Case Report

Malignant solitary fibrous tumor of the urinary bladder progressing to widespread metastases and death: a rare case report and literature review

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Abstract: Solitary fibrous tumor (SFT) of the urinary bladder has been rarely reported and malignant bladder SFT is even rarer. Here we present a case of an African-American male with SFT of the urinary bladder (intermediate risk) initially treated by cystoprostatectomy at the age of 59 years. Eight years later, he developed recurrence with widespread metastases to the liver, lungs, and abdominal cavity. He then received temozolomide and bevacizumab with good disease control. However, treatment was paused due to declining performance status. Follow-up at 1 year demonstrated growth of the metastatic lesions. Despite restarting therapy, the patient expired, 11 years after the original diagnosis. Autopsy was performed and revealed widespread metastases within the abdominal cavity (abdominal sarcomatosis) as well as liver, bilateral lung, and diaphragmatic involvement. The cause of death was determined to be metastatic SFT. A comprehensive literature review was performed. Although SFTs are commonly considered benign, a subset of SFTs of the urinary bladder behave aggressively. Risk assessment and proper follow-up for recurrence and metastasis is necessary. The patient was also found at autopsy to have two gastrointestinal stromal tumors (GISTs) in the stomach and near the gastroesophageal junction. To the best of our knowledge, this is the first reported case of a primary urinary bladder SFT resulting in death or having concurrent, multifocal GISTs, and only the second case of a bladder SFT that developed metastases after the initial diagnosis.

Keywords: Solitary fibrous tumor, urinary bladder, metastases, gastrointestinal stromal tumor

Introduction

Solitary fibrous tumor (SFT) is a rare fibroblastic neoplasm that accounts for less than 2% of all soft tissue masses. It is characterized histologically as having a haphazard distribution of ovoid to spindle cells and so-called “staghorn” vessels, and a recurrent molecular NAB2-STAT6 gene rearrangement [1]. Although it was initially identified in the pleura, SFT can involve any anatomic site including the visceral organs, bones, and superficial and deep soft tissues [1]. These tumors are observed in middle-aged adults ranging between 20 and 70 years old with no sex predilection. Although the majority have a favorable clinical course, SFT is notoriously difficult for prognostication because of the propensity for late relapse or even metastases in 10-40% of cases [2]. Surgical management is the mainstay of treatment for SFT with emphasis on obtaining tumor-negative margins. SFT primary to the urinary bladder has been rarely reported. To date, 29 cases were reported in the English literature. Among these, 3 cases displayed local recurrence and only 1 case progressed with metastases with limited follow-up [3]. Herein we report a case with SFT of the urinary bladder who developed recurrence with widespread metastases and eventually expired in the 11th year post-diagnosis.

Case presentation

The patient was a 59-year-old African-American male who first presented for urologic evaluation due to an episode of painless hematuria. The physical exam was remarkable only for mild lower abdominal tenderness. Workup with com-
Computed tomography (CT) imaging revealed a 9.1 × 8.5 × 8 cm heterogeneously enhancing mass apparently arising from the dome of the bladder. He underwent cystoscopy with bladder wash cytology which was negative for malignancy. Repeat CT demonstrated expansion of the mass to 12 × 11 × 10 cm. Repeat cystoscopy revealed reduced bladder volume due to mass effect but was otherwise unremarkable. Laparoscopy revealed a large, dense, vascular mass arising from the bladder in the midline and occupying most of the lower abdomen. There were no obvious intra-abdominal metastases. Biopsy of the mass revealed plump spindle cells, “staghorn” vessels and no necrosis. The tumor cells were positive for CD34, CD99, and BCL2 by immunohistochemical (IHC) stains, which supported a diagnosis of SFT. Given the size/extent of disease, the patient subsequently underwent a laparoscopic radical cystoprostatectomy with extensive pelvic lymphadenectomy and urinary diversion. Gross examination of the specimen showed a 12 × 12 × 11 cm well-circumscribed tumor located in the fundus of bladder with no mucosal involvement. The tumor had tan-white, firm, and solid cut surfaces. Histologic sections demonstrated similar morphology when compared with the biopsy with extensive (15%) necrosis and no prominent cytologic atypia. The tumor was diagnosed as SFT of the bladder. The mitotic rate was 1/10 high-power fields (HPF). All margins and submitted lymph nodes (33) were negative for tumor and the tumor was staged as pT2bN0 according to the American Joint Committee on Cancer (AJCC), 8th Edition. The patient was discharged on post-operative day 7 in good condition.

