Case report Open Access

A case of an inflammatory myofibroblastic tumor of the bladder in a young woman

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Abstract

We herein report a relatively rare case of an inflammatory myofibroblastic tumor (IMT) of the bladder in a young woman. In contrast to IMTs of other organs, IMTs of the lower urinary tract involve the mixture of a non-neoplastic lesion with a neoplastic lesion; therefore, the determination of whether other neoplastic lesions are present is important. In this case, magnetic resonance imaging confirmed a pedunculated lobulated tumor that protruded from the posterior left bladder wall into the lumen. The lobulated area exhibited a low-intensity signal on T1-weighted imaging (T1WI) and a high-intensity signal similar to that of urine on T2-weighted imaging (T2WI). Morphologically, the peduncle resembled a submucosal mass with faint low-intensity signaling on T1WI and faint high-intensity signaling on T2WI. These findings differ from those of a neoplastic lesion of the bladder mucous membrane. The presence of lesions from the muscularis propria to the submucosa and in areas with markedly high-intensity signals exhibiting a nodular fasciitis pattern on T2WI is considered useful for the differential diagnosis of IMT on imaging examination.

Keywords: Inflammatory myofibroblastic tumor, Urinary bladder, Inflammatory pseudotumor, Magnetic resonance imaging, Submucosal tumor

Introduction

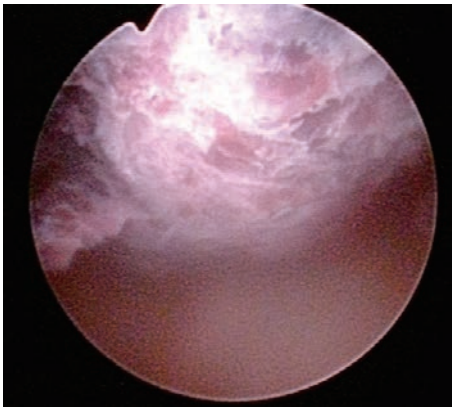
An inflammatory myofibroblastic tumor (IMT) is a neoplastic lesion composed of proliferations of spindle-cell myofibroblasts. It is considered a type of inflammatory pseudotumor (IPT)[1] and is characterized by inflammatory cell infiltration of lymphocytes and plasma cells. We herein present a case of a young woman with an IMT of the bladder together with a brief discussion of the literature.

Case Presentation

A woman in her twenties with an unremarkable medical history presented with a 1-month history of lower abdominal discomfort. The patient had visited her local physician and was prescribed antibiotics for suspected cystitis; however, the symptom did not improve. Abdominal ultrasonography revealed a bladder mass. The mass was first assumed to be proliferative cystitis but was later suspected to be a bladder tumor because of its increase in size over a 2-week period. The patient was then referred to our hospital for further treatment.

Biochemical blood testing revealed slightly decreased hemoglobin and hematocrit levels of 9.9 g/dl and 31.1%, respectively, and a slightly increased white blood cell count (8,800/μl), platelet count ($55.6\times10^4/\mu$l), and C-reactive protein level (0.6 mg/dl). Other test results were within the reference ranges. The serum levels of the tumor markers carcinoembryonic antigen and cancer antigen 19-9 were also within the reference ranges. Urinalysis revealed high urinary protein (3+), a red blood cell count of 20 to 30/hpf, and a white blood cell count of 20 to 30/hpf. Two urine cytology tests were negative.

Figure 1.



The tumor surface is covered with tissue and mucus, while the internal portion is uniformly white.

Cystoscopy revealed a tumor with a surface that was covered with tissue and mucus (Figure 1). The internal portion of the tumor was uniformly white. Ultrasonography (Figure 2) revealed a lobulated polypoid mass measuring 4.5 cm in diameter that protruded from the left bladder wall into the lumen. The tumor margin had high echogenicity, whereas the internal portion had low echogenicity. Wall thickening and protrusion of the tumor into the lumen were observed from the left bladder wall (where the tumorous mass was attached) to the dorsal wall. Continuous blood flow was confirmed from the wall to the center of the tumor. Magnetic resonance imaging (MRI) (Figure 3) confirmed a pedunculated lobulated tumor protruding from the posterior left bladder wall into the lumen.

