

Clinics in Diagnostic Imaging (50)

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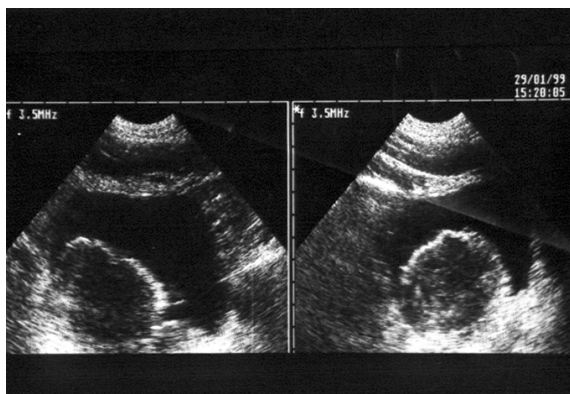


Fig. 1 US scans of the bladder taken in the longitudinal (left) and transverse (right) planes.

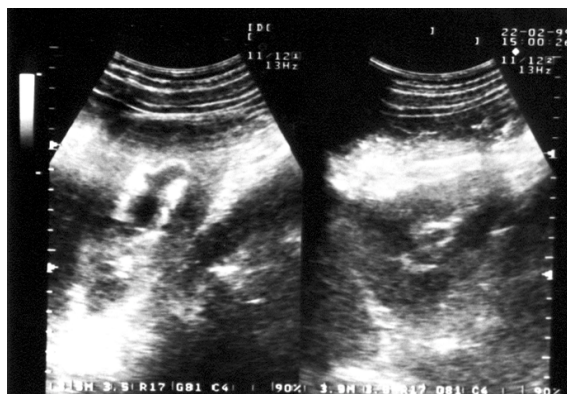


Fig. 2 US scans of the right kidney taken in the longitudinal (left) and transverse (right) planes.

CASE PRESENTATION

A 39-year-old man presented with painless macroscopic haematuria. He had been treated for suspected urinary tract infection for one week and when the haematuria did not resolve, he was referred to our institution. On retrospective questioning, he was found to have had multiple episodes of painless haematuria over the past year. There was no other significant past medical history. The physical examination was unremarkable. Ultrasonography (US) was performed. What do the US scans (Figs. 1 and 2) show?

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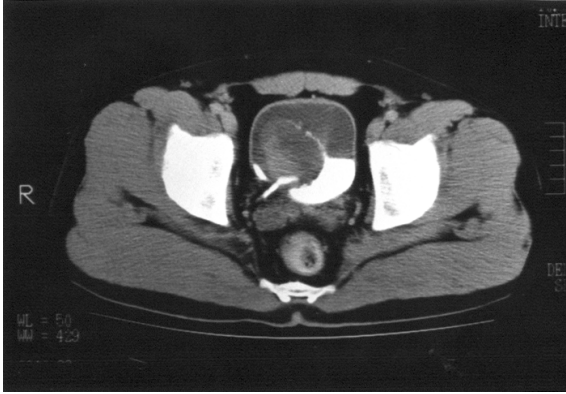


Fig. 3 Enhanced CT of the pelvis shows a rounded mass of heterogeneous density located at the right posteroinferior aspect of the bladder.

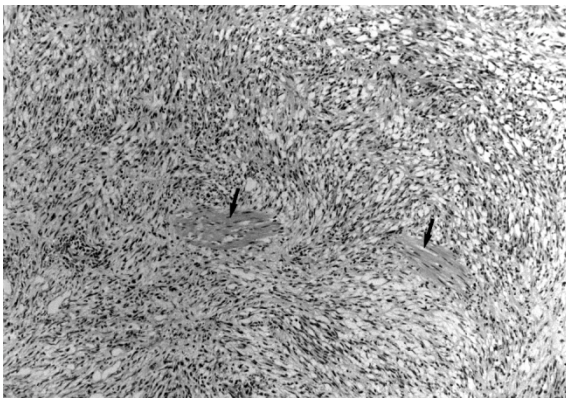


Fig. 4 Photomicrograph shows a leiomyosarcoma invading and partially obliterating the muscularis propria of the bladder wall. Residual smooth muscle bundles are present (arrows) (Haematoxylin and eosin, x200 original magnification).