Oncologic follow-up was accomplished with yearly CT imaging of the abdomen and pelvis. At serial appointments, the patient had no urinary or abdominal symptoms and had no abnormal physical exam or CT findings suggestive of recurrent or metastatic disease for 8 years.

In the 9th post-operative year, a routine renal ultrasound (US) incidentally noted three new heterogeneous liver lesions, the largest measuring 4.0 cm. Abdominal magnetic resonance imaging (MRI) showed multiple hypervascular liver nodules, as well as multiple enhancing mesenteric nodes which were consistent with peritoneal carcinomatosis. Subsequently, a percutaneous liver biopsy was performed. Final pathology was consistent with metastatic SFT, identical in morphology to the originally resected tumor. Next-generation sequencing studies were performed by Foundation Medicine, Inc. on formalin-fixed, paraffin-embedded liver tissue and identified a TP53 R248W gene alteration and NAB2-STAT6 fusion. Additional findings included stable microsatellite status and low tumor mutational burden (3 mutations per megabase).

A chest CT was obtained for completion of staging and revealed scattered nodules in both lungs, consistent with metastatic disease. The patient was started on sunitinib (37.5 mg daily) but did not tolerate the therapy and subsequent CT imaging revealed significant progression of peritoneal disease. Sunitinib therapy was discontinued, and the patient was switched to temozolomide (150 mg/m²) and bevacizumab (5 mg/kg). Approximately nine months after the initiation of this regimen, he had increasing fatigue, anorexia, and weight loss. Anti-cancer therapy was paused due to declining performance status; imaging demonstrated stability of metastatic lesions, so the patient was monitored off-therapy. Symptomatically, the patient did well in the interim, but follow-up at one year demonstrated growth of the metastatic lesions. At that point, temozolomide and bevacizumab therapy was resumed. After restarting treatment, the patient had multiple hospital admissions, including for deconditioning, an infected perinephric abscess, and COVID-19 infection.

Despite continued treatment, a subsequent US demonstrated extensive progression of mesenteric disease. Interventional radiology performed another image-guided biopsy of the abdominal masses, which again revealed histology similar to the original tumor. Confirmatory IHC stains showed the tumor cells were positive for STAT6, while negative for CD34. Due to continued declining functional status and intractable disease progression, further oncologic treatment was deferred. The patient transitioned to hospice care and expired, 11 years after the original diagnosis. An autopsy was performed. Initial internal examination revealed abdominal sarcomatosis: numerous white, soft tumor nodules on the peritoneal surfaces, omentum and organs and throughout the retro-
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Figure 1. Gross autopsy findings. A: Dense fibrous adhesions throughout the abdomen. Numerous white soft tumor nodules are present on peritoneal surfaces, diaphragm, omentum (white arrow), and mesentery, as well as the liver (black arrow) and lungs. B: Tumor nodules on liver surface (black arrow).


peritoneum (Figure 1A, 1B). The mesentery was diffusely replaced and distorted by numerous soft, white tumor nodules. The entire small and large bowel was encased by tumor as well (Figure 2A). Firm, red-white, well-circumscribed nodules were noted throughout the pleura and pulmonary parenchyma, up to 2.1 cm in greatest dimension (Figure 2D). The liver was riddled with tan-pink to purple, firm, well-circumscribed
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masses, up to 6.5 cm in greatest dimension (Figure 2G). The cut surfaces of the nodules in the liver, lungs and intestine were white and red with focal hemorrhage and necrosis. The histology and IHC profile of the nodules were similar to the originally diagnosed SFT (Figure 2B, 2C, 2E, 2F, 2H and 2I).

In addition, within the gastric body, adjacent to the lesser curvature, as well as near the gastroesophageal (GE) junction, two similar-appearing 1 cm well-circumscribed subserosal calcified nodules were discovered. The histology and IHC studies revealed spindle cell lesions (Figure 3A) that were positive for DOG1 (Figure 3C, 3D), CD117 (Figure 3E, 3F), and smooth muscle actin (SMA), focally positive for desmin, while negative for STAT6 (Figure 3B) and S-100, consistent with gastrointestinal stromal tumors (GISTs).

