

# Further observations on the “spaghetti sign” in upper urinary tract hemorrhage

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## Abstract

The “spaghetti sign” is recognized as a radiological sign of upper urinary tract hemorrhage. The sign was first described in the urinary bladder during intravenous urography, but it has subsequently been described on retrograde pyelography and in the urographic phase of contrast CT. We report the observation of the “spaghetti sign” in the bladder on ultrasonography and on Magnetic Resonance Urography (MRU), modalities in which the sign has not been previously described. We suggest that the observations may provide a useful guide when ultrasonography and/or MRU are employed in the search for the source of massive hematuria. We also report two additional cases of hematuria in whom the “spaghetti sign” is demonstrated in the urographic phase of contrast CT.

**Keywords:** Hematuria, magnetic resonance urography, spaghetti sign, ultrasonography

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## INTRODUCTION

The determination of the source of bleeding in hematuria is a common challenge in clinical and radiological practice. Tubular filling defects in the urinary bladder in a patient with hematuria were first observed on intravenous urography and captioned the “Spaghetti sign” because of their visual resemblance to spaghetti.<sup>[1]</sup> These filling defects represent blood clot casts (or molds) of the ureter, and their presence confirms that the source of hemorrhage must be supravescical.

The “spaghetti sign” has also been described in the excretory phase of contrast CT and on retrograde pyelography.<sup>[2-5]</sup>

This presentation documents for the first time, to the best of the authors’ knowledge, the observation of the

“spaghetti sign” in the bladder during ultrasonography and on magnetic resonance urography (MRU).

We also include two additional cases in which the sign is demonstrated during computed tomography (CT) urography.

## SUBJECTS AND METHODS

The imaging modalities which we most frequently employ in the investigation of patients presenting with hematuria include abdominal ultrasound, plain abdominal CT, and CT urography if indicated after a plain study. In selected cases, especially in pregnant women and young children, we employ abdominal MRU in place of CT because of radiation concerns.

Depending on the imaging findings, we perform cystoscopy and ureteroscopy for tissue biopsy or removal of urinary tract calculi, as appropriate.

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In the last 3 years, we encountered four patients with massive hematuria in whom a positive “spaghetti sign” was exhibited. In all of them, the source of hematuria was traceable to renal pathology.

## CASE REPORTS

### Case 1

A 62-year-old man presented at the emergency department with a 12-h history of severe lower abdominal pain, dysuria, and hematuria, with passage of blood clots.

On examination, his urinary bladder was distended, and he had marked suprapubic tenderness.

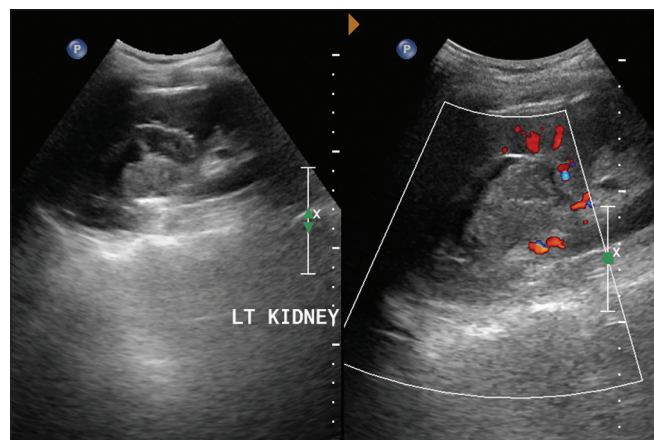
Apart from a high blood pressure of 170/100 mmHg, no other abnormality was shown. His bleeding and clotting profiles and his hematocrit value were normal.

A plain abdominal X-ray was noncontributory. An abdominal ultrasound showed a 5.5 cm × 4.5 cm moderately echogenic mass lesion in the superior aspect of the left renal pelvis, which on color Doppler was hypovascular in comparison with the renal parenchyma [Figure 1]. The right kidney was normal.

