Review Article

# Bladder perivascular epithelioid cell neoplasm: Review on clinical features of this rare tumor

Zeno Adriaenssens[1], Robby Peter Anna Lamoury[1], Sam Tilborghs[1], Stefan De Wachter[1,2], Tim Brits[2]

[1]Department of Anatomy, Faculty of Medicine and Health Sciences, University of Antwerp, Antwerpen, [2]Department of Urology, Antwerp University Hospital, Edegem, Belgium

## Abstract

Perivascular epithelioid cell neoplasm (PEComa) is a rare mesenchymal tumor composed of histologically and immunohistochemically distinctive perivascular epithelioid cells. They can arise in various localizations such as the bladder. A total of 36 case reports regarding bladder PEComa have been described in the literature. Eleven reviews regarding this tumor have been published in literature so far primarily focusing on anatomic pathology. Through these reviews, it is known that in bladder PEComa, the melanocytic marker Human Melanoma Black-45 is expressed in 100% of cases whereas variable expression can be seen in multiple other melanocytic and myoid markers such as smooth muscle actin, *Melan-A*, *CD117*, *S100*, *CD31,* and CD34. Since current reviews mainly emphasize anatomic pathology, we perform a review focusing on the clinical aspects of PEComa at the level of the clinician. A manual electronic search of the PubMed/Medline and Web of Science Core Collection databases was conducted. Search was done on (perivascular epithelioid cell neoplasms [MeSH terms]) AND (Bladder). All case reports and reviews were encompassed until March 15, 2023, to identify studies that assessed bladder PEComa. The age of presentation is relatively low with a median age of 37 years. There is a female predominance with a female/male ratio of 1.5. The tumor shows no preference in anatomical localization within the bladder. Even involvement of the bladder neck, proximal urethra, and distal ureter has been described. The clinical presentation consists in the majority of patients of symptoms related to the urinary tract such as hematuria, dysuria, passage of urine sediment, frequency, and urgency. Other symptoms include abdominal discomfort and dysmenorrhea. In clinical examination, an abdominal mass can be found based on the size and location of the tumor. Further examination usually encompasses cystoscopy due to the hematuria and radiological investigations such as ultrasound (US), computed tomography, and magnetic resonance imaging. These radiological investigations reveal a heterogeneous solid mass with clear borders. In our center, we performed a transvaginal US additionally in a patient with bladder PEComa, which was the only investigation in our patient that concluded the mass was located in the Retzius space. For treatment, transurethral resection of the bladder tumor and partial cystectomy were both described in equal numbers. The choice of treatment depends on the localization and size of the tumor. Follow-up consists of imaging, but clear guidelines on this matter are lacking. Bladder PEComa is a rare condition and usually presents itself with nonspecific symptoms. Radiological investigations will reveal the tumor, but the final diagnosis is based on cytological and immunohistochemical features.

**Address for correspondence:** Dr. Zeno Adriaenssens, Molenbaan 47, 2920 Kalmthout, Belgium.
E-mail: zeno.adriaenssens@student.uantwerpen.be
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Since bladder PEComa is an entity with uncertain malignant potential, it is important to include this entity in the differential diagnosis when a patient presents with lower abdominal discomfort and lower urinary tract symptoms in combination with a mass in the pelvic region.

**Keywords:** Bladder, clinics, perivascular epithelioid cell neoplasm, systematic review

## INTRODUCTION

The World Health Organization defines perivascular epithelioid cell neoplasms (PEComas) as mesenchymal tumors composed of histologically and immunohistochemically distinctive perivascular epithelioid cells.[1] They were first described by Bonetti *et al.* in 1992, in angiomyolipoma, clear cell "sugar" tumors (CSST), and lymphangioleiomyomatosis.[2] PEComas have been documented in various anatomical localizations such as liver, kidney, lung, and uterus. However, only 36 cases of bladder PEComa are reported in the literature, which makes it an extremely rare condition. Until now 11 reviews on bladder PEComa were performed, most of them focusing on pathology, i.e., cytological and immunohistochemical features of the tumor. Through those reviews, we know that most of the PEComas are composed of a mixture of epithelioid cells and spindled cells, and that all cases described consistently express the melanocytic marker Human Melanoma Black-45 (*HMB45*).[3] However, there is a clear paucity of data regarding clinical features as no reviews focusing on PEComa emphasize these clinical features. In our center, a patient was treated with bladder PEComa which motivated us to review literature exploring the clinical presentation, imaging features, treatment modalities, and follow-up. This way we aim to improve the diagnostic accuracy of this tumor with unknown malignant potential at the level of the clinician in order to better understand how this condition can be recognized or when the suspicion of bladder PEComa should be raised.

