



A case of bladder perivascular epithelioid cell tumors

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ABSTRACT

Perivascular epithelioid cell tumor (PEComa) was introduced in the WHO classification of bone and soft tissue tumors in 2002, and Bladder PEComa is very rare. A 60-year-old man underwent TURBT after CT and cystoscopy revealed a 2.5 cm tumor with a concave center on the posterior wall of the bladder. Pathological examination revealed a perivascular epithelioid cell tumor, which was diagnosed as primary bladder PEComa after systemic examination. We report a case of partial bladder resection for bladder PEComa, a type of mesenchymal tumor that does not originate from the bladder mucosa.

1. Introduction

Bladder PEComa is very rare, with 49 cases of primary bladder PEComa reported worldwide. Generally, a PEComa has a benign course, though malignant transformation has been reported in a few cases. We report our experience with primary bladder PEComa, which are rarely reported worldwide, including a review of the literature.

2. Case presentation

In June 2018, A 60-year-old man tested positive for occult blood in urine during a physical examination. The patient was referred to a proximal urologist, who performed an ultrasound examination and noted an elevated lesion in the bladder. In July 2018, the patient visited our hospital and was admitted, after which CT (Fig. 1) and cystoscopy findings (Fig. 2) revealed a tumor with central neovascularization about 2.5 cm in size on the posterior wall of the bladder. As a result, a transurethral resection of bladder tumor (TURBT) procedure was performed. Pathology revealed tumor lesions from the submucosa to the muscularis layer on HE staining, and the tumor itself was weakly acidic and somewhat pale, with abundant cytoplasmic spindle-shaped cells. Immunostaining results showed AE1/AE3 CK, c-kit, S-100, CA9, CD34, and CD56 were negative, Ki-67 was positive in less than 1%, synaptophysin was positive in some cells, and HMB-45, alpha-SMA, and CD10 were positive in most cells (Fig. 3). Finally these findings established the

diagnosis of PEComa. Based on the pathological diagnosis, it was determined that an additional partial cystectomy would be necessary, which was performed in August 2018 (one month after the initial TURBT). Since then, the patient has been followed including periodic contrast-enhanced CT scanning, with no evidence of recurrence or metastasis at the latest examination performed in February 2022.

3. Discussion

PEComa was introduced in the WHO classification of bone and soft tissue tumors in 2002, while Bonetti et al. first presented histopathological findings of a mesenchymal liver tumor derived from perivascular cells in 1992. Most of these tumors are benign and the average age of affected patients is 45 years. Studies have noted that the disease occurs in various organs, such as the uterus and kidneys, as well as retroperitoneum, with soft tissues as the predilection site, and the most common examples are angiomyolipoma (AML) of the liver, kidneys, and other organs, while lymphangioleiomyomatosis and CCST are common manifestations in the lungs.¹

In pathological findings, they are considered to be mesenchymal tumors composed of perivascular cells that show a focal association with vessel walls, and co-express melanocyte-related markers and smooth muscle markers.

Most PEComa tumors are benign, though some cases have been reported to have a malignant course. In 2005, a tentative histological

Abbreviations: PEComa, Perivascular epithelioid cell tumors; CT, Computed tomography; TURBT, transurethral resection of bladder tumor; HE, Hematoxylin and eosin.

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Fig. 1. A 26 × 25 mm-sized mass was found at the center of the posterior wall of the bladder. That showed clear borders, well-defined margins, and a uniform internal surface with contrast effect.



Fig. 2. Cystoscopy shows the center of the posterior wall of the bladder.

grading system for PEComa was presented based on six criteria; (1) tumor diameter ≥ 5 cm, (2) infiltrative growth pattern, (3) high nuclear atypia, (4) fission imaging showing mitotic count $\geq 1/50$ HPF, (5) presence of necrotic findings, and (6) vascular invasion.² A PEComa tumor is considered to be benign when none of those are noted, while uncertain malignant potential is considered when one characteristic is found, and malignant when two or more are present. In our case, only vascular invasion was observed, thus we determined uncertain

malignant potential based on histological results.