Tubular filling defects were noted layered posteriorly in the urinary bladder [Figure 2] and shifted with patient’s positioning. These were considered to be blood clot casts of the left ureter, giving a positive “spaghetti sign.”

A strong ureteric jet effect was observed on the right side but was completely absent on the left.

The prostate was not enlarged, and the liver, pancreas, and spleen were normal.



**Figure 1:** A 62-year-old man presenting with hematuria. Left renal ultrasound without and with color Doppler. There is a large mass in the renal pelvis, hypovascular relative to the surrounding renal parenchyma

Contrast CT of the abdomen and chest was performed for further evaluation of the renal mass.

This confirmed a mass arising from the superior aspect of the left renal pelvis, showing moderate contrast enhancement, and distorting the upper pole calyces. Transitional cell carcinoma was considered a high possibility. There was no perinephric tumor extension, regional lymphadenopathy, or chest metastases.

A three-way Foley’s catheter was introduced into the bladder for irrigation. In addition, a high oral fluid intake was instituted, and the clot retention was successfully cleared.

The patient was informed that surgery would be required as a definitive treatment, but he declined and left against medical advice.

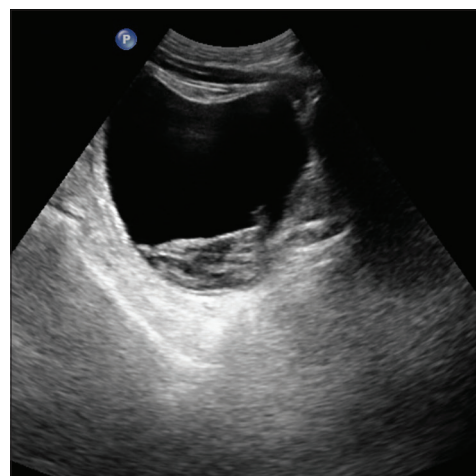
### Case 2

A 5-year-old boy was brought to the emergency department complaining of right upper flank pain and frank hematuria, with passage of clots. He had sustained trauma to the right flank several hours earlier, during horseplay with other boys.

On examination, he was tender in the right flank, but there was no palpable mass and the abdomen was soft and lax. The laboratory studies were normal, except for a slightly elevated prothrombin time of 49.5 s.

Abdominal ultrasound showed probe tenderness over the right renal angle, but no renal abnormality was detected. The left kidney, liver, pancreas, and spleen were normal.

The urinary bladder contained a large filling defect with linear strands in its base [Figure 3].



**Figure 2:** A 62-year-old man presenting with hematuria. Transverse ultrasound of the urinary bladder showing linear strands consistent with blood clots, layering in its posterior wall. The strands shifted with changes in patient’s position



**Figure 3:** A 5-year-old boy presenting with massive hematuria following blunt trauma to right flank. Sagittal ultrasound of the urinary bladder shows a large aggregate of linear strands of blood clot layering in its posterior wall

These moved with changes in patient's position and were considered to be blood clot molds of the ureter, probably the right, and indicating a positive "spaghetti sign."

As the source of hemorrhage was not determined by ultrasound, it was decided that MRU should be performed, which was done under sedation with chloral hydrate.

Both kidneys were shown to be essentially normal [Figure 4].

Linear filling defects were again demonstrated in the urinary bladder [Figure 5], consistent with a positive "spaghetti sign." Although the imaging studies failed to demonstrate renal injury, the history of blunt right renal angle trauma, renal angle tenderness, and a positive "spaghetti sign" point to the right kidney as the likely source of hemorrhage.

The bladder was irrigated with normal saline to remove the clots, and along with increased oral fluid intake, the urine became clear. A follow-up ultrasound study was entirely normal.

As the parents declined cystourethroscopy, the patient was discharged and advised for follow-up visits.

### Case 3

A 35-year-old woman was referred to us from another hospital where she presented with a 1-day history of right-side abdominal pain and gross hematuria. There was nothing of note in the medical history or clinical examination. An emergency abdominal ultrasound at the referring hospital was reported as showing mild right hydronephrosis and a filling defect in the bladder which was suggestive of a bladder mass.



