Case Report

Malignant solitary fibrous tumor of the urinary bladder

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Abstract

A 67-year-old man was examined for persistent pain over his lower abdomen and was found to have a large pelvic tumor. During surgery, we detected a 16 × 9 × 9 cm³ urinary bladder tumor with small intestinal adhesions, and performed partial cystectomy and segmental resection. Histological and immunohistochemical examinations established the diagnosis of malignant solitary fibrous tumor (SFT). Only 10 cases of urinary bladder SFT have been reported in the English literature; our patient is the second one with malignancy and had a longer follow-up period than the other case.

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1. Introduction

Solitary fibrous tumor (SFT) is a rare type of mesenchymal tumor. In the past, SFT was thought to be a pleural-based lesion; however, it has recently been reported to occur in many extrapleural sites with no mesothelial lining.1–5 SFTs are usually benign and slow growing, but malignant SFTs have been described to have a greater potential of recurrence or metastasis. Extrapleural SFTs of the urinary bladder are extremely rare, and only 10 cases have been reported in the English literature.6 Of these cases, only one showed malignant pathological feature.3 In this study, we present the second case of a patient with a malignant SFT.

2. Case report

A 67-year-old man had noticed a palpable mass in his right lower abdomen for 2 years. One month prior to admission, he felt pain during exercise and visited a hospital. Physical examination showed a movable firm mass over the right lower abdomen. Urinary analysis revealed microscopic hematuria, and urinary cytology was negative for urothelial carcinoma. The tumor markers including prostate specific antigen (PSA), alpha-fetoprotein (AFP), carbohydrate antigen (CA)-125, CA-153, carcinoembryonic antigen (CEA), and CA-19-9 were all within normal limit. Abdominal magnetic resonance imaging (MRI) showed a large soft-tissue mass, with the highest diameter being 16 cm, arising from the dome of the urinary bladder (Fig. 1). During surgery, we found that the tumor was adhered firmly to the small intestine. In addition, multiple engorged vessels extended from the urinary bladder to the tumor. We performed partial cystectomy and segmental resection of the intestine. The postoperative recovery course was uneventful. The patient has been well after an 18-month follow-up.

The mass was an encapsulated gray-white tumor, measuring 16 × 9 × 9 cm³ (Fig. 2). The transection was white and multinodular, with a 4 × 4 × 3 cm³ necrotic defect. Microscopic examination revealed that the tumor was composed of ovoid- to spindle-shaped cells. The tumor showed merged hyper- and hypocellular zones in short fascicular arrangement, and spiral to random patterns (patternless pattern). We observed bundles of collagen and sclerosing stromal cells scattered between tumor cells. We also observed branching and hemangiopericytoma-like...
vasculature with perivascular hyalinization. Necrotic areas of the tumor, mitotic activity $>4$ mitoses/10 high-power field (HPF) (Fig. 3A), and cellular atypia in local region indicated the malignancy of the tumor. Immunohistochemical examination revealed that the tumor cells were strongly positive for CD34 (Fig. 3B) and CD99, but negative for CD117, smooth muscle actin, and AE1/AE3.

3. Discussion

Ten cases of SFT of the urinary bladder have been reported in the English literature, and only one of them was malignant (Table 1). The long-term outcome of that malignant tumor is unknown. Tumor most often occurs in 50–60-year-old adults, and its common clinical manifestations are pain and palpable mass. Hematuria and dysuria are less common, but have also been reported. Symptomatic hypoglycemia is associated with the secretion of insulin-like growth factor-2 and was rarely observed in patients with SFTs of the urinary bladder.

Appearance of SFTs does not have a specific pattern. It often appears as a well-circumscribed, highly vascular mass without any invasion of nearby structures. MRI revealed that the mass was often isointense or hypointense on T1-weighted images (T1WI) and heterogeneous on T2WI. The heterogeneous appearance may be attributed to variation in cellular and collagenous content. Very low T2 signal regions may be observed and were attributed to the presence of interstitium rich with collagen and fibrotic tissue. In our case, iso-intensity of the mass in T1WI and heterogeneity in both T1WI and T2WI were consistent with the typical findings of SFTs. The tumor was well enhanced after contrast injection, which also fits with the hypervascular nature of the SFT. The central necrosis pattern observed in our case also indicated malignancy.

Histologically, SFTs are characterized by a branching hemangiopericytoma-like vasculature, usually with perivascular hyalinization. Spindle-shaped, monomorphic fibroblasts between collagen bundles are a common presentation. Variation in cellularity, with random distribution of hyper-
hypocellular zones, is commonly referred to as patternless architecture. Mitosis and necrosis are relatively rare in SFTs. The criteria for malignancy include large tumor size (>10 cm), hypercellularity, nuclear pleomorphism, tumor necrosis, >4 mitoses/10 HPF, and infiltrative margins. However, SFTs can have variable morphologies. Therefore, immunohistochemical staining plays an important role in the diagnosis. The tumor cells are often positive for CD34, CD99, and bcl-2, but they are negative for S-100 protein, actin, desmin, and keratin. We observed diffuse expression of CD34 and CD99 in our patient.

Gastrointestinal stromal tumor (GIST) is another condition considered in differential diagnosis, because the histological appearance of GIST is similar to that of SFTs, and GIST may also be positive for CD34 and bcl-2. However, GIST is often positive for CD117. Smooth muscle tumor cells have cigar-shaped nuclei, perinuclear vacuoles, and eosinophilic cytoplasm. While some smooth muscle tumor cells are positive for CD34, almost all cells were positive for smooth muscle actin. The tumor cells were negative for cytokeratin AE1/AE3 antigen, which is a specific marker for epithelium-derived tumor; this finding ruled out the possibility of a carcinoma. The spindle-shaped cells of the tumor originating from the nerve sheath have a special comma-shaped nucleus, and these cells are negative for CD34 and bcl-2 and positive for S-100 protein. We diagnosed SFT on the basis of the results of morphological and immunohistochemical examinations. Further, this tumor fits the malignancy criteria: large tumor size (16 cm), high cellularity, pleomorphism, six mitoses/10 HPF and up to three mitoses in an HPF (>4/10 HPF), necrosis, and infiltrative border in the muscle of urinary bladder.

Although extraperitoneal SFTs with benign histology can also show aggressive behavior, malignant SFTs account for higher rates of local recurrence, metastasis, and mortality. Complete resection of the tumor is the most recommended treatment if available, and it has a favorable long-term survival rate. Adjuvant radiation therapy can be considered in selected patients, but the efficacy of this treatment remains controversial. In contrast, patients with unresectable, recurrent, or metastatic SFT have a worse outcome. Radiation therapy can be administered to these patients as an adjuvant therapy or a primary therapy. Recently, antiangiogenic therapy has been considered to be a promising treatment for these patients, and, to date, sufficient evidence for systemic chemotherapy has not been obtained.

We performed partial cystectomy instead of radical cystectomy because the tumor could be completely resected with safe surgical margin and proper residual bladder volume. No adjuvant therapy was administered. The patient showed good recovery and did not show local recurrence on the CT image obtained after 18 months of follow-up. Thus, surgery seems to be an appropriate treatment modality for malignant SFT. However, more clinical data and longer follow-ups are required to evaluate the outcome of the disease.

**References**


