

Extragastrointestinal Stromal Tumor of the Urinary Bladder

A Case Report

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INTRODUCTION

A gastrointestinal stromal tumor (GIST) is primarily located in the gastrointestinal tract. It is classified as a mesenchymal tumor that shows a CD117 (c-KIT)-positive mesenchymal spindle or epithelioid neoplasm. Tumors outside the gastrointestinal tract that were histologically similar to GISTs and showed immunopositivity for CD117 were classified as extragastrointestinal stromal tumors. Although there were several reports of tumors outside the gastrointestinal tract, only a case of extragastrointestinal stromal tumor in the urinary bladder was reported.^(1,2) Herein, we report another case arising in the urinary bladder presenting with submucosal polypoid mass.

polypoid mass measuring 2.6 × 2.4 cm and multiple small calcified spots in the bladder base (Figure 1). No evidence of an intra or extraluminal mass was observed in the gastrointestinal tract.

The patient was taken to the operating room for a cystoscopic examination. A thumb tip-adult size polypoid submucosal mass with a smooth surface mucosa and a broad base was detected in the bladder dome. Thereafter, the mass was cystoscopically removed.

The cystoscopically resected specimen consisted of multiple chips of yellow, pink, and gray, soft rubbery tissue, measuring 3.0 × 2.0 × 2.0 cm in aggregate. Tumor cells were found to be scattered in

CASE REPORT

A 42-year-old man presented with hematuria since three weeks ago. He had no irritative or obstructive urinary symptoms. Urine analysis was normal and other laboratory studies were within normal limits.

Computed tomography scan revealed an intravesical solid



Figure 1. Computed tomography scan revealed an intravesical polypoid mass with small multiple calcified spots within the contrast-filled urinary bladder lumen.

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the submucosa, and the overlying transitional epithelium was intact without ulceration. It was composed of sheets or nests of eosinophilic cells without any organoid features and loose edematous well-vascularized stroma, and with varied cellularity (Figure 2). In some areas, the tumor cells proliferated along the abundant vascular structures, and were arranged in a perithelial pattern. Occasionally, individually-scattered tumor cells were noted in the edematous stroma. The tumor cells were chiefly composed of polygonal epithelioid cells and were rarely composed of little spindle cells. The nuclei were ovoid or round and often centrally located. Some had bland or hyperchromatic nuclei. Occasionally, intranuclear inclusion bodies were noted. The cytoplasm was bright plump eosinophilic with relatively distinct cytoplasmic margins; occasionally, irregular margins were observed. Furthermore, eosinophilic cytoplasm was not stained by periodic acid-Schiff staining. Neither mitosis nor necrosis was observed. The stroma had a loose edematous myxoid or a focally liquefied appearance with high vascularity.

Immunohistochemically, the tumor cells strongly expressed CD117 (Figure 3) and weakly expressed CD34, but they were negative for smooth muscle actin, pan-cytokeratin, and S-100 protein.

During 20-month follow-up, using transabdominal ultrasonography and cystoscopic examination, no recurrence of tumor growth was noted.

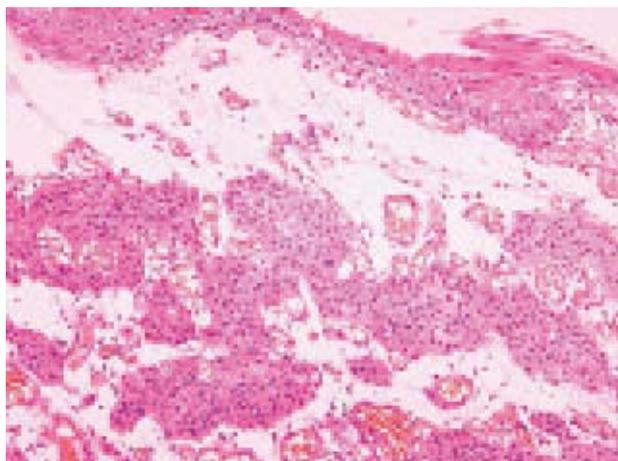


Figure 2. The tumor shows sheet or nests of polygonal epithelioid cells having bland or hyperchromatic nuclei and bright eosinophilic cytoplasm with relatively distinct cytoplasmic margins (hematoxylin and eosin stain $\times 100$).