49 cases of primary bladder PEComa were reported worldwide, with surgical treatment including TURBT in 21 cases, partial bladder resection in 19 cases, tumor enucleation in 2 cases, total cystectomy in 3 cases, and unknown in 4 cases.^{3–5} Among all reported cases, 3 (6.1%) patients developed local recurrence, 3 (6.1%) developed metastasis, and 1 (2.0%) died after metastasis appeared. In a report of 234 PEComa cases, the metastasis rate after surgery was 9.8% and the local recurrence rate after surgery was 6.4%.¹ The recurrence and metastatic rates after surgery for bladder PEComa are considered to be comparable to those of PEComa. Metastasis in the reported bladder PEComa cases included lung/brain metastasis (TURBT), large mesh/peritoneal dissemination (tumor enucleation), lung/bone to gallbladder metastasis (total cystectomy), and skeletal muscle metastasis (TURBT). Six of the seven patients with recurrence or metastasis (details unknown in one case) were classified with malignant potential lesions based on histology results.

Since a bladder PEComa, a type of mesenchymal tumor, does not originate from bladder mucosa, radical surgical treatment with a transurethral procedure alone is insufficient. However, radical cystectomy is unlikely to be necessary because PEComa is a type of mesenchymal tumor and not a multicentric, so-called “field-defect” tumor like conventional urothelial bladder cancer. In a previous case, an emergency partial bladder resection was performed due to difficulty with control of bleeding after transurethral resection of the tumor, though that is not appropriate from the viewpoint of safety for a PEComa, as it is rich in blood vessels. It is considered that a partial bladder resection should be performed as a reliable surgical treatment for primary bladder PEComa, especially in cases with histological findings indicating malignant potential.

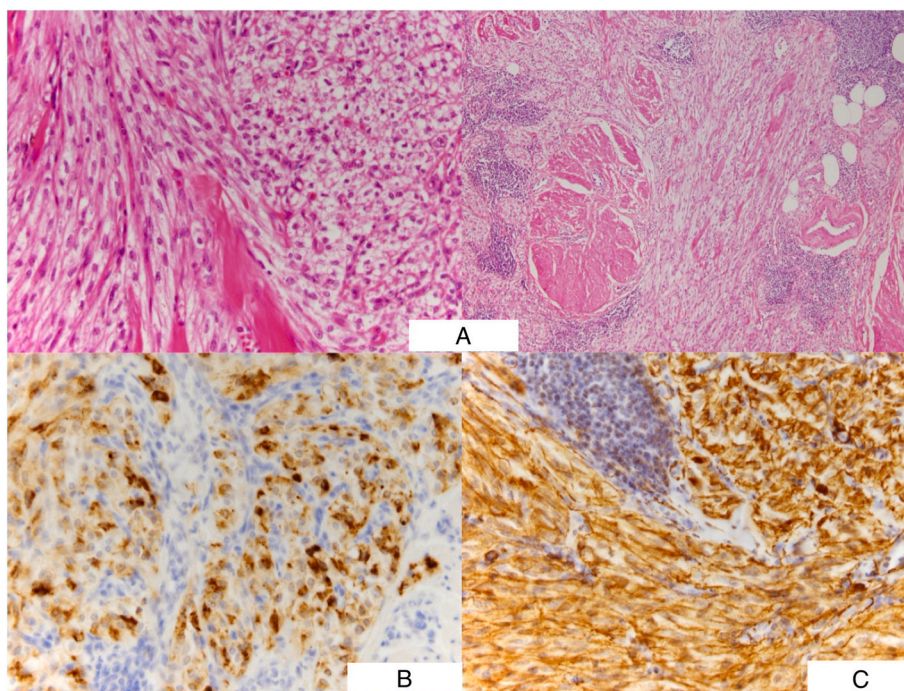


Fig. 3. (A) HE shows that vascular invasion was observed, (B) HMB45 positive, (C) αSMA positive.

4. Conclusion

This report of primary bladder PEComa brings the total number of presented cases to 50. Although rare, especially primary bladder PEComa, the number of reported cases has been increasing in recent years. Because of limited findings presented thus far, it is difficult to conclude regarding a precise treatment plan. Therefore, it is necessary to collect and analyze more cases in the future.

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Declaration of competing interest

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