Discussion

Solitary fibrous tumor (SFT) is a rare mesenchymal neoplasm that has been reported to arise from many organs including the urinary bladder [4]. Our PubMed search using the key words “solitary fibrous tumor” and “bladder” revealed 29 cases of urinary bladder SFT in the English literature (Table 1). SFT is most commonly diagnosed between the fifth and seventh decades of life with an equal distribution among men and women [5]. Based on our search, the urinary bladder SFT occurs 3.3 times more in males than females with an age ranging from 11 to 85 years (mean =54.2; Male: 55.4; Female: 50.1).

From the published data, including the present case, predominant symptoms include hematuria (37%), urinary obstruction (20%), and abdominal/pelvic pain (20%). To a lesser extent, urinary urgency and frequency (17%), dysuria (13%), and abdominal bloating/fullness (7%) were observed at presentation. However, asymptomatic cases diagnosed incidentally may also occur [4]. Additionally, hypoglycemia can be the initial indicator of SFT, which is due to a paraneoplastic syndrome known as Doege-Potter syndrome. This syndrome is secondary to ectopic secretion of insulin-like growth factor II by the tumor [6]. Radiologic features of SFTs of the bladder are considered nonspecific [7]. Sonography may show a variable pattern of echogenicity with relatively well-defined margins [7]. On non-contrast CT scans, SFT demonstrates soft-tissue attenuation while on contrast CT, strong enhancement and associated cysts, regions of hemorrhage, and/or necrosis can be observed [7]. MRI shows predominantly low or intermediate signal intensity on both T1- and T2-weighted images, dependent on collagenous and fibrous stromal content, vascularity, and chronicity of...
Table 1. Reported cases of solitary fibrous tumor of the urinary bladder and the present case