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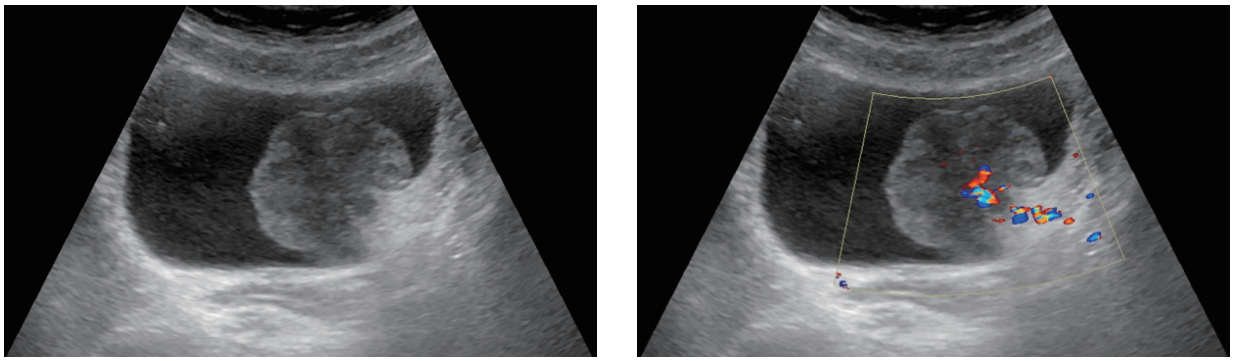
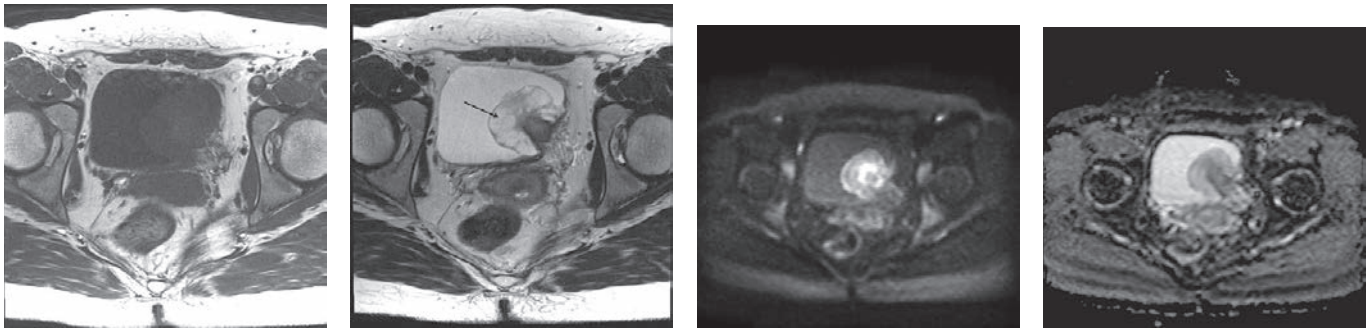
Figure 2.



A lobulated polypoid mass protrudes from the left bladder wall into the lumen. Wall thickening and protrusion of the tumor into the lumen are observed from the left bladder wall, where the tumorous mass is attached, to the dorsal wall. The posterior wall at the site of tumor attachment is thickened. There is a continuous blood flow signal from the wall to the central tumor portion.

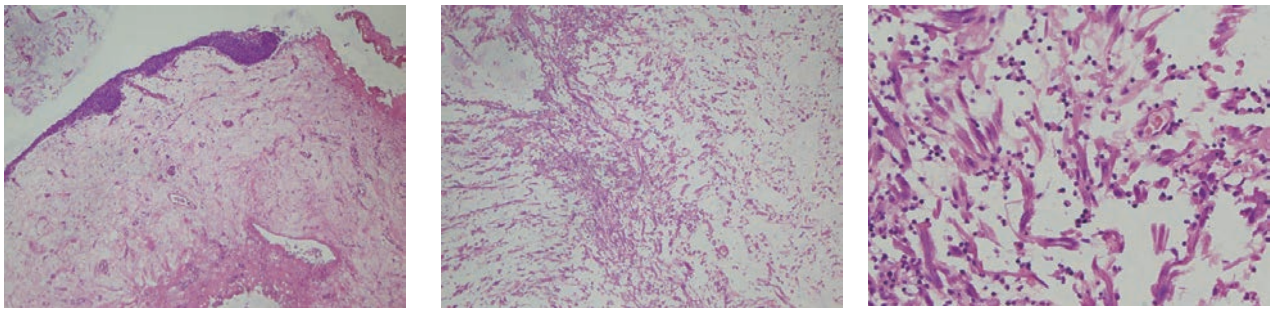
Figure 3.



