal mucosa [1-3].

Albeit commonly asymptomatic, AOU may manifest with variable symptoms such as flank or back pain, fever, voiding dysfunction and haematuria, resulting from superimposition of urolithiasis, obstruction or urinary infection. Whereas ureteroceles predominate in females (4-7:1), AOU-related stones tend to be more common in males. Occasionally, a long-standing calculus which cannot pass the narrow ureteral orifice may cause mural oedema, ischaemia, pressure necrosis and haemorrhage of the AOU. In this exceptional case, the complicated AOU resulted in a large, bleeding filling defect at both endoscopy and ultrasonography, which raised the suspicion of tumour [1-4]. In the patient reported here the use of CT provided the correct diagnosis by recognition of the characteristic site of an AOU protruding into the bladder lumen at the distal ureteral orifice. An uncomplicated ureterocele is heralded by the classical “cobra head” sign described at intravenous pyelography contrast-enhanced CT and MR-urography. Compared with the normal thin, uniform nonenhancing walls, haemorrhagic AOU is depicted with preserved usual and regular contour, and thickened hyperattenuating periphery reflecting intramural haemorrhage [4, 5]. Furthermore, CT allows comprehensive diagnosis of coexistent lithiasis, upstream hydronephrosis and renal function and aids in the differential diagnosis from transitional cell carcinoma and other non-neoplastic bladder lesions [5-8].

Surgical treatment is required for symptomatic or complicated AOUs, to prevent sepsis and renal function deterioration. Transurethral unroofing represents the preferred first-line approach. Other options such as ureteropelostomy, ureterocele excision with ureteral reimplantation, and nephroureterectomy are reserved for complex cases [1-4].

Differential Diagnosis List: Orthotopic ureterocele complicated by lithiasis and haemorrhage, Ectopic ureterocele, Primary megaureter, Congenital or acquired bladder diverticulum, Retained foreign body, Inflammatory pseudotumour, Bladder abscess, Urinary tuberculosis, Submucosal tumour e.g. leiomyomas, Transitional cell bladder carcinoma, Extrinsic compression from extravesical mass

Final Diagnosis: Orthotopic ureterocele complicated by lithiasis and haemorrhage.

References:


**Figure 1**

**a**

Description: The left kidney (calipers in A) showed upper normal limit size and parenchymal thickness, mild pyelocalyceal dilatation (thick arrows), without appreciable calculi and perirenal fluid. **Origin:** Tonolini M, Department of Radiology, “Luigi Sacco” University Hospital – Milan (Italy)

**b**

Description: The left kidney (calipers in A) showed upper normal limit size and parenchymal thickness, mild pyelocalyceal dilatation (thick arrows), without appreciable calculi and perirenal fluid. **Origin:** Tonolini M, Department of Radiology, “Luigi Sacco” University Hospital – Milan (Italy)
Description: In the urinary bladder, a large (nearly 5 cm) left posterolateral well-demarcated roundish echogenic intraluminal mass (caliper) was present, with dilated ipsilateral pelvic ureter (thick arrow). The bladder was well-distended and otherwise normal. Origin: Tonolini M, Department of Radiology, “Luigi Sacco” University Hospital – Milan (Italy)
Description: Obtained to assess suspected pyelonephritis, CT showed normal thickness and enhancement of the renal parenchyma on the affected left side, minimal perinephric fat stranding (*), mild hydronephrosis and a 6-mm calyceal calculus (arrows). Origin: Tonolini M, Department of Radiology, “Luigi Sacco” University Hospital – Milan (Italy)
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Description: The urinary bladder showed normal mural thickness, a thin-walled sizeable left intravesical ureterocele (thin arrows) and another stone (arrowheads) at the uretero-vesical junction.

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Origin: Tonolini M, Department of Radiology, “Luigi Sacco” University Hospital – Milan (Italy)
Description: Unenhanced CT acquisition showed the known left ureterocele with increased size, thickened hyperattenuating wall (50-58 Hounsfield Units, thin arrows) containing a stone (arrowheads). Note some intravesical air (+), dilated ipsilateral ureter (thick arrows). Origin: Tonolini M, Department of Radiology, “Luigi Sacco” University Hospital – Milan (Italy)
Description: Nephrographic-phase post-contrast CT (b...d) confirmed left ureterocele with thickened hyperattenuating wall (thin arrows) containing a stone (arrowheads); another calculus at the ureterovesical junction (arrow in c) causing upstream hydronephrosis (thick arrows). Origin: Tonolini M, Department of Radiology, “Luigi Sacco” University Hospital – Milan (Italy)
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Description: Excretory-phase acquisition (e...g) showed the ureterocele (thin arrows) as filling defect in the opacified urinary bladder. Note calculus at the ureterovesical junction (arrow). Origin: Tonolini M, Department of Radiology, “Luigi Sacco” University Hospital – Milan (Italy)
Description: Excretory-phase acquisition (e.g.) showed the ureterocele (thin arrows) as a filling defect in the opacified urinary bladder. Note calcific stones, ipsilateral hydronephrosis (thick arrow). Origin: Tonolini M, Department of Radiology, “Luigi Sacco” University Hospital – Milan (Italy)
Description: Additionally, excretory-phase acquisition depicted opacified left hydronephrosis (thick arrow) with opacified collecting system, dilated and mildly distorted upper calyces from probable chronic pyelonephritis. Lower pole calyceal lithiasis was not present anymore. Origin: Tonolini M, Department of Radiology, “Luigi Sacco” University Hospital – Milan (Italy)