Case Report

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Imaging and Clinical Findings of Primary Malignant Fibrous Histiocytoma of the Urinary Bladder: A Case Report

방광의 일차 악성 섬유성 조직구종의 영상 및 임상 소견: 증례 보고

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Primary malignant fibrous histiocytoma (MFH) is a malignant tumor of mesenchymal origin that rarely occurs in the urinary tract, particularly in the urinary bladder. Unlike urothelial carcinoma, which accounts for most bladder cancers, it occurs in the submucosal portion of the bladder wall and consists of the lamina propria, muscularis propria, and adventitia. It is presumed to originate from poorly differentiated pluripotent mesenchymal cells in which fibroblasts and histiocytes are partially differentiated. Radiologically, it is known as the "non-papillary tumor" and is commonly diagnosed as a large mass without necrosis, which shows invasion beyond the muscularis propia. Although the prognosis of this rare malignancy depends on pathological parameters, it generally has a poor prognosis with high local tumor recurrence. Here, we present a case of primary MFH in the urinary bladder with clinical symptoms of lower abdominal pain without gross hematuria that recurred rapidly and showed an aggressive disease course.

Index terms Malignant Fibrous Histiocytoma; Urinary Bladder; Magnetic Resonance Imaging

INTRODUCTION

Primary malignant fibrous histiocytoma (MFH) is a rare mesenchymal-derived sarcoma often found in the deeper layers of the urinary bladder wall, beyond the uroepithelium. These layers consist of the lamina propria, muscularis propria (de-

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trusor muscle), and adventitia. Histologically, they are presumed to originate from poorly differentiated pluripotent mesenchymal cells in which fibroblasts and histiocytes are partially differentiated (1, 2). MFH has been reported to have a very low prevalence but is the most common soft tissue sarcoma in adults. There are only a few reports of primary MFH of the urinary bladder in previous international literature. Unlike patients with urothelial carcinoma, who mainly complain of painless hematuria, patients with MFH may complain of nonspecific abdominal pain and a palpable mass without hematuria due to compression of the surrounding organs, making it difficult to diagnose (3). Understanding this rare malignancy can help clinicians and radiologists make appropriate diagnoses and prevent delays in diagnosis and treatment. Herein, we present a case of primary MFH of the urinary bladder with abdominal pain and a nonpapillary mass on cystoscopy and magnetic resonance imaging without lymph node metastasis at the time of diagnosis, which recurred rapidly and showed a very aggressive course.

CASE REPORT

A 58-year-old male with lower abdominal pain presented to the outpatient clinic of the Department of Urology with lower abdominal pain. He had no remarkable medical history and no other symptoms such as gross hematuria. A 3D urography CT scan showed about 3.4 cm × 2.8 cm × 3.0 cm sized heterogeneously enhancing irregular-shaped solid mass in the right anterolateral dome of the urinary bladder. Perivesical fat infiltration was observed in the anterior aspect of the mass (Fig. 1A). There was no remarkable intraabdominal lymphadenopathy suggestive of metastasis. No other enhancing lesions were observed, suggesting a multiplicity of lesions in both the urinary tract and urinary bladder. Pelvic dynamic contrast-enhanced MRI revealed that the mass in the urinary bladder dome showed intermediate signal intensity on T1- and T2-weighted images, with perivesical fat infiltration, marked diffusion restriction, and heterogeneous enhancement (Fig. 1B, C). There was no evidence of invasion into adjacent structures such as the prostate gland or pelvic wall. It has been suggested that tumors with relatively intact overlying mucosa may have the potential to be intramural. Furthermore, it does not show enhanced papillary projections to the mucosal side, which is considered different from urothelial carcinoma. Various types of mesenchymal tumors have been considered for the differential diagnosis, with leiomyomas and paragangliomas being representative examples. However, leiomyomas typically exhibit a characteristic low signal intensity on T2-weighted image (T2WI) without diffusion restriction, which diminishes the likelihood of diagnosis. In contrast, paragangliomas tend to present with intermediate signal intensity on T2WI, along with diffusion restriction and avid enhancement, necessitating a differential diagnosis. In this case, the presence of perivesical fat infiltration and locally aggressive features raised concerns regarding malignant mesenchymal tumors. Diagnostic cystoscopy revealed a solid mass protruding into the lumen of the urinary bladder with no papillary appearance and relatively intact overlying mucosa (Fig. 1D). Subsequent transurethral resection of the bladder was performed, and the mass was pathologically diagnosed as MFH. Three weeks later, a robot-assisted laparoscopic partial cystectomy was performed on the bladder mass. On postoperative histopathological examination, gross examination revealed an ulcerated infiltrating