## MATERIALS AND METHODS

A comprehensive literature search was conducted using the following databases: PubMed/MEDLINE (Ovid) and Web of Science Core Collection were considered for the collection of articles related to the topic of interest. A flowchart of the selection process is depicted in Figure 1. The primary key terms used during data collection were "perivascular epithelioid cell neoplasms (Medical Subject Headings terms)" and "bladder" using Boolean logic (AND). Duplicated articles were excluded from the study in advance. Based on these key terms and following in-depth screening of the relevance of each article, only articles relevant to this review were included. Articles were first screened and selected based on their abstract. If relevant, studies were read in detail. Database searches were supplemented by hand searching the reference lists of eligible articles. Afterward, reports were assessed for eligibility. Reports irrelevant after reading the manuscript were excluded. Two reports were thus excluded since they described PEComa not involving the bladder. There was no date restriction on the searched articles with the last search dated March 15, 2023.

During this systematic review, the main focuses were the clinical presentation, imaging features, treatment modalities, and follow-up.

## DISCUSSION

Bladder PEComa is an extremely rare tumor with only 37 cases presented in literature so far, our case included. Having such few cases makes it very hard to state clear guidelines for clinicians on this tumor. Earlier reviews mainly discussed cytological and immunohistochemical features of PEComa, primarily focusing on pathologists. Wu *et al.* performed statistical analysis of immunophenotypes in the published cases reporting these data, demonstrating 100% expression of the melanocytic marker *HMB45 (*34/34).[3] In the case described by Zeng *et al.* and in our case, both tumors also tested positive for *HMB45* expression, thereby keeping the expression rate at 100% in bladder PEComa. Second, based on the same data presented by Wu *et al.*, we can state that 79.3% (23/29) of previously presented cases, including our case, express smooth muscle actin.[3] Furthermore, we see variable expression of multiple melanocytic and myoid markers in bladder PEComa such as *Melan-A*, *CD117*, *S100*, *CD31*, and *CD34*.[4] Finally, bladder PEComa might be associated with transcription factor binding to IGHM enhancer 3-rearrangement, which is not necessarily associated with poor outcome.[5]

In this article however, our aim is to describe bladder PEComa not based on cytological examinations but purely based on clinical characteristics.

### Demographic features

Bladder PEComa patients described in literature tend to have a relatively low age with a median of 37 years, ranging