Fig. 5 Photomicrograph shows leiomyosarcoma penetrating into the perivesical fat (upper field) and invading into a blood vessel (arrow) (Haematoxylin and eosin, x200 original magnification).

IMAGE INTERPRETATION

US scans showed a large soft tissue mass within the bladder. It had lobulated margins and was located on the right posterior bladder wall (Fig. 1). Moderate hydronephrosis affecting the right kidney was also present (Fig. 2). Computed tomography (CT) of the abdomen and the pelvis was performed to delineate the extent of the disease. CT showed a 4 cm x 5 cm x 5 cm heterogeneous lesion at the right inferoposterior aspect

of the bladder, with involvement of the right ureteric orifice (Fig 3). There was associated dilatation of the right ureter and pelvicalyceal system (not shown).

Cystoscopy was performed under general anaesthesia. An extensive non-papillary fleshy tumour involving the right ureteric orifice was found. The lesion was resected transurethrally to its base. Histopathological examination revealed a high-grade leiomyosarcoma that invaded the smooth muscle of the bladder wall (Fig. 4).

DIAGNOSIS

Leiomyosarcoma of bladder

CLINICAL COURSE

Cystoprostatectomy was offered but the patient insisted on deferring the operation. He presented again two months later with haematuria, clot retention and anaemia. Total cystoprostatectomy and ileal conduit was then performed. At surgery, extensive tumour recurrence at the posterior bladder wall, with trigone and bladder neck involvement, was found. Histopathological examination of the resected specimen revealed a high-grade, stage 3 spindle cell tumour. Both the perivesical fat and the right postero-lateral resection margins were positive for tumour (Fig. 5).

The patient was then referred for radiotherapy. After two sessions of radiotherapy, however, he was admitted for abdominal discomfort, distension and nausea. A repeat CT of the pelvis showed a large soft tissue pelvic mass which extended to the pelvic inlet (Figs. 6A-B). Abdominal radiograph showed multiple dilated bowel loops and air-fluid levels, typical of small bowel obstruction (Fig. 7). CT of the thorax showed small nodules at the lung bases, as well as mediastinal lymphadenopathy (not shown). The subacute intestinal obstruction was attributed to bowel oedema from radiation, and decision was made to treat the patient conservatively.

The radiotherapy was continued, and the patient's intestinal obstruction subsequently resolved with improvement of his clinical condition. Repeat CT of the pelvis performed upon completion of radiotherapy showed significant diminution in the size of the tumour mass (Fig. 8). Chemotherapy was considered but the patient's condition suddenly worsened when he developed severe hypercalcaemia and a massive right malignant pleural effusion. He died five months from the initial diagnosis of bladder leiomyosarcoma.

DISCUSSION

Macroscopic haematuria is not an uncommon symptom. In fact, it is the most common presenting symptom of urinary tract malignancies⁽¹⁻³⁾. Tan et al⁽¹⁾ reported that

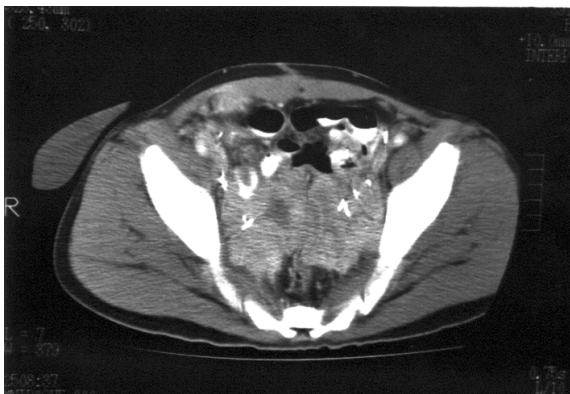


Fig. 6A Enhanced CT taken at the level of the mid-pelvis shows a large recurrent lobulated heterogeneous tumour occupying most of the pelvic cavity.