**Figure 4:** A 5-year-old boy presenting with massive hematuria following blunt trauma to right flank. Coronal T2-weighted static fluid magnetic resonance urography shows both kidneys to be essentially normal

Contrast abdominal CT showed two nodules abutting the right renal pelvis, with features suggesting angiomyolipomas. At the excretory phase, tubular filling defects were observed in the dependent portion of the urinary bladder [Figure 6]. There was no evidence of a bladder mass.

A three-way Foley's catheter was introduced into the urinary bladder, and irrigation with normal saline initially yielded a lot of clots, but eventually, the urine became completely clear. The patient was discharged on antibiotics and is being followed up, as she may require angioembolization.

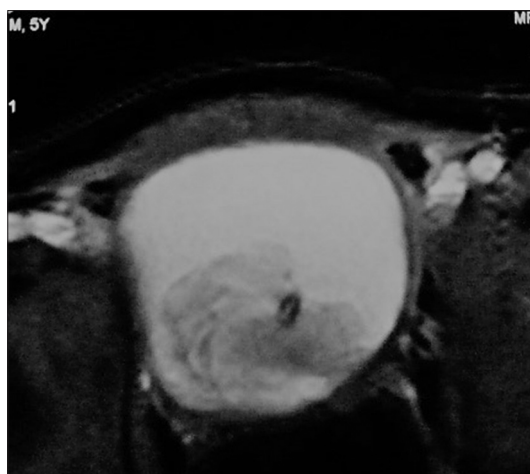
### Case 4

A 61-year-old woman presented with gross, painless hematuria, which had occurred intermittently for 4–5 months. She was anemic but was not diabetic or hypertensive, and clinical examination revealed no other significant finding. A plain CT abdomen showed no urinary tract calculus or pelvicalyceal distension.

Contrast CT showed a pedunculated filling defect in the right renal pelvis [Figure 7]. This was considered as papilloma or transitional cell carcinoma, and a right ureteroscopy was advised.

The left kidney was normal. The liver, gallbladder, pancreas, and spleen were also normal.

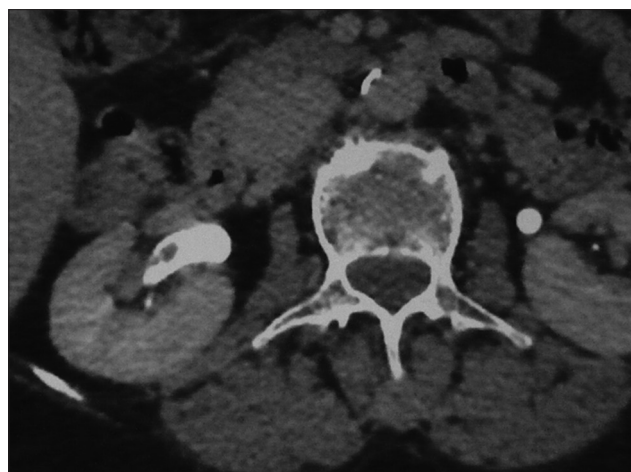
A coronal reformatted CT image in the excretory phase showed convoluted filling defects in the urinary bladder [Figure 8], considered to be blood clot casts of the right ureter, giving a positive "spaghetti sign."



**Figure 5:** A 5-year-old boy presenting with massive hematuria following blunt trauma to the right flank. Axial T2-weighted fat-saturated static fluid magnetic resonance urography showing the urinary bladder containing frond-like tubular filling defects, representing blood clots, lying posteriorly and aggregated more on the right side. The central low-signal intensity shadow is the tip of a Foley's catheter



**Figure 6:** A 35-year-old woman presenting with right abdominal pain and massive hematuria. Axial image of the urinary bladder in the excretory phase of contrast computed tomography showing tubular filling defects in its most dependent part. The air-filled balloon of a Foley's catheter is noted in the center of the bladder



**Figure 7:** A 61-year-old woman presenting with gross, painless hematuria. Axial image of contrast computed tomography shows a pedunculated mass in the right renal pelvis. Biopsy came back as papillary urothelial carcinoma

Cystoureteroscopy was performed under general anesthesia, and a mass was confirmed projecting into the right renal pelvis. A biopsy was taken, and the histology report returned as papillary urothelial carcinoma.