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Fig. 1. Primary malignant fibrous histiocytoma of the urinary bladder in a 58-year-old male patient complaining of lower abdominal pain.
A. Axial and sagittal contrast-enhanced abdomen and pelvis CT images show about 3.4 cm × 2.8 cm × 3.0 cm sized heterogeneously enhancing solid mass (*) in the right anterolateral portion of the urinary bladder dome with suspicious perivesical fat infiltrations at the anterior aspect of the mass (arrows).
B. Axial and sagittal T2-weighted images show an intermediate signal intensity mass (*) in the right urinary bladder dome with perivesical tumor infiltration (arrow).
C. Sagittal diffusion-weighted image (b-value, 1000 s/mm^2) (left) shows marked diffusion restriction of the mass. Sagittal contrast-enhanced fat-suppressed T1-weighted image (right) shows heterogeneous enhancement of the mass.

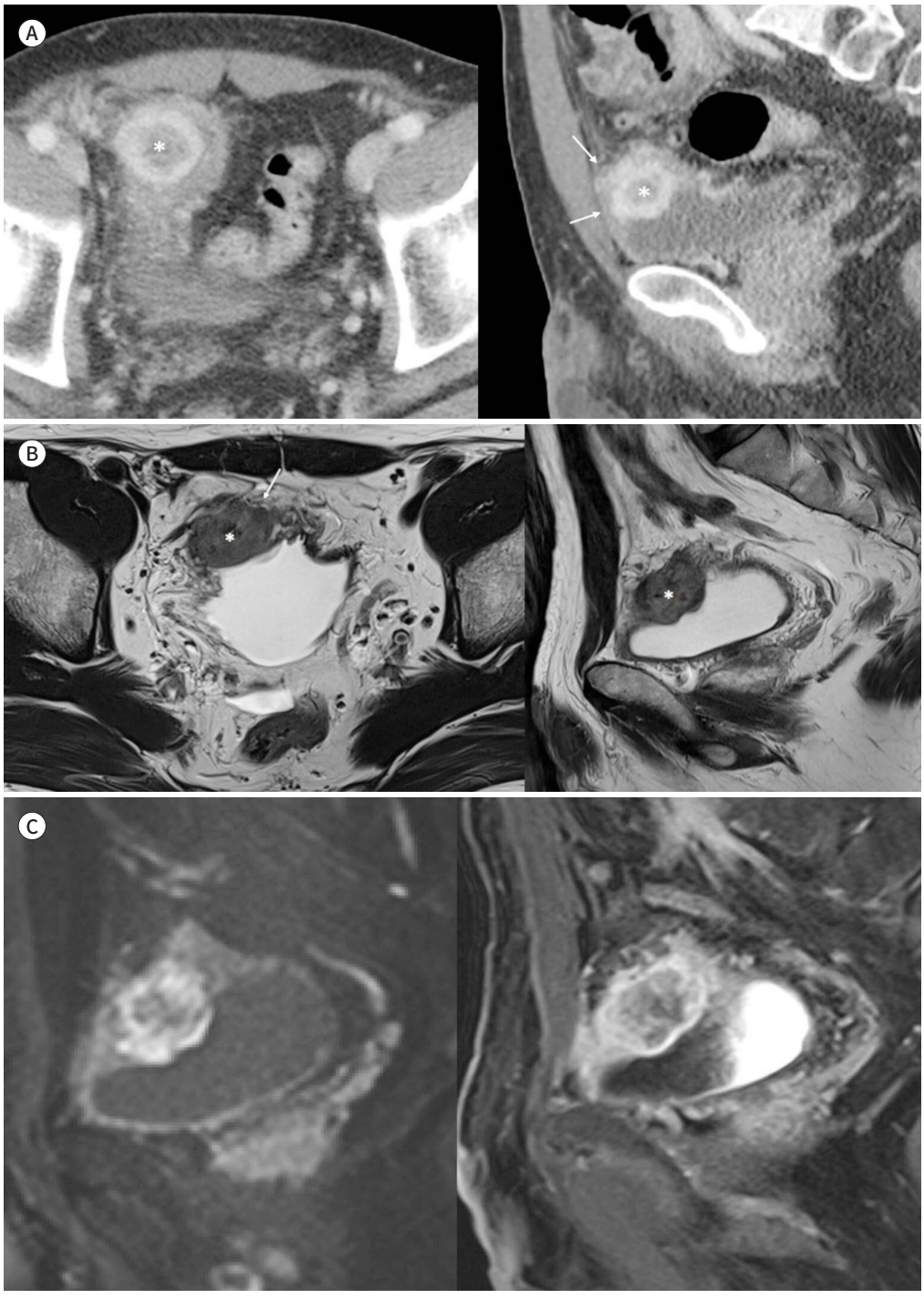
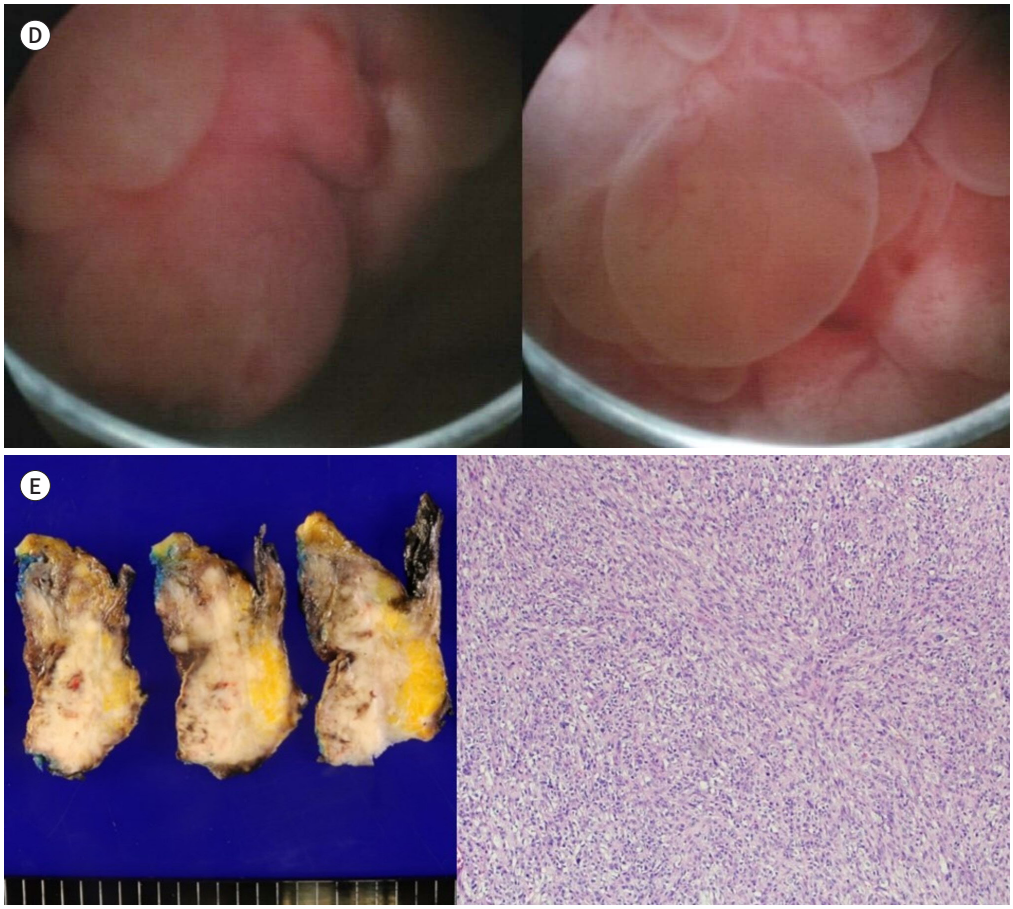