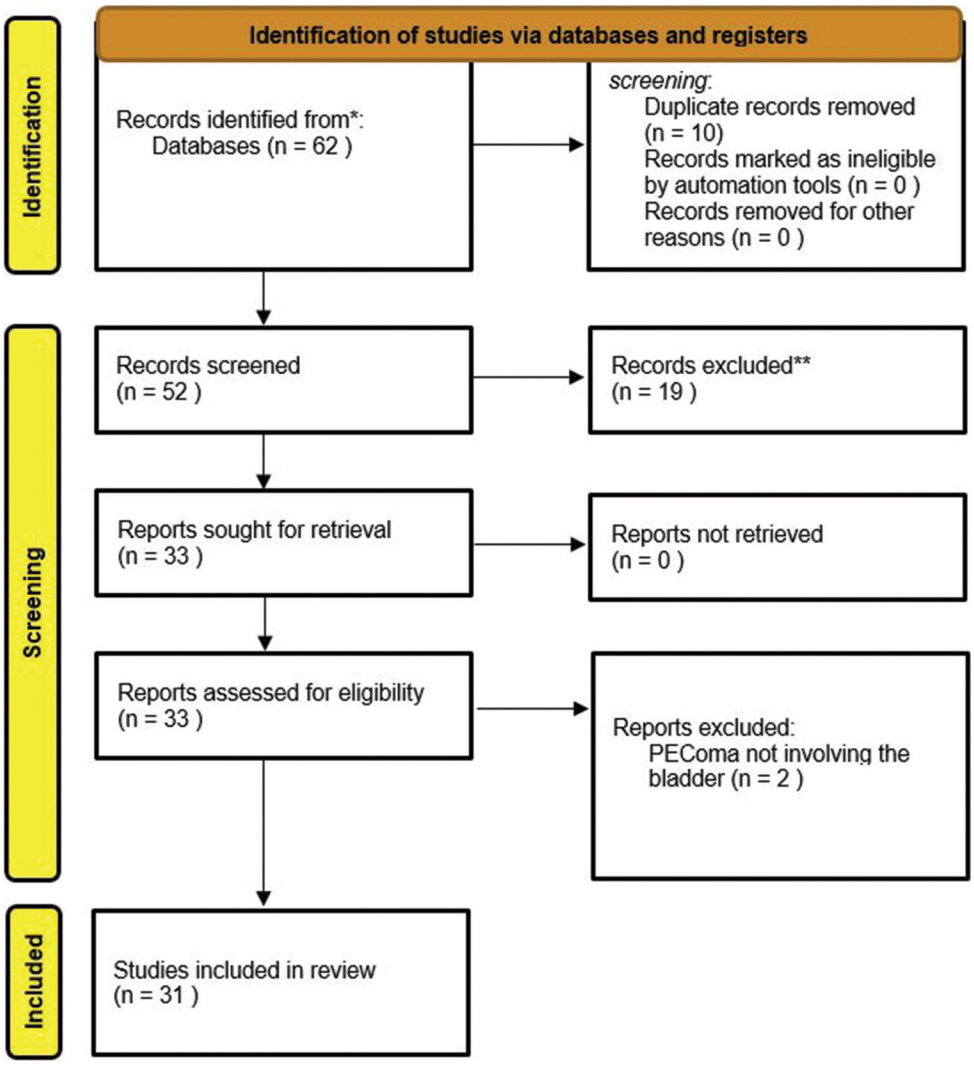


**Figure 1:** Preferred Reporting Items for Systematic Reviews and Meta-Analyses flow

from 16 to 78 years of age.[3,6] Thereby, more than 80% of cases (30/37) present between the third and fifth decades of life. Concerning sex, bladder PEComa has a slight female predominance with a female/male ratio of 1.5. Based on our literature search, the size of bladder PEComa varies from a small lesion of 0.6 cm to an enormous mass of 18.8 cm.[7,8] Consequently, the median size of bladder PEComa when diagnosed is 4.0 cm. The bladder tumor can arise at almost every anatomical location and does not really have a preferential site. Even involvement of the bladder neck, proximal urethra, and distal ureter has been described.[8-10] A wide overview of those characteristics is presented in Table 1.

## Clinical signs and symptoms

The majority of patients present themselves with symptoms related to the urinary tract [Table 1]. Only 5 of the 35 cases in which the clinical presentation is discussed report asymptomatic patients. In three of those asymptomatic patients the diagnosis was established through in identical findings on MRI, US or physical examination performed for other purposes.[11-13] Of the two remaining asymptomatic patients, it is not described how the diagnosis was established.[3] Subsequently, it was remarkable that in 13 cases, the patient had hematuria, and in 12 out of 35 cases, the patient had some kind of persistent lower abdominal discomfort upon presentation at the physician's office. Two patients presented themselves with dysmenorrhea as the main complaint. Other clinical symptoms described are dysuria, passage of urine sediment, urinary frequency, odynuria, and urgency.[6,14,15]

## Imaging modalities

### *Cystoscopy*

The imaging characteristics of bladder PEComa have some specificity.[16] The most used imaging technique in the described cases is cystoscopy (27/35). On cystoscopy, PEComa of the bladder is often described

