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Case Report

Bladder PEComa: A case report and literature review

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ABSTRACT

The location of PEComa is rare in bladder, and the misdiagnosis rate is very high before operation. The characteristic of the bladder PEComa is that the CT is a round, elliptical or irregular cystic mass with a large volume, more than 5 cm or more, and the edge is clear. The solid part of the scanning tumor is obviously enhanced, and the rich blood supply artery and the drainage vein can be seen.

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Introduction

The knowledge of perivascular epithelioid cell tumor (PEComa) originated from 3 types of tumors: renal and hepatic angiomyolipoma, clear cell "sugar" tumor, lymphangiomyomatosis (LAM) [1]. In 1992, Bonetti et al [2] first defined PEComa as a tumor composed of cells with similar morphological and immune phenotypes, such as clear cell "sugar" tumor, angiomyolipoma, and LAM. 2002 World Health Organization Classification of Tumors: Pathology and Genetics of Tumors of Soft Tissue and Bone defined PEComas as a group of mesenchymal tumors. Immunohistochemistry has clear characteristics of perivascular epithelioid cells. PEComatous tumors were newly identified in 2015 World Health Organization Classification of Lung Tumors, including (a) lymphangiomyoma (LAM); (b) PEComa, benign (with clear cells); (c) PEComa, malignant. Relevant literature [3] summarized

the pathomorphological characteristics of PEComa as follows: (1) Tumor cells are arranged radially or sleeve-like around dilated blood vessels. (2) The tumor cells were epithelioid and could be divided into 3 types: the transparent cells with clear cytoplasm and vacuolated cytoplasm; the cytoplasm is eosinophilic with nucleated atypical eosinophilic cells. Both cytoplasm and nucleus were spindle cells, which were similar to smooth muscle cells. Each tumor may be composed of 3 of these cells in different proportions. Each tumor may be composed of the above 3 kinds of cells in different proportions. (3) Tumor interstitial rich in blood vessels, blood vessels for thin wall, occasionally for thick wall or hyaline degeneration. Characteristic expression of melanocyte markers and muscle cell markers are the immunohistochemical characteristics of PEComa, among which positive melanin markers represented by HMB45 and Mart-1 are of great significance for diagnosis, and HMB45 is the most sensitive [4]. Except for some lymphomas, Mum-1 is only expressed in some malignant melanoma and transparent cell sarcoma, and PEComas are stained with Mum-1 to differentiate melanoma from transparent cell sarcoma [5].

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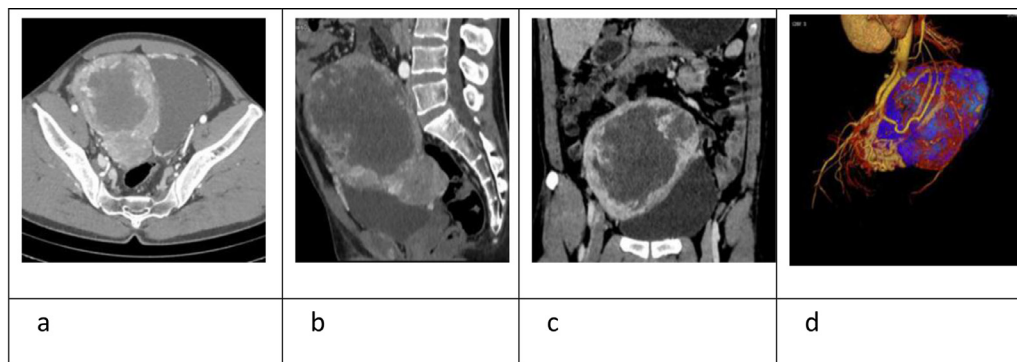


Fig. 1 – CT examination. 1a (axial arterial stage): a cystic solid mass with clear edges was observed in the lower abdomen and pelvic cavity. 1b (sagittal arterial stage): the tumor edge was significantly enhanced in the arterial stage, but there was no enhancement in the central low-density area, and the bladder was pushed to the left. 1c (coronary vein stage): the solid part at the edge of the tumor was further strengthened, and the right side of the bladder floor wall was invaded; 1d (angiography): the tumor was mainly supplied by the bilateral internal iliac artery, and increased and disordered venous drainage was also observed.

Case report

Clinical data: The patient, a 36-year-old male, presented dull pain in the lower abdomen accompanied by fatigue in the past 5 months, which lasted for several minutes at a time without relief, and showed no symptoms such as chills, fever, and nausea. No definite abnormality was found in the tumor markers.

Image data CT examination: Large mass in the lower abdomen and pelvis, about 18.8 cm × 12.1 cm × 13.6 cm in size, oval in shape, with cystic components accounting for 2 out of 3 of the tumor volume, uneven density on noncontrast scan, necrosis in the central area. The CT value in the noncontrast scan was about 27–44hu; the CT values in the arterial and venous phases were about 94–108HU and 96–113HU, respectively. The lesion was supplied by the branches of the bilateral internal iliac artery, with abundant internal vessels and multiple thickened and tortuously draining veins around it. The boundary between the lesion and the right side of the bladder floor and the right side of the ureter was not clear (Fig. 1 a–d). No definite enlarged lymph nodes were observed in the upper, middle, and lower abdomen and pelvic cavity. **Diagnostic opinion:** Lesion nature tend to be malignant in nature (gastrointestinal or reproductive system source) mesenchymal tumor may be.