<table>
<thead>
<tr>
<th>Author [Ref]</th>
<th>Year</th>
<th>Age/ Sex</th>
<th>Clinical presentation</th>
<th>Gross findings</th>
<th>Tumor size (cm)</th>
<th>Necrosis &amp; Mitosis</th>
<th>Treatment</th>
<th>Follow-up (months/ recurrence or metastasis)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Vargas et al. [15]</td>
<td>2021</td>
<td>64/F</td>
<td>lower abdominal fullness</td>
<td>N/P</td>
<td>N/P</td>
<td>N/P</td>
<td>Surgical resection</td>
<td>N/P</td>
</tr>
<tr>
<td>Sun et al. [16]</td>
<td>2020</td>
<td>52/M</td>
<td>Urinary urgency and frequency</td>
<td>N/P</td>
<td>7</td>
<td>Absent &amp; Low mitotic activity</td>
<td>Partial cystectomy</td>
<td>12 months/Absent</td>
</tr>
<tr>
<td>Kratiras et al. [17]</td>
<td>2019</td>
<td>31/M</td>
<td>Hematuria, dysuria, dull abdominal pain</td>
<td>N/P</td>
<td>5.3</td>
<td>Present &amp; 23/10 HPF</td>
<td>Transurethral resection followed by radical cystectomy</td>
<td>5 months/Absent</td>
</tr>
<tr>
<td>Rovegno et al. [18]</td>
<td>2019</td>
<td>69/M</td>
<td>Abdominal pain, hematuria, lower urinary tract symptoms</td>
<td>N/P</td>
<td>10</td>
<td>Present (&gt;10%) &amp; 0/10 HPF</td>
<td>Mass resection, radical cystoprostatectomy</td>
<td>N/P</td>
</tr>
<tr>
<td>Urbina-Lima et al. [6]</td>
<td>2019</td>
<td>61/M</td>
<td>Generalized asthenia, weight loss and hypoglycemia</td>
<td>N/P</td>
<td>23</td>
<td>N/P &amp; 2/10 HPF</td>
<td>Partial cystectomy</td>
<td>N/P</td>
</tr>
<tr>
<td>Kouba et al. [3]</td>
<td>2017</td>
<td>33/F</td>
<td>Hypoglycemia</td>
<td>Well circumscribed mass with white, myxoid-like appearance</td>
<td>3.5</td>
<td>N/P &amp; 0/10 HPF</td>
<td>Transurethral resection</td>
<td>132 months/Absent</td>
</tr>
<tr>
<td>**Kouba et al. [3]</td>
<td>2017</td>
<td>41/M</td>
<td>Urinary obstruction</td>
<td>Well circumscribed mass with white, myxoid-like appearance</td>
<td>5.7</td>
<td>N/P &amp; 4/10 HPF</td>
<td>Surgical resection</td>
<td>12 months/Recurrence at 12 months</td>
</tr>
<tr>
<td>Tong et al. [19]</td>
<td>2017</td>
<td>85/F</td>
<td>Dysuria</td>
<td>Grayish yellow kernel, solid and tenacious cut surface with grey red outer membrane</td>
<td>12</td>
<td>N/P</td>
<td>Complete resection</td>
<td>3 months/Absent</td>
</tr>
<tr>
<td>Tanaka et al. [4]</td>
<td>2016</td>
<td>60/M</td>
<td>Incidental US finding</td>
<td>Tumor with regular surface</td>
<td>8</td>
<td>N/P &amp; 0/10 HPF</td>
<td>Wide excision</td>
<td>24 months/Absent</td>
</tr>
<tr>
<td>Tanaka et al. [4]</td>
<td>2016</td>
<td>60/M</td>
<td>Incidental US finding</td>
<td>Well-circumscribed mass</td>
<td>4</td>
<td>Absent &amp; 1/10 HPF</td>
<td>Partial cystectomy and radical prostatectomy</td>
<td>10 years/Absent</td>
</tr>
<tr>
<td>Mustafa et al. [20]</td>
<td>2016</td>
<td>36/F</td>
<td>Urgency, straining to void and increased abdominal girth</td>
<td>N/P</td>
<td>10</td>
<td>Absent &amp; &lt;2/10 HPF</td>
<td>Cystectomy with mass excision</td>
<td>N/P</td>
</tr>
<tr>
<td>Dozier et al. [5]</td>
<td>2015</td>
<td>41/M</td>
<td>Weight loss, progressive abdominal bloating</td>
<td>Gray-white mass with ill-defined whorled-like pattern and randomly assorted tan fleshy nodules</td>
<td>28</td>
<td>Present &amp; 10/10 HPF</td>
<td>Complete surgical resection</td>
<td>8 months/Absent</td>
</tr>
<tr>
<td>Tian et al. [21]</td>
<td>2014</td>
<td>66/M</td>
<td>N/P</td>
<td>Encapsulated gray-white tumor with multinodular and focally necrotic cut surface</td>
<td>7.9</td>
<td>Present &amp; N/P</td>
<td>N/P</td>
<td>2 years/Absent</td>
</tr>
<tr>
<td>*Otta et al. [10]</td>
<td>2014</td>
<td>78/M</td>
<td>Hematuria, acute urinary retention</td>
<td>N/P</td>
<td>N/P</td>
<td>Transurethral resection</td>
<td>41 months/Recurrence at 5 months</td>
<td>N/P</td>
</tr>
<tr>
<td>Mozafarpour et al. [22]</td>
<td>2014</td>
<td>54/M</td>
<td>Intermittent abdominal pain, hematuria</td>
<td>Encapsulated gray creamy spongy tissue</td>
<td>13.9</td>
<td>Present &amp; &lt;3/10 HPF</td>
<td>Radical surgical excision</td>
<td>12 years/Absent</td>
</tr>
<tr>
<td>Spairani et al. [23]</td>
<td>2014</td>
<td>60/M</td>
<td>Voiding difficulty</td>
<td>Lobulated unencapsulated, well-circumscribed mass with smooth, firm, and white-tan cut surfaces</td>
<td>9</td>
<td>Absent &amp; 1/10 HPF</td>
<td>Partial cystectomy</td>
<td>11 months/Absent</td>
</tr>
<tr>
<td>Cheng et al. [24]</td>
<td>2012</td>
<td>67/M</td>
<td>Low abdominal pain, microscopic hematuria</td>
<td>Encapsulated gray-white tumor with multinodular and focally necrotic cut surface</td>
<td>16</td>
<td>Present &amp; &gt;4/10 HPF</td>
<td>Partial cystectomy and segmental resection of the intestine</td>
<td>18 months/Absent</td>
</tr>
</tbody>
</table>