**Table 1: Summary of clinical features of 37 bladder perivascular epithelioid cell neoplasm cases published in English literature**

| Case number | Age (years) | Sex | Maximum tumor size (cm) | Anatomical location | Clinical symptoms | Imaging features | Treatment | Follow-up (months) |
|---|---|---|---|---|---|---|---|---|
| 1 | 18 | Male | 5.50 | Muscularis propria; between prostate and left obturator muscle | Asymptomatic patient | MRI: For lumbar hernia showed solid mass. US: Heterogeneity mass | Surgical intervention | 12; NED |
| 2 | 55 | Female | 0.60 | Lamina propria of bladder floor | Transient lower abdominal pain for 1 year | US: 5 mm mass in bladder floor. Cystoscopy: 1 cm solid mass | TURBT | 48; NED |
| 3 | 33 | Female | 4.00 | Left inferior portion of bladder | Dysmenorrhea | CT: Sharply circumcised mass. Cystoscopy: Bulging mass with intact mucosa | Partial cystectomy | 72; NED |
| 4 | 19 | Female | 3.00 | Left lateral vesical wall | Hematuria | US/cystoscopy: Intraluminal polypoidal pedunculated mass | TURBT | Never reported back and lost in follow-up |
| 5 | 48 | Male | 3.00 | Posterior midline vesical wall | Dysuria, passage of urine sediment, lower abdominal discomfort | Cystoscopy: Lobular mass with mild bullous edema (coexistent enterovesical fistula found) | Laparotomy, partial cystectomy, partial small bowel resection. Adjuvant IFN-α immunotherapy | 48; NED |
| 6 | 39 | Male | 5.00 | Urachal cyst | NA | NA | Partial cystectomy | 8; NED |
| 7 | 24 | Female | 3.00 | Posterolateral | History of chronic pelvic pain | US: Solid right adnexal lesion protruding into posterolateral bladder wall. MRI: Homogenous soft-tissue mass. Cystoscopy: Mucosal-covered solid mass | Laparoscopic partial cystectomy | 3; NED |
| 8 | 36 | Male | 4.80 | Submucosal mass, anterior bladder wall | Hematuria | Cystoscopy revealed the mass | Partial cystectomy | 10; NED |
| 9 | 37 | Male | NA | Bladder dome | Hematuria | Cystoscopy revealed the lesion | TURBT | 21; NED |
| 10 | 26 | Female | 5.00 | Anterior bladder wall | Mass palpated on routine examination after hysterectomy | "Radiologic studies identified the mass" | Partial cystectomy after embolization | No follow-up information |
| 11 | 26 | Female | 5.40 | Right anterior wall | Ureterovaginal fistula 4 days posthysterectomy | Cystoscopy: Extrinsic mass effect on right bladder. CT: Pelvic mass. MRI: Enhanced venous structures | Arterio-embolization with partial cystectomy and ureteral reimplantation | No follow-up information |
| 12 | 23 | Male | 9.20 | Left lateral wall | 2-year history of frequent miction and odynuria | US: Inhomogeneous round mass. CT: Cystic mass with a thick irregular wall | Partial cystectomy | No follow-up information |
| 13 | 42 | Male | 6.00 | Right lateral wall | Good health, incidental finding on ultrasound screening | CT: 6 cm mass. PET: Hypermetabolic urinary bladder wall mass. Cystoscopy: Extrinsic mass | TURBT + robot-assisted laparoscopic partial cystectomy | No follow-up information |
| 14 | 44 | Female | 2.70 | Junction anterior wall of vagina and left posterior bladder wall | 3 months dysmenorrhea | MRI: Demonstration of the mass. Diagnostic laparoscopy: Endometriotic cyst. Cystoscopy: Extension of nodule to bladder | Open partial cystectomy, colpectomy, right ovariectomy, pelvic lymphadenectomy | 30; NED |
| 15 | 39 | Male | 3.00 | Superior and medically to the left ureteric orifice | Painless hematuria for 1 month | US: 3 cm mass left vesicoureteric junction. Cystoscopy: 3×3 solid mass | TURBT | 3; NED |
| 16 | 16 | Female | 3.00 | Left posterior wall | 3-year history of vague abdominal discomfort and 1 month frequent micturition | US: 2 cm solid mass. CT showed mass. MRI: Sharply circumscribed soft-tissue mass, wide base. Cystoscopy: Yellowish solid mass | TURBT | 13; NED |
| 17 | 54 | Male | 3.00 | Right anterior wall | Gross painless hematuria | US/MRI: Suspicious right anterior wall bladder tumor | TURBT + partial cystectomy | 11; NED |
| 18 | 55 | Female | 5.00 | Bladder wall | Hematuria | Cystoscopy: Solid mass suspicious for urothelial carcinoma | TURBT + radical cystectomy (6 m after TURB) and ChT | 6; infiltrative mass bladder and sigmoid colon. 10; widespread abdominal masses |