A B C D

Axial (A) T1-weighted, (B) T2-weighted, (C) diffusion-weighted, and (D) apparent diffusion coefficient magnetic resonance imaging of the pelvis confirms a submucosal, pedunculated, lobulated mass in the posterior left bladder wall. The tumor is protruding into the lumen. The peduncle has a morphology of a submucosal mass. (← : nodular fasciitis pattern on T2-weighted images).

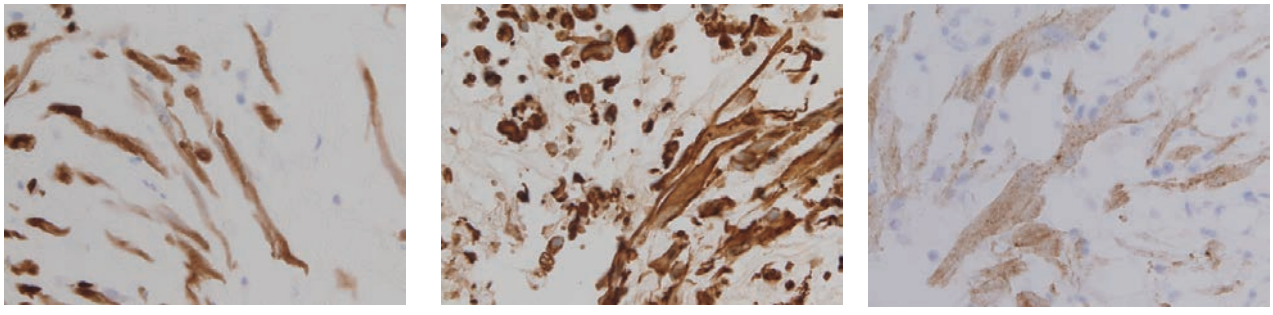
Figure 4.



A B C

A myxoid stroma with few atypical cells in the urothelial subcutaneous layer is observed. Multiple spindle-shaped cells with lymphocytic invasion are observed by hematoxylin and eosin staining (A) 20 × , (B) 40 × , and (C) 100 × .

Figure 5.



A B C

Immunostaining is positive for cytokeratin (AE1/AE3), vimentin, and ALK-1 staining (400 ×).

The lobulated area exhibited a low-intensity signal on T1-weighted imaging (T1WI) and a high-intensity signal similar to that of urine on T2-weighted imaging (T2WI). The morphology of the peduncle was similar to that of a submucosal mass, with a faint low-intensity signal on T1WI and a faint high-intensity signal on T2WI. Diffusion-weighted imaging revealed some areas in the central portion of the tumor with high-intensity signals; the apparent diffusion coefficients were slightly decreased.

A definitive diagnosis was difficult to achieve based on the above imaging findings; therefore, transurethral resection of the bladder tumor was performed.

Pathological examination revealed a notably myxoid stroma with few abnormal morphologies of the urothelial subcutaneous stroma. There were multiple spindle-shaped mononuclear cells with amphophilic cytoplasm, swollen nuclei, irregular shapes, and lymphocyte invasion (Figure 4). Immunostaining of the spindle cells was positive for AE1/AE3, vimentin, myogenin, and activin-like kinase 1 (Figure 5) and slightly positive in some areas for desmin, smooth muscle actin, and calponin. Immunostaining results for p63, GATA binding protein 3, and uroplakin III were negative (Table 1). Based on these findings, a diagnosis of IMT was made. Postoperative MRI showed a possible residual tumor, for which additional excision was proposed. However, the patient requested observation and follow-up.