Fig. 6B Enhanced CT taken at the level of the hip joints show extension of tumour into the ischiorectal fossae.

of all the patients evaluated for haematuria, about half were found to have a urological lesion. Of these, 20% was due to urolithiasis and 14% was attributed to urological malignancy. In their series, transitional cell carcinoma was the most common malignancy. Therefore macroscopic haematuria should always be carefully evaluated and not ignored.

Most investigators are in agreement that macroscopic haematuria presenting in an adult patient will require imaging of the upper urinary tract and endoscopy of the bladder to exclude malignancy. Intravenous urogram combined with cystoscopy used to be the most commonly-used combination of modalities. Recently, in a series published by Yip et al⁽⁴⁾ of 468 patients with first presentation of painless haematuria, 18% had bladder cancer while 5% had renal cancer. These authors found that US was significantly more sensitive in the detection of bladder tumours compared to urography while in the upper tract, there was no significant difference in the sensitivity of lesion detection. US is now being more frequently used in the imaging for haematuria as it does not carry radiation hazards and is applicable in patients with contrast agent allergy and renal impairment.

The diagnostic pathway for haematuria can be

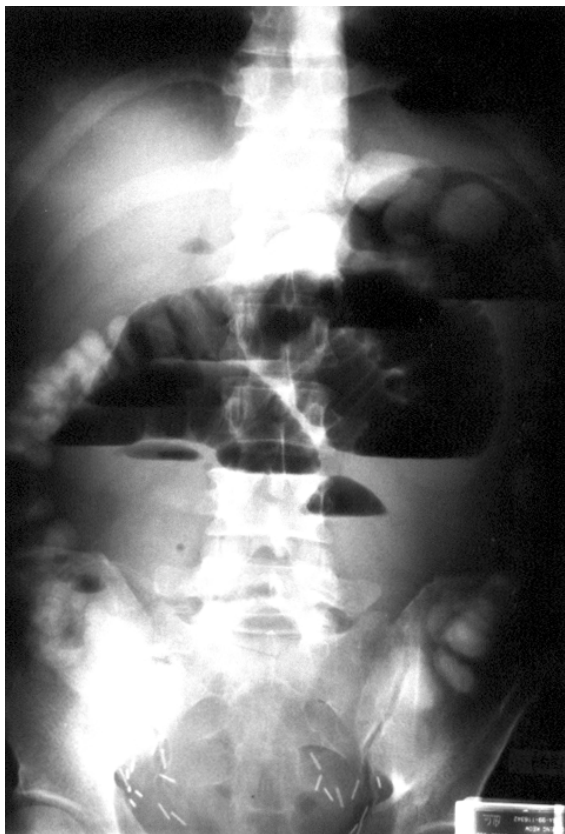


Fig. 7 Erect abdominal radiograph shows multiple loops of dilated small bowel, with several air-fluid levels

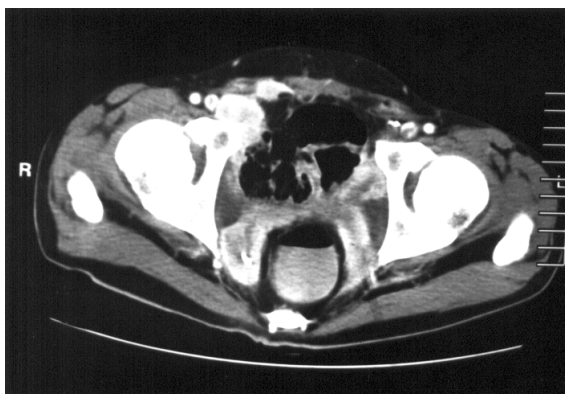


Fig. 8 Enhanced CT of the pelvis taken at the same level as Fig. 6B shows significant reduction in the size of the tumour following further radiotherapy.