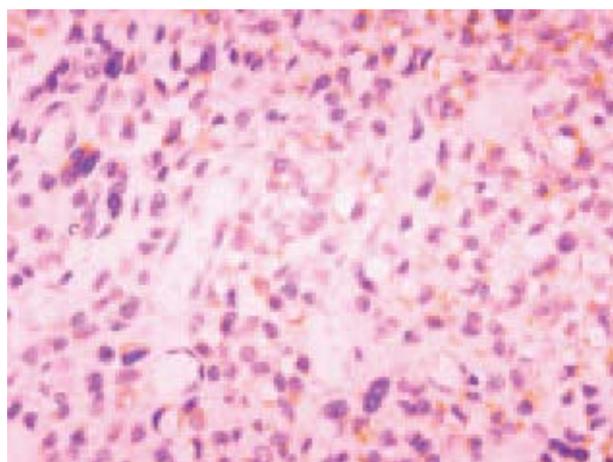


Figure 3. The stain-positive tumor cells for CD117 (Immunohistochemical stain, $\times 200$).

DISCUSSION

Most of the GISTs were previously classified as leiomyoma or neurogenic tumors because of similar histologic and immunophenotypic features of smooth muscle or neurogenic differentiation.⁽³⁾ Recently, however, they expressed a growth factor receptor with tyrosine kinase activity, known as KIT, which appeared to play a key role in committing primitive mesenchymal cells towards interstitial cells of Cajal differentiation.^(3,4) True neural and smooth muscle neoplasms of the gastrointestinal tract entirely lack KIT mutations. Some GISTs contain activating mutations in the KIT proto-oncogene, which appears to be a strong candidate for the molecular pathogenesis of GISTs.⁽⁵⁾ Therefore, KIT expression was proposed to be both the most sensitive and specific phenotypic marker of GISTs showing differentiation towards the interstitial cell of Cajal.⁽⁴⁾

Extragastrintestinal stromal tumors are relatively rare compared with their gastrointestinal counterpart. Approximately, 7% of the tumors occurred in the soft tissues of the peritoneum, omentum, and mesentery.⁽⁶⁾ Extragastrintestinal stromal tumors usually present during adult life in the form of enlarging masses with variable duration. Unlike their gastrointestinal counterparts, these lesions tend to be large when are first detected. Most of them are firm, fleshy, grey, and red masses with occasionally cystic change.

Histologically, there are two types; epithelioid and spindle cells. The former is noted where the tumor is composed of round cells with varying pleomorphic cell size and an eosinophilic or clear cytoplasm. The nuclei are uniform and round to ovoid, and have a vesicular chromatin. Occasionally, hyperchromatism, multinucleation, a prominent cytoplasmic vacuole, and a peripherally located nucleus are also seen. The epithelioid type was previously classified as leiomyoblastoma. The spindle pattern more closely resembles the conventional smooth muscle tumors, which are short and fusiform in contrast to the smooth muscle cells that show short fascicles and whorls, and their nuclei tend to be uniform and round.⁽³⁾

In addition to the fact that the cells were consistent positive for KIT (CD117), some GISTs showed immunopositivity for CD34 and S-100 protein and were rarely positive for smooth muscle actin and desmin.

The clinical behavior of an extragastrintestinal stromal tumor is difficult to predict. However, it is well-accepted that the malignant behavior of GISTs is strongly correlated with tumor size (> 5 cm in greatest diameter) and mitotic count (\geq 5 mitosis per 50 high-power field). Based on this rule, our case is benign. However, it should be noted that the lesions that are very small (even < 2 cm) and have low mitotic rates (even < 5 per 50 high-power field) occasionally metastasize.⁽³⁾ Therefore, a prolonged follow-up period is necessary for almost any GIST that exhibits the potential to behave in a malignant fashion.

It was known that there was no effective therapy except complete surgical resection until the recent development of the KIT inhibitor imatinib mesylate. This inhibitor attacks a specific molecular target in GISTs, by employing tyrosine kinase of KIT inhibition as well as disrupting signal transduction at mitosis. It induces growth arrest and apoptosis of tumor cells.⁽⁷⁾

CONFLICT OF INTEREST

None declared.

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