*Contd...*

**Table 1: Contd...**

| Case number | Age (years) | Sex | Maximum tumor size (cm) | Anatomical location | Clinical symptoms | Imaging features | Treatment | Follow-up (months) |
|---|---|---|---|---|---|---|---|---|
| 19 | 65 | Male | 2.50 | Right bladder wall | Recurrent hematuria and back pain (2 months) | US: Lesion 2 cm right bladder. CT: Heterogeneous enhancement, infiltration bladder wall. PET: Increased uptake at skeletal segments: L1, L5, and left iliac wing | TURBT + ChT (gemcitabine) | 6; stable disease |
| 20 | 27 | Female | 4.20 | Midline base of the wall, bladder neck, and proximal urethra | Intermittent painless gross hematuria | Cystoscopy/CT: Hypervascular solid round mass. MRI: Extravesical tumor in perivesical fat of Retzius space | TURBT + partial cystectomy and right pelvic node dissection | 6; metastasis and recurrence |
| 21 | 39 | Female | 4.00 | Bladder neck and anterior bladder wall into Retzius space | 8-year history of right lower abdominal pain and occasional urinary urgency | US: Possible bladder lesion. MRI: Soft-tissue lesion. Cystoscopy: Immediate visualization | TURBT + robotic laparoscopic partial cystectomy | 6; NED |
| 22 | 29 | Female | 1.50 | Right wall | LUTS | CT: 1.5 cm hypodense nodule in right bladder. Cystoscopy: Confirmation | TURBT | 25; NED |
| 23 | 44 | Male | NA | Bladder wall | NA | NA | NA | 13; NED |
| 24 | 49 | Female | 5.80 | Left lateral wall | Recurrent lower abdominal pain for 1 month. Painless mass touched above uterus on PE | US: Elliptic mass, clear border, slight irregular wall. CT: Homogenous cystic nodule | Partial cystectomy | 18; NED |
| 25 | 57 | Female | 4.00 | Anterior wall | Recurrent lower abdominal discomfort | CT: 4 cm spherical mass on anterior bladder wall. Cystoscopy: Round-shaped, solid | Explorative laparotomy with partial cystectomy | 24; NED |
| 26 | 27 | Male | 3.00 | Anterior bladder neck | Painless gross hematuria and acute urinary retention | Cystoscopy: 3 cm polypoid, solid, vascular tumor at the anterior bladder neck | TURBT | 12; NED |
| 27 | 74 | Female | 3.00 | Left posterolateral wall | Single episode of hematuria | CT: Mass in left posterolateral bladder wall. Cystoscopy: Confirmation. MRI: 3 cm lesion | TURBT + radical cystectomy | NA |
| 28 | 36 | Male | 18.80 | Right outer wall of the bladder bottom and right ureter | 5 months dull pain in lower abdomen and fatigue | CT: Large oval cystic mass, abundant vessels | Partial cystectomy | NA |
| 29 | 78 | Female | 5.00 | Bladder wall | 3×recurrent lower abdominal discomfort. 2×painless hematuria. 2×asymptomatic | US: Solid hypoechoic masses. CT: Nodular or irregular soft-tissue density masses with clear boundaries. Cystoscopy: Intraluminal round or papillary masses | NA | NA |
| 30 | 37 | Female | 6.00 | Left bladder neck | | | TURB | 13; recurrences 40; NED |
| 31 | 31 | Female | 5.50 | Bladder wall | | | TURB | 10; recurrences 30; NED |
| 32 | 26 | Male | 1.50 | Right anterior wall | | | TURB plus ChT | 12; NED |
| 33 | 55 | Female | 4.00 | Right anterior wall | | | Partial cystectomy | 134; NED |
| 34 | 34 | Female | 3.00 | Right posterior wall | | | Partial cystectomy | 30; NED |
| 35 | 30 | Male | 6.90 | Right lateral wall | | | Partial cystectomy | 23; NED |
| 36 | 66 | Female | 1.50 | Right posterior bladder wall | Repeated UTI for 3 years (DMT2), asked for review of urinary tract (microscopic hematuria) | US: Strong echogenic mass, clear borders. CT/MRI: Round nodular mass, clear borders. Cystoscopy: Reddish-brown round mass | Transurethral ERBT | 6; NED |
| 37 (our case) | 26 | Female | 7.40 | Detrusor muscle, bladder neck | Abdominal discomfort and minor urinary frequency | US: Mass present. CT: Sharply bounded mass. Cystoscopy: External compression bladder fundus. MRI: Same mass. TVUS: Inhomogeneous mass in Retzius space | Robotic-assisted laparoscopic partial cystectomy | 3; NED |

