



Ureteric Telangiectasia: An Unusual Cause of Chronic Unilateral Hematuria and Treatment

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Abstract

Chronic Unilateral Hematuria (CUH) is defined as unilateral hematuria seen on flexible cystoscopy with normal standard biochemical and radiological investigations. An 18-year-old gentleman was referred to our department for unilateral painless CUH secondary to mid and distal ureteric telangiectasia. He was a smoker but did not have a personal or family history of cancer or bleeding diathesis.

He underwent diagnostic ureteroscopy and nephroscopy with samples taken for cytology and histology, both of which were negative for a high-grade malignancy. Ureteric stenting was performed after ureteroscopy in preparation for nephroscopy, appeared to tamponade the telangiectasia, and resolve the bleeding completely. This case identifies a unique cause of CUH and its treatment.

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Case report description

Chronic unilateral hematuria is defined as unilateral hematuria seen on flexible cystoscopy with normal standard biochemical and radiological investigations. Ureteropyeloscopic evaluation is recommended and lesions can either be identified (discrete or diffuse) or not identified with differentials including calculi, malignancy, hemangiomas, varices, minute venous rupture and telangiectasia [1]. Patients are usually asymptomatic but can occasionally present with massive hematuria causing clots resulting in renal colic or anemia necessitating packed cell transfusion [2].

Treatment options include oral homeostatic agents (carbazochrome sodium sulfonate, tranexamic acid and aminocaproic acid), pelvicalyceal instillation (silver nitrate and povidone-iodine [3]) endourological therapy (diathermy fulguration and

Holmium: yttrium aluminium garnet (YAG) or Neodymium: YAG laser ablation [4]), segmental renal artery segmental embolization [5] or nephrectomy. Treatment failure has been reported more often in diffuse lesions [6] but fortunately, no such cases have been reported after 1999, which may be in part due to improved diagnostics¹, highlighting the duality of the challenge this condition poses.

Mr H A A is an 18-year-old gentleman who was referred to Urology for intermittent painless gross hematuria of 2 months duration precipitated by exercise. His past medical history includes left shoulder surgery for a labral tear but was otherwise unremarkable. He is an active smoker (4 cigarettes a day for a year), a student and does not have any family history of cancer or bleeding diathesis.

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On examination, no telangiectasia seen, no flank masses felt, external genitalia was normal and no renal or bladder mass or hydronephrosis observed on bedside ultrasonography. Urinalysis confirmed the presence of red blood cells (615 per high-powered field, HPF) but no malignant cells were seen nor bacteria grown. His serum haemoglobin concentration (15.3g/dL), platelet count ($245 \times 10^9/L$) clotting studies (Prothrombin Time, PT 11.2s, Activated Partial Thromboplastin Time, APTT 29.6s) and kidney function (estimated Glomerular Filtration Rate, eGFR 95ml/min/1.73m²) were normal. Computed Tomography (CT) Urogram did not detect any calculi, masses, filling defects or vascular thromboses, fistulae or malformations.

A flexible cystoscopy (Figure 1) was performed which visualised a jet of hematuria from the left ureteric orifice but not the right and a normal bladder and urethra.

Left retrograde pyelography did not reveal any filling defects (Figure 2) and he underwent diagnostic ureteroscopy, which re-

vealed telangiectasia over the distal and mid ureter (Figure 3) and biopsies of the adjacent mucosa were taken. The mucosa of the proximal ureter and pelvi-ureteric junction appeared normal and samples for cytology were taken. A 4.8Fr multi length Boston Scientific Percuflex Plus ureteral stent was placed and the patient counselled for left diagnostic ureteropyeloscopy and hemostasis.

Mr H underwent left ureteric stent removal and diagnostic ureteropyeloscopy two weeks after and the previously seen telangiectasia in the distal and mid ureter had completely resolved (Figure 4). All calyces were individually inspected and found to be normal and further samples taken for cytology.

A 4.8Fr multi length Boston Scientific Percuflex Plus ureteral stent was again placed at the end of the procedure and removed two weeks after. The histology and cytology results from both procedures have been summarised below in Table 1.

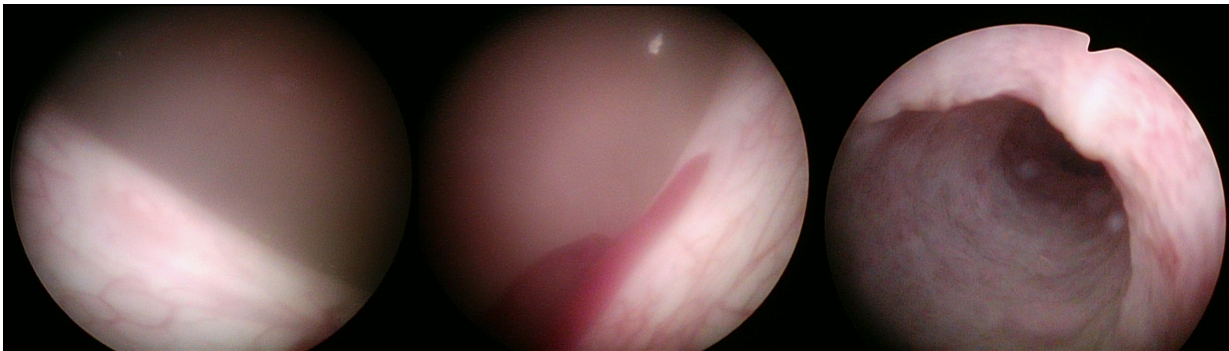


Figure 1: Flexible cystoscopy demonstrating a normal right ureteric orifice (left), a jet of blood from the left ureteric orifice (center) and a normal urethra (right).



