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

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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.

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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.<sup>[1]</sup> They were first described by Bonetti et al. in 1992, in angiomyolipoma, clear cell "sugar" tumors (CSST), and lymphangioleiomyomatosis.<sup>[2]</sup> 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).<sup>[3]</sup> 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).<sup>[3]</sup> 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.<sup>[3]</sup> 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.<sup>[5]</sup>

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.<sup>[3,6]</sup> 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.<sup>[7,8]</sup> 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.<sup>[8-10]</sup> 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.<sup>[11-13]</sup> Of the two remaining asymptomatic patients, it is not described how the diagnosis was established.<sup>[3]</sup> 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.<sup>[6,14,15]</sup>

## **Imaging modalities**

## Cystoscopy

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

Table 1:	Summ	ary of cli	inical features	of 37 bladder perivascu	lar epithelioid cell neoplasm	cases published in English literature		
Case	Age (vears)	Sex	Maximum tumor size (cm)	Anatomical location	Clinical symptoms	Imaging features	Treatment	Follow-up (months)
-	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 vear	US: 5 mm mass in bladder floor. Cvstoscopv: 1 cm solid mass	TURBT	48; NED
8	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
2	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-reimmuntherand	48; NED
۶ ک	39 24	Male Female	5.00 3.00	Urachal cyst Posterolateral	NA History of chronic pelvic pain	NA US: Solid right adnexal lesion protruding into posterolateral bladder wall. MRI: Homogenous soft-tissue mass. Cystoscopy: Mucosal-covered solid mass	ratial cystectomy Laparoscopic partial cystectomy	8; NED 3; NED
ω	36	Male	4.80	Submucosal mass, anterior bladder wall	Hematuria	Cystoscopy revealed the mass	Partial cystectomy	10; NED
9 10	37 26	Male Female	NA 5.00	Bladder dome Anterior bladder wall	Hematuria Mass palpated on routine	Cystoscopy revealed the lesion "Radiologic studies identified the mass"	TURBT Partial cystectomy after	21; NED No follow-up
11	26	Female	5.40	Right anterior wall	examination atter nysterectomy Ureterovaginal fistula 4 days posthysterectomy	Cystoscopy: Extrinsic mass effect on right bladder. CT: Pelvic mass. MRI: Enhanced vennus structures	embolization Arterio-embolization with partial cystectomy and ureteral reimplantation	information No follow-up information
12	23	Male	9.20	Left lateral wall	2-year history of frequent mirtion and odviniria	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	No follow-up information
14	44	Female	2.70	Junction anterior wall of vagina and left posterior bladder wall	3 months dysmenorrhea	uses Diagnostic laparoscopy: Endometriotic cyst. Cystoscopy: Extension of nodule to bladder	opercounty Open partial cystectomy, colpectomy, right ovariectomy, pelvic lymohadenectomy	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. Costosconor: 3x3 solid mass	TURBT	3; NED
16	16	Female	3.00	Left posterior wall	3-year history of vague abdominal discomfort and 1	opticocopy: 50% on a mass Substantial mass. CT showed mass. MRI: Sharply circumarcribed soft-tissue mass. wide base Cvetosconv: Vallowich 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 cvstectomy	11; NED
18	55	Female	5.00	Bladder wall	Hematuria	Cystoscopy: Solid mass suspicious for urothelial carcinoma	TÚRBT + radical cystectomy (6 m after TURB) and ChT	6; infiltrative mass bladder and sigmoid colon.
								10; widespread abdominal masses
								Contd

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Table 1: (	Sontd							
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 scolaral commonts: 11, 15, and loft illoc wing	TURBT + ChT (gemcitabine)	6; stable disease
20	27	Female	4.20	Midline base of the wall, bladder neck, and	Intermittent painless gross hematuria	sketetar segments. L1, L2, and retuined wing Cystoscopy/CT: Hypervascular solid round mass. MRI: Extravesical tumor in perivesical far of Retrins snare	TURBT + partial cystectomy and right	6; metastasis and recurrence
21	39	Female	4.00	Bladder neck and anterior bladder wall into Batzius snare	8-year history of right lower abdominal pain and occasional	US: Possible bladder lesion. MRI: Soft-tissue lesion. Cystoscopy: Immediate visualization	TURBT + robotic laparoscopic partial	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	AN 202	Bladder wall	NA	NA LIC TILICITIC CONTRACTOR OF CONTRACTOR	NA	13; NED
24	47	remale	0.8.0	Lett lateral wall	Recurrent lower appominal 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; NEU
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. Cvstoscopv: Confirmation. MRI: 3 cm lesion	TURBT + radical cvstectomv	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	US: Solid hypoechoic masses. CT: Nodular	NA	NA
30	37	Female	6.00	Left bladder neck	discomfort. 2×painless hematuria. 2×asymptomatic	or irregular soft-tissue density masses with clear boundaries. Cystoscopy: Intraluminal	TURB	13; recurrences 40; NED
31	31	Female	5.50	Bladder wall		round or papillary masses	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 21	34	Female	3.00	Right posterior wall			Partial cystectomy	30; NED
36 36	66 66	Male Female	0.90	kight lateral wall Right posterior bladder wall	Repeated UTI for 3 years (DMT2), asked for review of urinary	US: Strong echogenic mass, clear borders. CT/MRI: Round nodular mass, clear borders. Cystoscopy: Reddish-brown round	Partial cystectomy Transurethral ERBT	23; NED 6; NED
37 (our case)	26	Female	7.40	Detrusor muscle, bladder neck	tract (microscopic hematuria) Abdominal discomfort and minor urinary frequency	mass 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

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

#### Ultrasound

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

## Computed tomography and magnetic resonance imaging

Computed tomography (CT) typically shows bladder PEComa as sharply circumcised cystic or nodular masses.<sup>[21]</sup> Second, CT defines this tumor as hypervascularized with abundant presence of vessels.<sup>[8,9]</sup> 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.<sup>[16]</sup> Again, the soft-tissue mass is described to be sharply circumcised with enhanced venous structures.<sup>[6,19]</sup>

#### Alternative imaging modalities

One study performed positron emission tomography on the bladder US showing a hypermetabolic mass.<sup>[12]</sup> 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



Figure 2: Example of Perivascular epithelioid cell neoplasm during cystoscopy<sup>[17]</sup>

characteristics of bladder PEComa on different imaging modalities.

## 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.<sup>[10,12,22,23]</sup> 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.<sup>[24]</sup> 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	2: Six	cases	in w	hich	transu	irethral	resection	of	bladde
tumor	was	followe	d by	add	itional	surgica	I treatme	nt	

Case	Treatment	Reason
Chan <i>et al</i> .	TURBT + robotic-assisted	Uncomplete removal of
Abou Ghaida <i>et al</i> .	TURBT + partial cystectomy	Uncomplete removal of bladder tumour
Williamson <i>et al</i> .	TURBT + radical cystectomy (6m after TURBT) and ChT	Recurrence and widespread abdominal metastasis found 6-10 months post-TURBT
Russell <i>et al</i> .	TURBT + partial cystectomy and right pelvic node dissection	Progressive worsening clinical condition after post-TURBT persistent hematuria
Tarplin <i>et al</i> .	TURBT + robotic laparoscopic	Uncomplete removal of
Tricard <i>et al</i> .	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.<sup>[9]</sup>

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.<sup>[25]</sup> 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.<sup>[26]</sup> In the two other cases, the metastatic manifestation presented only after initial TURBT treatment.<sup>[9,24]</sup> 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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## **Conflicts of interest**

There are no conflicts of interest.

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