NA: Not applicable, UTI: Urinary tract infection, LUTS: Lower urinary tract symptoms, PE: Physical examination, DMT2: Diabetes mellitus type 2, TURB: Transurethral resection of bladder, MRI: Magnetic resonance imaging, US: Ultrasound, CT: Computed tomography, PET: Positron emission tomography, TVUS: Transvaginal US, TURBT: TURB tumor, IFN-α: Interferon-alpha, ChT: Chemotherapy, ERBT: En bloc resection of bladder tumor, NED: No evidence of disease

as intraluminal polypoid solid masses with or without pedicle. Furthermore, they show to be round-shaped having yellowish to reddish-brown color [Figure 2].[4,6,17] The lesions are also reported to be strongly vascularized.[8,18] Finally, on cystoscopy, extrinsic mass effect on the bladder is seen in extraluminal PEComas.[12,19]

### *Ultrasound*

When US is performed in bladder PEComa, a heterogenic mass with hyperechoic center and hypoechoic surroundings is found.[11,15] Furthermore, the tumour is often described as round or elliptic masses with clear borders.[17,20]

### *Computed tomography and magnetic resonance imaging*

Computed tomography (CT) typically shows bladder PEComa as sharply circumcised cystic or nodular masses.[21] Second, CT defines this tumor as hypervascularized with abundant presence of vessels.[8,9] On MRI, multiple cases including a case of PEComa we had in our center report iso-intense signalization on T1-weighted images and heterogeneous signals in T2-weighted images.[16] Again, the soft-tissue mass is described to be sharply circumcised with enhanced venous structures.[6,19]

### *Alternative imaging modalities*

One study performed positron emission tomography on the bladder US showing a hypermetabolic mass.[12] In the case of PEComa in our center, a transvaginal ultrasound (TVUS) was performed. This has not been described in previous reports so far. In addition, despite CT, MRI, US, and cystoscopy, this preoperatively performed TVUS was the only imaging modality to precisely locate the extraluminal bladder PEComa in the Retzius space and not involving reproductive organs. Therefore, TVUS proved itself important in the exact localization and involvement of possible surrounding tissues. Figure 3 provides characteristics of bladder PEComa on different imaging modalities.

