

ROBOTIC-ASSISTED SURGICAL MANAGEMENT OF A PRIMARY PROSTATIC PERIVASCULAR EPITHELIOID CELL TUMOR (PECOMA): A RARE AND CHALLENGING CASE

MANEJO CIRÚRGICO ASSISTIDO POR ROBÓTICA DE TUMOR PRIMÁRIO DE CÉLULAS EPITELIOIDES PERIVASCULARES (PECOMA) DA PRÓSTATA: UM CASO RARO E DESAFIADOR

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Received: 08/31/2026. Accept: 09/20/2026

RESUMO

Os tumores de células epitelioides perivasculares (PEComas) são neoplasias mesenquimais raras, caracterizadas por diferenciação miomelanocítica e comportamento biológico variável, sendo o acometimento primário da próstata excepcional. Relata-se o caso de um homem de 68 anos, com múltiplas comorbidades, que apresentou sintomas obstrutivos do trato urinário inferior e hematúria macroscópica persistente. Os exames de imagem evidenciaram uma massa sólida em íntimo contato com a próstata e o assoalho vesical, sem definição etiológica pré-operatória. Foi realizada ressecção transuretral da próstata com finalidade diagnóstica e alívio sintomático. O estudo anatomopatológico revelou neoplasia fusocelular maligna com extensas áreas de necrose, e a imuno-histoquímica demonstrou positividade para marcadores musculares lisos e melanocíticos, com negatividade para citoceratinas, confirmando o diagnóstico de PEComa maligno prostático. De acordo com os critérios de Folpe, o tumor foi classificado como de alto risco para comportamento agressivo. O caso ilustra as dificuldades diagnósticas dessa entidade rara, frequentemente confundida com patologias urológicas comuns, e reforça a importância da imuno-histoquímica para o diagnóstico definitivo. A ressecção cirúrgica completa permanece o pilar do tratamento, enquanto terapias sistêmicas direcionadas, como inibidores da via mTOR, podem ser consideradas em casos selecionados. O seguimento clínico prolongado é fundamental devido à imprevisibilidade do comportamento biológico desses tumores.

PALAVRAS-CHAVE: Tumor de células epitelioides perivasculares (PEComa); Próstata; Neoplasias prostáticas; Tumores raros; Oncologia urológica; Cirurgia robótica assistida; Prostatectomia radical; Imuno-histoquímica.

ABSTRACT

Perivascular epithelioid cell tumors (PEComas) are rare mesenchymal neoplasms characterized by a distinctive myomelanocytic immunophenotype and variable biological behavior, with primary prostatic involvement being exceptionally uncommon. We report the case of a 68-year-old man with multiple comorbidities who presented with progressive lower urinary tract symptoms and persistent gross hematuria. Imaging studies revealed a large solid mass closely related to the prostate and bladder floor, without definitive preoperative etiological characterization. Transurethral resection of the prostate was performed for diagnostic purposes and symptomatic relief. Histopathological examination demonstrated a malignant spindle-cell neoplasm with extensive tumor necrosis, and immunohistochemical analysis showed positivity for smooth muscle and melanocytic markers, with negativity for epithelial markers, confirming the diagnosis of malignant prostatic PEComa. According to Folpe criteria, the tumor was classified as high risk for aggressive behavior. This case highlights the diagnostic challenges posed by this rare entity, which often mimics common urological conditions, and underscores the critical role of immunohistochemistry in achieving an accurate diagnosis. Complete surgical resection remains the cornerstone of treatment, while targeted systemic therapies, particularly mTOR inhibitors, may be considered in selected cases. Long-term follow-up is mandatory due to the unpredictable biological behavior of these tumors.

KEYWORDS: Perivascular epithelioid cell tumor (PEComa); Prostate; Prostatic neoplasms; Rare tumors; Urologic oncology; Robotic-assisted surgery; Radical prostatectomy; Immunohistochemistry.