Fig. 1. Primary malignant fibrous histiocytoma of the urinary bladder in a 58-year-old male patient complaining of lower abdominal pain.
D. A non-papillary mass protruding into the lumen of the urinary bladder with an intact overlying mucosa was identified using conventional cystoscopy.
E. Gross specimen cut sections (left) show a gray-white firm mass with infiltrating margins and a low-power field microscopic image (right) reveals high cellular proliferation of spindle cells in a storiform pattern (hematoxylin & eosin stain, × 100).



white-gray firm tumor tissue involving the entire wall. While there was no evident involvement of the perivesical fat tissue on gross examination, microscopic involvement was observed. A low-power view revealed high-cellular proliferation of spindle cells in a storiform pattern (hematoxylin and eosin stain, × 100) (Fig. 1E). Immunohistochemical analysis revealed positive results for Vimentin, α1-antichymotrypsin, and CD68 but negative for S-100, Desmin, CD34, CD31, and CD117. The mass was finally confirmed to be an MFH and pathologically staged as T3a N0 (TNM stage by AJCC, 8th ed) (4).

A ^{18}F-FDG PET/CT scan performed a week post-surgery revealed the emergence of metastasis in the right external iliac lymph node. Contrast-enhanced abdominal and pelvic CT tomography performed 1 month after surgery showed multiple external and internal iliac lymph node metastases and heterogeneously enhancing nodules at the partial cystectomy site, perivesical space, anterior peritoneum, and mesentery, suggesting local tumor recurrence and metastatic seeding nodules. The treatment plan was changed from adjuvant radiation therapy

to palliative chemotherapy (AIM regimen: Doxorubicin, Ifosfamide, Mesna) based on the CT scan findings. The patient expired 4 months later after relapse despite palliative chemotherapy.

This retrospective study was approved by the Institutional Review Board of Soonchunhyang University Seoul Hospital (IRB No. 2022-08-008) and the requirement for informed consent was waived.

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DISCUSSION

This is a rare case of primary MFH of the urinary bladder that presented with non-specific symptoms, an advanced tumor stage at diagnosis, and an aggressive clinical course despite surgery and subsequent chemotherapy.

To the best of our knowledge, only a few cases of primary MFH arising in the urinary bladder have been reported. Previous studies have reported a total of 33 cases up until 2020 (5-7). We summarized the clinical and radiological features of bladder MFHs described in previous case reports, mainly in cases in which the radiological findings were well described (Supplementary Table 1 in the online-only Data Supplement).

At the time of diagnosis, the clinical manifestations of bladder MFH included hematuria in 81% of cases, and abdominal pain and/or urinary symptoms in 43% of cases (2). Although hematuria is a classic symptom of bladder cancer, MFH may present only with symptoms of abdominal pain, as in our case, which is thought to be related to the fact that MFH is a tumor of mesenchymal origin. Urothelial carcinoma arises from urothelial cells that line the urinary bladder, whereas MFH arises from mesenchymal cells such as fibroblasts and histiocytes in the submucosal layer of the bladder wall (7). It typically appears as a high-grade tumor containing multinucleated giant cells with high cellularity, nuclear pleomorphism, and abundant atypical mitoses (8).

Bladder neoplasms are broadly divided into either epithelial or nonepithelial (mesenchymal) neoplasms, 95% being epithelial. Neoplasms derived from mesenchymal tissues include benign tumors, such as leiomyomas, paragangliomas, fibromas, and solitary fibrous tumors, and malignant tumors, such as rhabdomyosarcoma, leiomyosarcoma, lymphoma, and MFH (8). There is a significant overlap in radiological findings, which makes diagnosis difficult. Nonetheless, epithelial neoplasms tend to manifest as irregular masses with internal filling defects within the urinary bladder and papillary projections to the mucosal side. This is because they originate from the superficial layer of the bladder wall. By contrast, mesenchymal tumors are located in the submucosal portion of the bladder wall and typically present as relatively smooth intramural lesions. These differences in imaging characteristics correspond to the underlying histological variances and cellular origins. However, these distinctions can vary based on whether the lesion is benign or malignant as well as the degree of cellularity of the tumor.