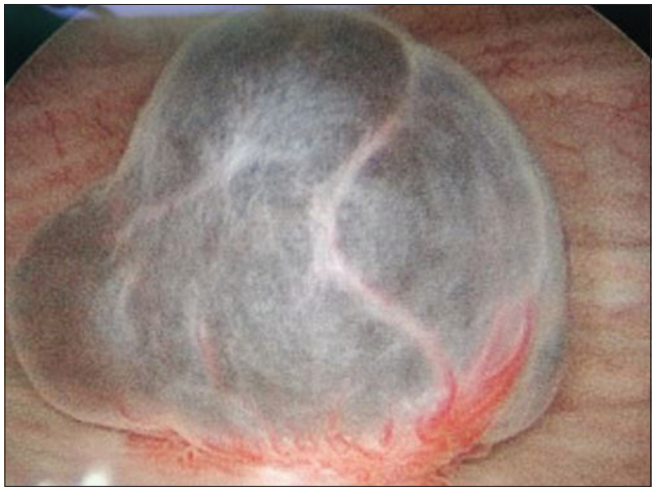

**Figure 2:** Example of Perivascular epithelioid cell neoplasm during cystoscopy[17]

## Treatment

When it comes to the treatment of bladder PEComa, surgical resection is the performed intervention in all cases. Transurethral resection of bladder tumor (TURBT) and partial cystectomy are by far the most used techniques. They were almost equally performed in the reported cases. TURBT is especially used in cases where the intraluminal tumor only invades submucosa or lamina propria. When the tumor invades the muscular layers of the bladder wall or has extraluminal extent, partial cystectomy should be the treatment procedure of choice. So far, the literature also describes five cases in which TURBT was followed by partial cystectomy and one case in which TURBT was followed by radical cystectomy [Table 2]. In four of those cases, the subsequent partial cystectomy was performed short after TURBT failed to remove the bladder PEComa completely.[10,12,22,23] In the fifth case, 6 months after TURBT, an infiltrative mass invaded the entire thickness of the bladder wall and a metastatic nodule in the sigmoid colon was found. Therefore, radical cystectomy was carried out and the nodule in the sigmoid colon was synchronously resected. Unfortunately, 10 months after initial TURBT, follow-up imaging showed widespread abdominal masses that were considered metastasis. The patient underwent palliative chemotherapy but succumbed to disease 2 months later.[24] The last case described persistent postprocedural hematuria for which the patient required continuous bladder irrigation. Progressively worsening conditions necessitated emergent exploratory laparotomy with transvesical tumor excision. Since cutting edges were positive, a third surgical resection involving new partial cystectomy with right pelvic lymph node dissection was performed. Six months later, multiple PEComa

**Table 2: Six cases in which transurethral resection of bladder tumor was followed by additional surgical treatment**

| Case | Treatment | Reason |
|---|---|---|
| Chan *et al*. | TURBT + robotic-assisted laparoscopic partial cystectomy | Uncomplete removal of bladder tumour |
| Abou Ghaida *et al*. | TURBT + partial cystectomy | Uncomplete removal of bladder tumour |
| Williamson *et al*. | TURBT + radical cystectomy (6m after TURBT) and ChT | Recurrence and widespread abdominal metastasis found 6-10 months post-TURBT |
| Russell *et al*. | TURBT + partial cystectomy and right pelvic node dissection | Progressive worsening clinical condition after post-TURBT persistent hematuria |
| Tarplin *et al*. | TURBT + robotic laparoscopic partial cystectomy | Uncomplete removal of bladder tumour |
| Tricard *et al*. | TURBT + Radical cystectomy | Uncomplete removal of bladder tumour |

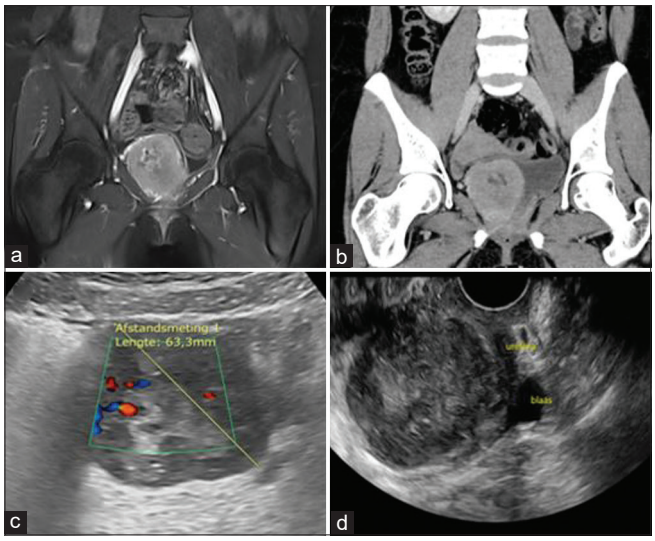


**Figure 3:** Different imaging characteristics of bladder perivascular epithelioid cell neoplasm in one patient. (a) Magnetic resonance imaging T1 iso-intense mass with central necrotic component located between anterior vaginal wall and right bladder wall. The mass is sharply demarcated. (b) Computed tomography scan with intravenous contrast medium showing a perivesical mass with cystic or necrotic components and punctiform calcifications. (c) Ultrasound abdomen with a large heterogeneous structure in the pelvis with hyperreflective center. Differentiation between the mass and surrounding tissues is impossible due to insufficient bladder filling. (d) Transvaginal ultrasound showing a solid tumor with central necrosis. The tumor is located anteriorly of the bladder and urethra in the Retzius space