1. INTRODUCTION

Perivascular epithelioid cell tumors (PEComas) comprise a rare and heterogeneous group of mesenchymal neoplasms characterized by the presence of epithelioid and spindle cells arranged around blood vessels and by a distinctive immunophenotypic profile, with co-expression of melanocytic and smooth muscle markers^{1,2}. Since their initial description, PEComas have been recognized as a distinct entity within the classification of soft tissue tumors.

These neoplasms may arise in various anatomical sites, with a predilection for the kidney, gynecologic tract, and gastrointestinal tract³. Involvement of the male genitourinary tract is uncommon, and primary prostatic involvement is exceptionally rare, with only a very limited number of cases reported in the literature^{4,5}. This rarity contributes to significant diagnostic challenges, as the clinical presentation often mimics more common urologic conditions, such as benign prostatic hyperplasia and epithelial prostatic neoplasms.

Histopathologically, PEComas exhibit variable architecture, composed of clear or eosinophilic cells with abundant cytoplasm and a characteristic perivascular distribution. Definitive diagnosis relies fundamentally on immunohistochemistry, with positivity for melanocytic markers such as HMB45 and Melan-A, together with expression of smooth muscle markers including actin, caldesmon, and desmin, representing a defining feature^{1,2}. This immunophenotypic profile enables differentiation from other mesenchymal and epithelial neoplasms of the genitourinary tract.

Although some PEComas demonstrate indolent behavior, there is consistent evidence of tumors with malignant potential, associated with an increased risk of local recurrence and metastasis⁶. Histological features such as large tumor size, necrosis, marked nuclear atypia, and increased mitotic activity have been correlated with worse prognosis⁷. Given the scarcity of available data and the clinical and histological variability of these tumors, case reports remain essential to expand understanding of the biological behavior, diagnostic criteria, and therapeutic strategies for prostatic PEComas. In this context, we describe a case of primary prostatic PEComa, emphasizing its clinical, pathological, and immunohistochemical findings in correlation with the current literature.

2. CASE REPORT

A 68-year-old male patient with a history of chronic kidney disease (CKD), smoking, and benign prostatic hyperplasia (BPH) presented with dyspnea at rest, anorexia, and changes in urinary pattern. Prominent lower urinary tract obstructive symptoms were noted, associated with persistent and large-volume gross hematuria lasting five days, refractory to medical management with tranexamic acid. During the initial diagnostic workup, the patient was referred for evaluation of a solid protruding lesion in the vesical region identified on imaging studies.

On admission, he was alert and oriented, breathing comfortably on room air. Digital rectal examination revealed a markedly enlarged prostate (grade 3), with preserved limits, predominantly firm and heterogeneous consistency, and an irregular surface, with left-sided asymmetry predominance. No clearly delimited nodules were identified. The examination was minimally painful, with no evidence of fixation to adjacent structures.

Laboratory evaluation showed a total serum prostate-specific antigen (PSA) level of 2.71 ng/mL (reference value ≤ 2.50 ng/mL), with a free-to-total PSA ratio of 10%.

Transabdominal and suprapubic prostate ultrasound performed in May 2024 demonstrated a solid expansive lesion measuring approximately 6.3×5.5 cm, located at the bladder base and in close contact with the prostate, raising suspicion for significant prostatic protrusion or bladder neoplasm. The prostate volume was estimated at 58 g, with dimensions of $4.5 \times 4.9 \times 4.7$ cm, globular shape, regular contours, preserved anatomical limits, and heterogeneous echotexture. The examination supported the clinical picture of infravesical obstruction, showing a pre-void bladder volume of 350 mL and a post-void residual volume of 238 mL. Based on these findings, cystoscopy was indicated for further diagnostic clarification.



Figure 1. Pelvic magnetic resonance imaging (MRI), sagittal plane, demonstrating a large expansive mass in the prostatic region, with heterogeneous signal intensity and central hypointense areas, which is producing mass effect on the urinary bladder anteriorly and posterior displacement of the rectum, without evident invasion of adjacent bony structures in the presented sections.

Subsequently, urethrocystoscopy revealed an unremarkable penile urethra and a prostatic urethra with signs of previous resection of the median lobe, with preservation of the lateral lobes. The bladder showed intact mucosa, diffuse trabeculation, and multiple diverticula, findings consistent with a high-pressure bladder secondary to chronic bladder outlet obstruction.