Table 1. Immunostaining of spindle cells

AE1/AE3	++	Vimentin	++
Myogenin	++	ALK-1	++
Desmin	+	SMA	+
Calponin	+	p63	−
GATA3	−	Uroplakin III	−

++: positive, +: slightly positive, − : negative

Discussion

The term IPT was first used by Umikar and Iverson[2] in 1954 to describe a tumorous lesion in the lung. In 1990, Pettinato et al.[3] first reported and defined a neoplastic and myofibroblastic lesion as an IMT. Thus, IMT as a group of IPTs is a relatively new concept. An IMT is classified as a rare benign/malignant or intermediate type of tumor that may cause distant metastasis.[1] However, IPT is not uniformly observed in all organs. For example, IPT of the lower urinary tract most commonly occurs in the bladder and is referred to one the following: reactive pseudosarcomatous response, IPT, pseudosarcomatous fibromyxoid tumor, nodular fasciitis, pseudomalignant spindle cell proliferation, or pseudosarcomatous myofibroblastic proliferation. Currently, there is no consensus regarding whether IMT is neoplastic or non-neoplastic.

Images of an IMT may show a lobulated solid tumor mass; however, IMTs have no specific findings. Furthermore, high fluorodeoxyglucose uptake occurs on positron emission tomography/computed tomography; therefore, differentiation of IMT from a malignant tumor is difficult. Additionally, because no specific findings on blood examination allow for a definitive diagnosis, the aforementioned histopathological diagnosis is considered necessary. IMT of the bladder infiltrates the muscularis propria and surrounding tissues with possible tumor necrosis.[4-7] Some characteristic findings may confirm this malignancy, such as the submucosal morphology of the base and formation of a lobulated mass that protrudes into the bladder lumen, as in the present case. In addition, MRI findings of a mass with accompanying wall thickening and a notable submucosal lesion are characteristic of IMT of the bladder.[8-10] An important differential diagnosis in patients with bladder cancer is an epithelial tumor, which often originates from the mucous membrane of the bladder and progresses to the submucosal and muscularis propria. Therefore, the presence of lesions progressing from the muscularis propria to the submucosa or the presence of a submucosal lesion that is more notable than a lesion of the mucous membrane that protrudes into the bladder lumen may be an important imaging finding for the differential diagnosis of IMT.

In our patient, the lobulated area that protruded into the lumen exhibited a markedly high-intensity signal on T2WI. This signal intensity level was the same as that noted for urine in the bladder and was considered to be of a higher intensity than that for general soft tumors and bladder cancer. This may have occurred because the lobulated area was rich with myxoid stroma. Only one case in the literature exhibited the same findings[10]; other reported cases exhibited high-intensity signals that were typical of soft tissue tumors. In our patient, the base and neck areas of the tumor also exhibited signals that were similar to those of typical soft tissue tumors, which was consistent with previously reported cases.[8,9] These variations in imaging patterns may be the result of different tissue types and growth morphologies of IMTs. Similar to IMTs of other sites, IMT of the lower urinary tract presents with the following three growth morphologies: a nodular fasciitis pattern, a fibrohistiocytoma pattern, and a desmoid scar pattern. The proportion of each pattern differs according to the tumor and site.[4] The nodular fasciitis pattern is characterized by the proliferation of tumor cells within a rich mucus stroma. Furthermore, interstitial edema is reportedly the most prominent characteristic of IMT of the lower urinary tract.[5,6] In our patient, the lobulated area with markedly high-intensity signals on T2WI corresponded with this finding as confirmed by pathologic examination of the surgical specimen, which was mainly taken from the lobular area. Furthermore, notable myxoid stroma of the urothelial subcutaneous layer was observed, consistent with the imaging findings. Therefore, a markedly high-intensity signal on T2WI is important for the differential diagnosis of IMT of the urinary tract with a nodular fasciitis pattern in the main body.

In summary, we report a rare case of IMT of the bladder. Differential diagnosis of a neoplastic lesion is considered important for IMT of the lower urinary tract. On MRI, a submucosal lesion is more notable than a mucous membrane lesion that protrudes into the bladder lumen, and areas of the nodular fasciitis pattern with markedly high-intensity signals on T2WI are considered to be important for the differential diagnosis of IMT.

Conflict of interest

The authors certify and declare that no part of the research presented has been funded by any industry sources and that there is no conflict of interest.

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