further expedited by combining US and diagnostic cystoscopy at the same sitting on the day of the clinic visit⁽⁵⁾. In our department, ultrasound machines are readily available in most of the clinic consultation rooms. We have found clinic US to be useful in detecting gross kidney and bladder lesions. With its use, a more focused diagnostic pathway could be afforded. In some cases of bladder tumours detected by clinic US, patients could be listed for definitive endoscopic surgery while awaiting additional imaging, such as CT for staging purposes.

Apart from urolithiasis and urological malignancies, other causes of macroscopic haematuria include urinary

tract infections, urinary tuberculosis, inflammatory conditions of the urinary tract, trauma, prostate disease and polycystic kidneys⁽⁵⁾. Medical conditions such as glomerulonephritis and blood dyscrasias may also cause haematuria. Further investigations should therefore be conducted, where appropriate, should initial imaging studies and endoscopy fail to determine the cause of haematuria.

However, careful history taking and examination combined with urine analysis will often provide clues to the diagnosis. Useful points to consider include: the demographic features of the patient, pattern of haematuria, associated symptoms, presence of features of malignancy. It is most important not to assume the diagnosis and treat as for urinary tract infections without performing necessary imaging and endoscopic examination since haematuria can be intermittent. The cessation of bleeding may therefore give a "false" sense of security.

Soft tissue sarcomas are a group of malignancies arising in extraosseous tissues derived from the mesoderm. Less than 5% of soft tissue sarcomas arise from the genitourinary tract. They constitute only 1-2% of all malignant genitourinary tumours⁽⁶⁾. The histological diagnosis of leiomyosarcoma by appropriate immunohistochemical studies is essential. In our patient, the tumour showed immunohistochemical reactivity for vimentin and smooth muscle actin, the latter supporting a smooth muscle origin of this spindle cell tumour. High mitotic activity, foci of necrosis and invasion into smooth muscle bundles of the muscularis propria were also noted. Differential diagnoses included sarcomatoid carcinoma, pseudosarcomas and postoperative spindle cell nodules⁽⁷⁾.

Russo et al⁽⁶⁾ studied the factors predictive of outcome in 43 cases of genitourinary sarcoma, 10 of which were bladder sarcomas. Of these 10 cases, 8 presented with painless macroscopic haematuria and 2 with dysuria. A reduced survival was noted in patients with tumours greater than 5 cm in size, high grade tumours, and positive surgical resection margins. Interestingly, bladder sarcomas were noted to have a higher survival rate than paratesticular, prostate and kidney sarcomas, in that order. This was probably because of the alarming presenting sign of haematuria. In general, patients with high-grade invasive tumour of greater than 5 cm in size or evidence of metastatic disease carry a poorer prognosis.

Current data on leiomyosarcoma covers a relatively small number of cases with diverse regimens of treatment. The exact role of chemotherapy and radiotherapy remains unclear although radiotherapy did bring about marked reduction in the size of the tumour in our patient. Complete surgical resection remains the

treatment modality that affords long term survival. In summary, urological sarcoma is a rare entity that carries a poor prognosis in the majority of cases. Early recognition and diagnosis may allow a better survival rate.

ABSTRACT

A 39-year-old man presented with intermittent painless macroscopic haematuria. He was subsequently diagnosed to have a high grade leiomyosarcoma of the bladder. He underwent total cystoprostatectomy and radiotherapy but died of metastatic disease five months after the initial diagnosis. Aspects of the imaging and diagnostic pathway of painless haematuria are discussed. The management of leiomyosarcomas in the urinary system is highlighted.

Keywords: leiomyosarcoma, bladder tumour, ultrasonography, computed tomography

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