The patient was referred to an oncology center, where a right nephroureterectomy was performed.

She is still under follow-up.

## DISCUSSION

Hematuria is a common clinical problem and may vary in severity from microscopic to massive.



**Figure 8:** A 61-year-old woman presenting with gross, painless hematuria. Coronal reformatted image of the urinary bladder in the excretory phase of contrast computed tomography shows convoluted linear filling defects, aggregated more to the right side of the bladder

Locating the source of hemorrhage can be challenging, as it can originate anywhere in the urinary system, and often requires the combination of clinical and imaging findings.<sup>[6,7]</sup> In general, initial hematuria is associated with urethral lesions, terminal hematuria with lesions of the posterior urethra, bladder neck and trigone, and total hematuria with supravescical lesions.<sup>[8]</sup> These clinical patterns of presentation are however not invariable.

Since the “spaghetti sign” in the bladder indicates blood clot molds of the ureter, a positive sign invariably confirms that the source of bleeding is proximal to the bladder.

However, by virtue of its mode of formation, a positive “spaghetti sign” can only occur in the context of massive hematuria, as the hemorrhage must be intense enough for its clot to produce casts of the ureter. On the other hand, it is in a situation of massive hematuria that an urgent determination of the source of hemorrhage becomes mandatory.<sup>[9]</sup>

To determine the origin of hematuria, multidetector CT is now the gold standard. It is preferred to excretory urography in most cases,<sup>[6,7,10]</sup> with a sensitivity of 96%–100% and a specificity of 94%–100% in the detection of calculi, for example.

MRU is an evolving modality, and because it involves no ionizing radiation, it is useful in the imaging of pregnant women and young children, as in our Case 2. In pregnant women, MRU is of particular advantage in distinguishing physiological from calculus ureteric obstruction.<sup>[10,11]</sup>

The major drawbacks of MRU are its insensitivity to calculi and its long imaging time, with consequent susceptibility to motion artifacts.<sup>[10,11]</sup> In young children, sedation may be required, as in our Case 2.

As previously reported with intravenous urography and with the excretory phase of CT,<sup>[1-5]</sup> a positive “spaghetti sign” observed in the bladder on ultrasound (Cases 1 and 2) or on MRU (Case 2) helps direct the search of the source of hemorrhage to the upper urinary system.

It is important to distinguish the tubular filling defects constituting the “spaghetti sign” from those seen in cystitis glandularis. The latter are small, focal polypoidal bladder mucosal thickenings due to metaplasia of the urothelium and said to be associated with chronic bladder inflammation and considered by some as premalignant.<sup>[12-15]</sup> Apart from differences in clinical presentation, lesions of cystitis glandularis are static, while blood clots in the bladder shift with the patient’s position, gravitating to the most dependent part.

Blood clots originating from vesical or perivesical pathologies may also shift with gravitation, but the absence of the linear strands that characterize clots released from the ureters is an important differentiating factor. Although a large number of clots emanating from the ureter may clump together [Figures 2 and 3], the observation of a few separate strands is adequate to confirm their ureteric origin.

The ureteric jet effect, which demonstrates the flow of urine into the bladder from the ureters,<sup>[16-18]</sup> is also of value when a positive “spaghetti sign” is observed on ultrasonography. Because of the reduced rate of urine flow from a ureter clogged with blood clots, the jet effect from that ureter will be absent, as observed in our patient (Case 1), or markedly impaired compared with the normal side.

When a positive “spaghetti sign” is present, this discrepancy in the ureteric jet effect between the two sides serves to determine the side of disease.

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### Conflicts of interest

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