metastases were found in the abdominal wall, lung, and left hypochondriac lymph nodes.[9]

In total, 18 patients first received TURBT as treatment for bladder PEComa. Of those patients, six required additional surgical resection consisting of partial/radical cystectomy. In two of those patients, metastatic disease was found after TURBT of which one required emergent laparotomy due to persistent bleeding after TURBT and the other died because of widespread metastasis. Although TURBT is a minimally invasive way to treat bladder PEComa, it should always be considered properly whether it is the best treatment option. Avoidance of tumor seeding is essential and the fact that bladder PEComas are described as vascular lesions might also play a role in the decision-making.

### Follow-up

Due to the few cases presented, clear follow-up guidelines are lacking. When we, as clinicians, want to set up a follow-up plan for a patient diagnosed with bladder PEComa, the tendency of malignant degeneration of the tumor is essential. Few things have been written on this subject, and Folpe *et al*. proposed a classification for malignancy of PEComas, mainly based on gynecological tumors. The proposed criteria for malignant potential are: >5 cm, infiltrative growth, high nuclear grade and cellularity, mitotic rate >1/50 high-power field (HPF), necrosis, and vascular invasion.[25] In total, three of the earlier published cases of bladder PEComa showed metastatic disease status. In one case, the metastatic disease consisted of skeletal metastatic lesions already there on the first presentation.[26] In the two other cases, the metastatic manifestation presented only after initial TURBT treatment.[9,24] When we compare the three cases with metastatic disease, we notice that they have at least three out of six criteria proposed by Folpe *et al*. that increase the chance for malignant potential. Although most cases follow a benign disease course, the follow-up plan must include regular imaging to check for possible recurrence since three of the presented cases had a malignant disease course. The precise timing of those imaging modalities should depend on the malignant potential. To estimate this potential, the criteria proposed by Folpe *et al*. can still be used. The case of PEComa in our center had three out of six proposed criteria (>5 cm, necrosis and mitotic activity >1/50 HPF). Therefore, a board of experts decided to suggest strict follow-up of 5–10 years after initial diagnosis with regular CT thorax and abdomen or full-body MRI if the capacity allows this technique to be used.

## CONCLUSIONS

In this article, we reviewed the available literature for valuable clinical information on bladder PEComa. This way we want to provide the clinician tools to recognize bladder PEComa, how to treat it, and how to organize follow-up after treatment. Due to the uncertain malignant potential of the tumor, it is important to include bladder PEComa to the diagnostic landscape in selected cases. Most cases of bladder PEComa present between the third and fifth decades of life, with slight female predominance (female: male ratio is 1.5). Bladder PEComa can present itself asymptomatically, but most patients present with hematuria and persistent lower abdominal discomfort. On imaging modalities, it is mostly described as a well-defined polypoid solid mass. Concerning treatment, surgical resection remains the main treatment option. TURBT and robot-assisted laparoscopic partial cystectomy are equally used in the described cases. Selection between both treatment options should be based on the location and infiltration of the lesion. Besides, it should be kept in mind that bladder PEComas are often highly vascularized which could complicate TURBT. Finally, to compose the follow-up plan, regular imaging using cystoscopy, CT, or MRI depends on the potential malignancy. Therefore, criteria (>5 cm, infiltrative growth, high nuclear grade and cellularity, mitotic rate >1/50 HPF, necrosis, and vascular invasion) proposed by Folpe *et al*. can, in our opinion, guide to estimate this potential.

## Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. The patient has given her consent for the images to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

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Nil.

## Conflicts of interest

There are no conflicts of interest.

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