What was seen during the operation was that the tumor was located in the pelvic cavity with unclear boundary with the right outer wall of the bladder bottom and the right ureter, showing adhesive changes with a size of 18cm×14cm×10cm and intact capsule. There is no ascites in the abdominal cavity, no tumor implantation or metastatic nodules in serosal surface and liver of the abdominal cavity, and no enlarged lymph nodes near the inferior mesenteric artery. Surgical removal of part of bladder and right ureter.

The gross pathological data showed that the tumor size was 16 cm × 14 cm × 8 cm, the cut surface was gray-yellow and gray-red, the capsule was solid, and there were some gray-red gelatin like substances in the capsule.

Pathological diagnosis: Spindle cell tumor with massive hemorrhage and infarction. HE and immunohistochemistry (Fig. 2a–d): ki-67 (+, about 3%), melan-a (–), hmb-45 (+), s-100 (–), SDHB (+), dog-1 (–), CD117 (–), CD34 (–), SMA (+), Actin (–), EMA (–), CK (–), Vim (+). Combined with HE and immunohistochemical results, PEComa was suggested.

Discussion

The location of PEComa is uncertain, kidney [6], liver [7], and uterus [8] are common. It is rare in bladder, and the misdiagnosis rate is very high before operation. Literature reports [9] bladder PEComa mainly occurs in young and middle-aged women, aged 19–58 years old, with a median age of 36 years old. Most patients are found to have urinary symptoms such as gross hematuria during physical examination or seek medical treatment. It has also been reported in the literature [10] that there is no gender difference in the occurrence of bladder PEComa, but most cases are under 40 years old, with no specific symptoms, and some patients have clinical symptoms similar to bladder cancer. This case is a 35-year-old male patient with abdominal pain as the main clinical symptom, which is not completely consistent with the literature. According to literature reports [11], PEComa CT examination mostly presents cystic solid mass with clear edges, large volume, and more than 5 cm in diameter. The CT value of solid tumor components in noncontrast scan is lower than soft tissue density (about 30HU), and the density can be uniform or uneven. On contrast enhanced scanning, the cystic wall was obviously enhanced, but the liquid part was not enhanced [12]. In this case, the tumor size was about 18.8 cm × 12.1 cm × 13.6 cm, and the density was uneven on plain scan, with necrotic cystic changes in the central area. The CT value of plain scan was 27–44hu. CT value: 94–108hu. Continuous enhancement at venous stage, CT value: 96–113hu, consistent with literature reports. Pathologically, PEComa has abundant blood supply, and most tumors have disordered blood supply arteries

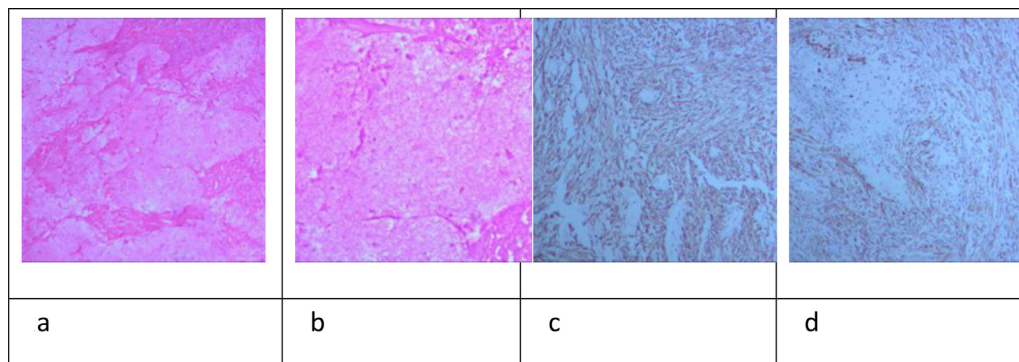


Fig. 2 – HE and immunohistochemistry. 2a and 2b (HE): spindle cell tumor with massive hemorrhage and infarction; Immunohistochemistry 2c, 2d: sma-1 (+), Vim (+), hmb-45 (+), s-100 (–).

and drainage veins, so persistent enhancement often occurs on images. In this case, the focal vessels were extremely abundant, and the supply arteries and drainage veins were also extremely disordered. Arteriovenous fistula-like changes were observed. It has been reported in the literature [13] that MRI showed uneven short T1 and slightly longer T2 signals in the whole tumor, but it has also been reported in the literature [14] that hemorrhagic or proteinaceous liquid components could occur in PEComa, that is, short T2 signal foci appeared in MRI. Benign PEComa is common, while metastatic PEComa is rare [15]. Routine radiotherapy and chemotherapy for PEComa have no obvious curative effect, and surgical resection is often chosen as the treatment method [16].

The cases of the bladder PEComa are rare and the image data is less. In combination with related literature [11,12] and performance in this tumor, the authors summarize characteristics of bladder PEComa: CT manifestations of class, oval or irregular shape pouch or solid mass, volume is bigger, often in more than 5 cm, margin out, noncontrast scan presented mixed density, necrosis, cystic change, enhanced scan presented solid tumor component significantly strengthened, and the rich arterial blood supply and venous drainage.

Differential diagnosis: (1) Bladder cancer: bladder cancer is usually manifested as protruding into the bladder cavity or asymmetric thickening of the bladder wall, and often presents clinical symptoms such as hematuria, frequent urination, and urgency [17]; (2) *Bladder lymphoma*: often accompanied by a history of chronic cystitis, CT, and MRI manifestations of submucosal mass and thickened bladder wall, often with clinical symptoms such as hematuria, dysuria, and increased nocturia [18].

In conclusion, it is difficult to diagnose bladder PEComa by imaging at present, and the diagnosis requires surgery and biopsy for pathologic diagnosis.

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