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<table>
<thead>
<tr>
<th>Reference</th>
<th>Year</th>
<th>Gender</th>
<th>Age</th>
<th>Symptoms</th>
<th>Diagnosis</th>
<th>Follow-up</th>
<th>Treatment</th>
<th>Pathologic Findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>Wang et al. [25]</td>
<td>2010</td>
<td>50/M</td>
<td>Terminal gross hematuria, residual urine sensation, urination pain</td>
<td>N/P</td>
<td>8</td>
<td>N/P</td>
<td>Surgical resection</td>
<td>9 months/Absent</td>
</tr>
<tr>
<td>Bruzzone et al. [26]</td>
<td>2010</td>
<td>74/M</td>
<td>Ovoidal mass coated by adipose tissue and with well delimited margins</td>
<td>10</td>
<td>N/P</td>
<td>Radical surgical excision</td>
<td>N/P</td>
<td></td>
</tr>
<tr>
<td>Heinzelbecker et al. [27]</td>
<td>2008</td>
<td>24/F</td>
<td>Hematuria</td>
<td>N/P</td>
<td>8.5</td>
<td>N/P</td>
<td>Transurethral resection follow partial cystectomy</td>
<td>2 years/Absent</td>
</tr>
<tr>
<td>Tzelepi et al. [28]</td>
<td>2007</td>
<td>59/F</td>
<td>Well circumscribed unencapsulated polyloid mass with white, solid and gelatinous cut surface</td>
<td>8.5</td>
<td>N/P &amp; &lt;1/10 HPF</td>
<td>Radical cystectomy</td>
<td>77 months/Absent</td>
<td></td>
</tr>
<tr>
<td>Kim et al. [29]</td>
<td>2004</td>
<td>56/M</td>
<td>Voiding difficulty, frequency</td>
<td>Encapsulated, rubbery mass with, creamy white, multinodular, whorled cut surface</td>
<td>12</td>
<td>N/P &amp; 2-3/10 HPF</td>
<td>Wide excision</td>
<td>12 months/Absent</td>
</tr>
<tr>
<td>Leite et al. [30]</td>
<td>2004</td>
<td>60/M</td>
<td>Incidental MRI finding (preoperative workup for prostate cancer)</td>
<td>Tumor with pale, elastic, fasciculate, homogeneous cut surface</td>
<td>3.2</td>
<td>Absent &amp; 3/10 HPF</td>
<td>Surgical resection</td>
<td>11 months/Absent</td>
</tr>
<tr>
<td>Corti et al. [31]</td>
<td>2001</td>
<td>50/M</td>
<td>Pelvic pain, dysuria, hematuria</td>
<td>Well-circumscribed firm nodular mass</td>
<td>6.5</td>
<td>Absent &amp; &lt;2/10 HPF</td>
<td>Cystoprostatectomy</td>
<td>18 months/Absent</td>
</tr>
<tr>
<td>Westra et al. [32]</td>
<td>2000</td>
<td>67/M</td>
<td>Incidental cystoscopy finding during TURP</td>
<td>Well-circumscribed mass with yellow-white, solid, and whirled cut surface</td>
<td>4</td>
<td>Absent/10/10 HPF</td>
<td>Cystoprostatectomy</td>
<td>9 months/Absent</td>
</tr>
<tr>
<td>Westra et al. [32]</td>
<td>2000</td>
<td>67/M</td>
<td>Incidental MRI finding (recurrent prostate cancer surveillance)</td>
<td>Well-circumscribed mass with yellow-white, solid, and whirled cut surfaces</td>
<td>N/P</td>
<td>Absent/N/P</td>
<td>Transurethral resection</td>
<td>1 month/Absent</td>
</tr>
<tr>
<td>Bainbridge et al. [33]</td>
<td>1997</td>
<td>50/F</td>
<td>Urinary frequency, &quot;prolapsing sensation&quot; upon voiding</td>
<td>N/P</td>
<td>5.2</td>
<td>Absent &amp; 1/10 HPF</td>
<td>Transurethral resection</td>
<td>18 months/Absent</td>
</tr>
<tr>
<td>Bainbridge et al. [33]</td>
<td>1997</td>
<td>42/M</td>
<td>Pelvic pressure</td>
<td>Well-circumscribed mural mass with gelatinous cut surface and numerous small cyst-like spaces</td>
<td>20</td>
<td>Absent &amp; 3/10 HPF</td>
<td>Partial cystectomy</td>
<td>6 months/Absent</td>
</tr>
<tr>
<td>Present case</td>
<td>2023</td>
<td>59/M</td>
<td>Well-circumscribed tumor with tan-white, firm, solid cut surface</td>
<td>12</td>
<td>Present (15%) &amp; 1/10 HPF</td>
<td>Cystoprostatectomy, Temozolomide, bevacizumab</td>
<td>11 years/Metastases to liver, lungs, peritoneum at 8 years. Died of disease at 11 years</td>
<td></td>
</tr>
</tbody>
</table>

*Cases with local recurrence. *Cases with metastases. *Year of report. M, male; F, female; N/P, not provided; ref., reference; HPF, high power field.
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the tumor [5, 7]. Non-specific symptoms and radiologic findings necessitate histopathologic diagnosis along with immunohistochemistry and other ancillary testing [4].