Figure 2: Left retrograde pyelography demonstrating smooth flow of contrast up the distal and mid ureter (left) across the proximal ureter (center) into the kidney (left).

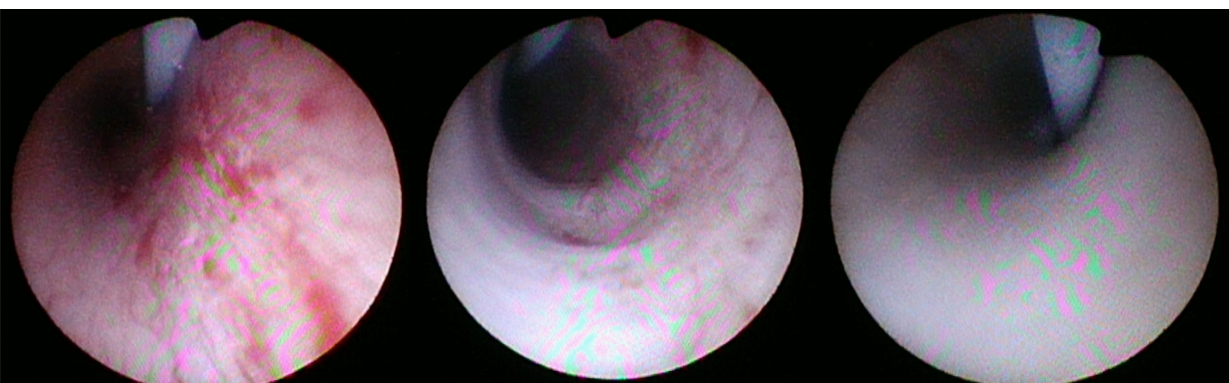


Figure 3: Intra-operative photographs demonstrating telangiectasia seen over left distal (left) and mid ureter (center) and normal proximal ureter (right).

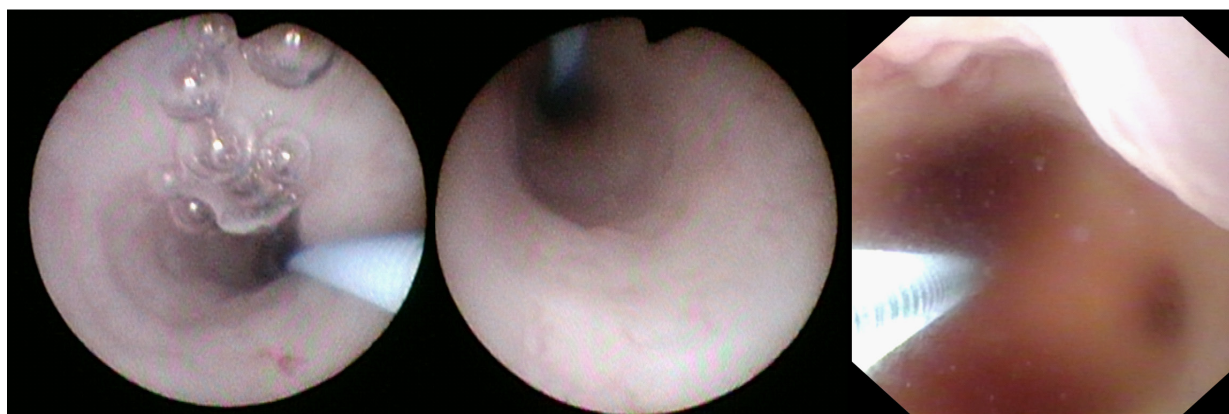


Figure 4: Intra-operative photographs demonstrating resolution of telangiectasia in the distal (left) and mid ureter (center) and normal calyces (right).

Table 1: Summary of histology and cytology results.

Site	Ureteroscopy		Ureteropyeloscopy
	Histology	Cytology	Cytology
Bladder	-	Benign	-
Left distal ureter	Fibrous connective tissue	Benign	-
Left mid ureter	Fibrous connective tissue	Benign	-
Left proximal ureter	-	Atypical cells	Atypical cells
Left kidney	-	-	Atypical cells

Discussion

Telangiectasia of the urinary tract is very uncommon but has been reported in Hereditary Hemorrhagic Telangiectasia (HHT) [7,8] and Ataxia Telangiectasia (AT) syndromes [9,10], though so far only in the kidney, bladder, prostate and urethra. Mr H’s telangiectasia were in the ureter and as he did not have any family history of bleeding diathesis or personal history of recurrent epistaxis with oral, nasal and cutaneous telangiectasia [11] or progressive cerebellar dysfunction [12], genetic tests for either disease were not offered [13].

Unilateral hematuria of ureteric origin has been described but only in the context of an idiopathic stricture in a 10-year-old boy [14]. A distal ureteric stricture 2 to 3cm from the vesicoureteric junction was seen on retrograde pyelography, which was too narrow to admit an ureteroscope, and a stent was placed for 8 weeks. Interestingly, even 8 months after stent removal he did not experience any further micro or macroscopic hematuria, had normal serum haemoglobin concentration and was weaned off the intravenous iron infusions which he was previously reliant on. Unfortunately, cytology and histology analyses as well as endoscopic inspection of the remaining urinary tract if performed were not detailed.

Mr H’s hematuria has not recurred and though histology of his distal and mid ureter did not suggest malignancy and no tumors were observed endoscopically, atypical cells were reported in his proximal ureter and kidney. As such, he has been planned for regular clinical review.

Conclusion

- Ureteric telangiectasia is an uncommon cause of chronic unilateral hematuria.
- Telangiectasia of the urinary tract can occur in isolation and are not limited to the kidney, bladder, prostate and urethra.
- A trial of ureteric stenting before hemostasis for hematuria from uretric telangiectasia is reasonable.

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