MFH of the urinary bladder can present as variable features on imaging. It may appear as a heterogeneously enhancing mass with ill-defined margins, areas of necrosis, or infiltration into the surrounding tissues. In addition, it may be associated with an irregular or thickened

bladder wall, similar to urothelial carcinoma. However, unlike urothelial carcinoma, which is often diagnosed when it is small owing to symptoms of hematuria, MFH is most often a large mass at the time of diagnosis. MFH tends to present as a large mass without central necrosis (9) and is characterized as a "non-papillary tumor" that tends to invade the deep muscular layer of the bladder wall, with 83% of cases known to be at T3 or higher stages (5). Generally, cancers derived from mesenchymal cells tend to be more diffuse in nature, whereas urothelial carcinomas tend to be more localized (9). However, imaging findings of MFH and other urinary bladder neoplasms can overlap, making them difficult to distinguish using imaging alone. In this case, we considered a primary diagnosis of a malignant mesenchymal tumor rather than urothelial carcinoma. When considering the distinguishing points between these two diseases, urothelial carcinoma typically shows enhanced papillary projection to the mucosal side, usually originates from the bladder base (80% at initial diagnosis), and presents synchronous and metachronous tumors in 30% of cases (8). Other potential differential diagnoses for malignant non-epithelial neoplasms include leiomyosarcoma, rhabdomyosarcoma, and lymphoma. Leiomyosarcoma, the most prevalent non-epithelial malignancy, typically presents as a sizable mass with evident intratumoral necrosis. Rhabdomyosarcoma manifests as a typical grape-like intraluminal mass, primarily affecting individuals under 10 years of age. Lymphoma, which is more common in middle-aged women, exhibits a characteristic homogeneous enhancement, aiding in differentiation (8, 9). Ultimately, a definitive diagnosis usually requires histological evaluation and immunochemical analysis (2). MFH is generally recognized as aggressive, with a high recurrence rate and poor prognosis. In our case, although prompt surgery and subsequent treatments were performed after diagnosis, rapid progression and recurrence of the cancer eventually led to death. Although rare, knowledge of primary MFH of the urinary bladder with nonspecific symptoms, an aggressive clinical course, and prompt treatment is essential.

Supplementary Materials

The online-only Data Supplement is available with this article at http://doi.org/10.3348/jksr.2023.0083.

Author Contributions

Conceptualization, L.E.J., K.J.H.; data curation, L.Y.J., H.S.S.; formal analysis, L.Y.J., H.J.; investigation, L.E.J.; methodology, C.Y.; resources, K.J.H.; software, L.Y.J.; supervision, L.E.J.; visualization, L.Y.J., J.S.Y.; writing—original draft, L.Y.J.; writing—review & editing, L.E.J., K.J.H.

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Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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방광의 일차 악성 섬유성 조직구종의 영상 및 임상 소견: 증례 보고

이윤정[1] · 이은지[1*] · 김재헌[2] · 진소영[3] · 홍성숙[1] · 황지영[1] · 장윤우[1]

원발성 악성 섬유성 조직구종은 요로, 특히 방광에서 흔하게 발생하지 않는 중간엽 유래의 악성 종양이다. 방광암의 대부분을 차지하는 요로 상피암과는 달리, 악성 섬유성 조직구종은 점막 고유층, 고유근층, 장막층으로 구성된 방광 벽의 요로 상피 하부에서 발생한다. 조직학적 기원은 섬유아세포와 조직구 세포가 부분적으로 분화된 저분화 중간엽 줄기세포에서 발생하는 것으로 추정된다. 영상의학적으로는 요로 상피암에서 흔히 볼 수 있는 유두상 성장 패턴을 보이지 않아 '비유두상 종양'으로 알려져 있으며, 흔히 고유근층 이상의 침범을 보이고 괴사를 동반하지 않는 거대 종괴의 형태로 진단된다. 이 드문 악성 종양의 예후는 다양한 병리학적 인자에 의해 결정되지만, 일반적으로 불량한 예후와 높은 국소 재발률을 보인다. 또한 무통성 혈뇨를 주로 호소하는 요로 상피암 환자와 달리 주변 장기를 압박하여 비특이적인 하복부 통증이 나타날 수 있어 진단에 어려움이 있을 수 있다. 저자들은 혈뇨 없이 하복부 통증을 주 증상으로 내원하여 방광의 원발성 악성 섬유성 조직구종으로 진단된 후 빠른 치료를 시작하였음에도 매우 공격적인 임상 경과를 보였던 증례를 보고하고자 한다.

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