Grossly, SFTs are typically well-circumscribed masses with nodular cut surfaces [8]. Occasionally, they may show hemorrhage, myxoid changes, and cystic degeneration [8]. Histologically, SFTs are typically composed of haphazardly arranged ovoid to spindle cells in the so-called “patternless pattern”, with pale eosinophilic cytoplasm in a background of variable collagenous stroma. Characteristic branching and hyalinized staghorn-shaped blood vessels are often appreciated [8]. Its wide histologic spectrum includes paucicellular lesions with abundant stromal keloidal-type collagen to highly cellular tumors and myxoid or lipomatous differentiation [8].

Immunohistochemistry plays a crucial role in the diagnosis [5]. The neoplastic cells often show strong and diffuse expression of CD34 [5, 8]. They are also commonly positive for CD99, vimentin and BCL2, while negative for CD117, SMA, desmin and S-100 [5]. NAB2-STAT6 gene fusions are pathognomonic for SFT and IHC staining for STAT6 is an extremely sensitive and specific surrogate marker for all fusions and is very helpful in the diagnosis of SFT [8].

Although the malignant criteria are defined as large tumor size, high mitotic count, cytologic atypia, necrosis, and infiltrative growth [5, 8], a new risk stratification model based on a point system is believed to be more accurate in predicting the clinical course of the disease. Points are assigned for age (<55 years (0 point), ≥55 years (1 point)); tumor size (<5 cm (0 point), 5 cm to <10 cm (1 point), 10 cm to <15 cm (2 point) and ≥15 cm (3 point)); mitotic figures/10HPFs (0 (0 point), 1-3 (1 point) and ≥4 (2 points)); and necrosis (<10% (0 point), ≥10% (1 point)). Based on total points, the tumor is then assigned a low risk (0-3 total points), intermediate risk (4-5 total points) and high risk (6-7 total points) [8, 9]. For our patient (age: 59 years; tumor size: 12 cm; mitoses: 1/10 HPF; necrosis: 15%; total score: 5 points), the risk of metastases based on the modified four-variable risk model was intermediate.

In general, the rate of recurrence (distant or local) has been reported as 10-30% or 10-40% for SFTs [2, 8]. Including the present case, our review of the literature discovered 25 cases of urinary bladder SFT where follow-up information was available. Three (12%) of these cases had progressive disease with local recurrence (at 5, 12, and 72 months) and two (8%), including our case, had metastatic disease (at 96 and 130 months) [3]. Due to the variable clinical course and unpredictable behavior of SFTs, surgery is recommended as the treatment of choice [5]. For unresectable tumors, chemotherapy and radiation therapy have also been offered with a variable success rate [5].

In our case, next generation sequencing identified NAB2-STAT6 gene fusions and a TP53 R248W gene mutation. This result is consistent with a prior study of 91 SFT cases in which TP53 mutations were found in 41% of malignant SFTs and suggested that dysfunction of TP53 contributed to malignant transformation [11]. TP53 could be a genomic driver for malignancy in our current case. As this is only the second case of urinary bladder with metastasis, the underlying genomic underpinning for aggressive behavior of bladder SFT needs more case studies.

Coexistence of other type(s) of tumor(s) with SFT is not common. According to the literature, bladder leiomyoma with synchronous SFT of the pleura [12], concurrent malignant SFT arising from the omentum and endometrial endometrioid adenocarcinoma [13], and pleural SFT, multiple GISTs, moyamoya disease, and hyperparathyroidism in a patient with neurofibromatosis type 1 [14] have been reported, all with an uncertain mechanism for coexistence.

In conclusion, SFT is a fibroblastic neoplasm that rarely arises from the urinary bladder and causes non-specific symptoms. The diagnosis depends primarily on histopathologic and immunophenotypic findings. The majority of cases behave indolently and have a favorable prognosis with complete resection. However, clinicians should be aware of the risk of recurrence and distant metastases, and patients should have appropriate follow-up.

We have described a case of primary SFT of the urinary bladder with initial treatment success from complete surgical resection but later recurrence with widespread metastases and eventual death. To our knowledge, this is the...
first reported case of a patient with urinary bladder SFT who died of the disease (autopsy-proven), and only the second case reported that developed metastases after the initial diagnosis. In addition, this malignant SFT coexisted with two GISTs within the stomach and near the GE junction, for which the clinical significance and pathobiology remain unclear.

Disclosure of conflict of interest

None.

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References


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