Solitary Fibrous Tumour of the Urinary Bladder in a Young Woman Presenting with Haemodynamic-Relevant Gross Haematuria

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1. Case report

A 24-yr-old woman was admitted as emergency to the Department of Urology with gross haematuria for 3 hr and lower abdominal pain beginning 2 hr earlier. There was no history of urological or gynaecological disease. Sonography revealed an endovesical structure suspicious for a clot retention. At an emergency endoscopy, a large tumour on the right cranialateral bladder wall covered with normal mucosa was detected. The origin of haemorrhage remained unclear. The gynaecological examination showed a tumour of 8.5 by 7.8 cm in size in the area of the right ovary suspected to be a germinal cell tumour. At excretory urography, the upper urinary tract on the left side was slightly...
dilated, and the indwelling catheter was displaced to the left (Fig. 1). Computed tomography revealed a large tumour filling the urinary bladder with suspected connection to the tumour of the right ovary (Fig. 2).

Due to the haemodynamically relevant haemorrhage with the haemoglobin dropping from 12.8 g/dl to 7.7 g/dl in 36 hr, a transurethral resection of the tumour was performed; the weight of the resected specimen was 166 g. Laparoscopy after 10 d showed no pathological findings in the right lower abdomen.

Histologically, the tumour revealed spindle cells arranged among a collagen background (Fig. 3). The immunohistochemical analysis showed a strong positivity of tumour cells to CD34 (Fig. 4) and Bcl2. In addition, they were positive for vimentin and CD99 but negative for EMA, MNF, CEA, Ca-125, α-SMA, and CK7. Furthermore, there was a low Ki-67 expression. According to these results, a solitary fibrous tumour of the bladder wall was diagnosed.

Partial cystectomy with total resection of the remaining tumour tissue was performed (Fig. 5) and regular clinical controls were recommended. In the following 2 yr, regular follow-up examinations with magnetic resonance imaging, transurethral biopsies in the first year, and cystoscopies in the second year revealed no recurrence of disease.

2. Discussion

This is the first case in the literature that reports on a solitary fibrous tumour (SFT) of the bladder wall in a
young woman and the first case that presented with acute haemodynamic-relevant gross haematuria.

A solitary fibrous tumour is a rare mesenchymal tumour entity commonly localized in the pleura. It derives from dendritic interstitial cells that express CD34. It is also known to occur in other extrapleural sites such as the central nervous system, gastrointestinal tract, nasal cavity, or breast. In the urinary tract, manifestations in the prostate, the seminal vesicle, the spermatic cord, the kidney, and the bladder have been described. SFT is extremely rare in the urinary bladder, and only nine cases in the English literature have been reported so far [1–6]. It particularly affects men between the ages of 40–70. The clinical presentation of SFT of the urinary bladder is variable. It can become manifest in different symptoms such as voiding difficulties, gross haematuria, urinary retention, or nonspecific abdominal pain. Other incidental findings have also been described.

Macroscopically, the tumour presents covered with intact mucosa, as in our case. Histologic characteristics include the patternless pattern, which is dominated by spindle cells arranged among a collagen background. Immunohistochemically, CD34 is a highly sensitive marker for SFT (90–95% of cases), as are CD99 (70% of cases) and Bcl2 [2,7]. The tumour reported herein was positive for CD34, CD99, and Bcl2 and, therefore, in combination with its histological pattern, clearly defined as SFT.

Even though extrathoracic SFTs are usually described as benign neoplasias, about 10% will metastasize or show recurrence [8]. Pleomorphism and a high mitotic rate are assumed indicators for malignant SFT; however, cases showing benign histology but with a malignant clinical course have also been reported, so the prognosis of extrapleural SFT is not predictable with certainty on histological patterns only [9,10]. Careful and long-term follow-up is highly important. In our case, the tumour showed low Ki-67 expression and low mitotic rate, both indicating rather benign histology.

In cases of mesenchymal tumour of the urinary bladder presenting with spindle-cell neoplasias, the differential diagnosis of SFT should be considered. It is strictly necessary—although sometimes difficult—to clearly differentiate it from other malignant entities such as sarcomatoid transitional cell carcinoma, inflammatory pseudotumour, leiomyosarcoma, or haemangiopericytoma.

Concerning the therapeutic options for SFT, it has been suggested that margin-free resection of the tumour is the most important determinant of the patient’s clinical outcome [9]. Therefore, complete but conservative surgical excision—depending on the tumour size—should be performed. In our case, partial cystectomy with margin-free resection zone was performed. The 2-yr follow-up revealed no recurrence of disease.

Conflicts of interest

The authors have nothing to disclose.

EU-ACME question

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Question:

What is the most suitable option for treatment of a solitary fibrous tumour of the bladder at present?

A. Vaporisation of the tumour mass.
B. Bacillus Calmette-Guérin (BCG) instillation therapy.
C. Radical cystoprostatovesiculectomy.
D. Margin-free resection of the tumour mass.
References