The ureteral orifices were in their usual anatomical positions, patent, with bilaterally visible urinary efflux. A sessile lesion measuring approximately 6 cm was identified on the left lateral bladder wall, contiguous with the bladder neck and prostate. These endoscopic findings supported the diagnosis of infravesical obstruction associated with a large expansive lesion.

Given these findings, a robotic surgical approach was indicated for both diagnostic and therapeutic purposes, considering the extent of the lesion and the need for complete resection with enhanced anatomical precision. The patient underwent robotic-assisted radical prostovesiculectomy combined with partial cystectomy. Bilateral double-J ureteral stents were also placed to prevent ureteral injury.

The surgical specimen was submitted for histopathological examination. Macroscopically, multiple irregular brownish elastic tissue fragments were identified, weighing 97.0 g in total and measuring $13.5 \times 7.3 \times 2.4$ cm in aggregate (Figure 2). Histological analysis with hematoxylin-eosin staining demonstrated a malignant spindle cell neoplasm with extensive areas of tumor necrosis (Figure 2). Due to the undifferentiated spindle cell morphology, immunohistochemical studies were requested for accurate characterization and classification of the neoplasm (Figure 3).

The exclusion of epithelial neoplasms, particularly prostatic adenocarcinoma, was supported by the absence of epithelial marker expression and reinforced by positivity for melanocytic and smooth muscle markers, establishing the characteristic immunophenotypic profile of perivascular epithelioid cell tumors. The combined morphological and immunohistochemical findings were consistent with the diagnosis of malignant PEComa involving the prostate and bladder base. The presence of significant cytologic atypia and extensive tumor necrosis supported a classification associated with aggressive biological behavior.

Immunohistochemical analysis demonstrated positivity for caldesmon, calponin, and desmin, with focal positivity for HMB45 and Melan-A, confirming myomelanocytic differentiation. The tumor was negative for AE1/AE3 (cytokeratins), CD117 (c-KIT), CD34, LHK8 (MUC1), and S100. The absence of cytokeratin expression represents a critical finding for excluding epithelial neoplasms, particularly prostatic adenocarcinoma, further supported by the co-expression of melanocytic and smooth muscle markers, thereby establishing the characteristic immunophenotypic profile of perivascular epithelioid cell tumors.

The combined morphological and immunohistochemical findings were consistent with the diagnosis of malignant PEComa involving the prostate and bladder base. The presence of marked cytologic atypia and extensive tumor necrosis supports classification as a lesion with aggressive biological behavior.

Following confirmation of malignant PEComa involving the prostate and bladder base, the case was discussed in a multidisciplinary setting, considering the

local extent of the disease, the aggressive histological features evidenced by extensive necrosis, and the patient's clinical comorbidities. Up to that point, transurethral resection had played a fundamental role in establishing the diagnosis and providing partial relief of infravesical obstruction



Figure 2. Macroscopic appearance of the surgical specimen, showing an irregular solid mass, brownish yellow in color, and measuring $13.5 \times 7.3 \times 2.4$ cm; consistent with prostatic mesenchymal neoplasia.

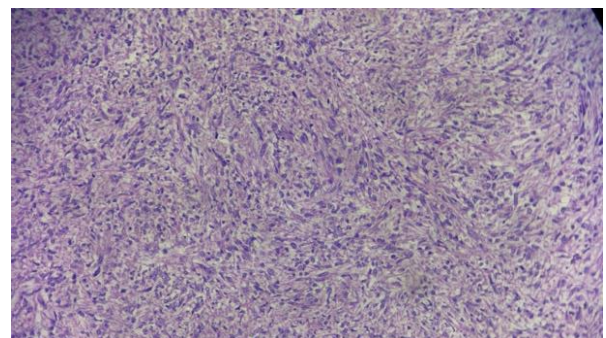


Figure 3. Photomicrograph demonstrating a neoplastic proliferation composed of epithelioid and spindle cell arranged in solid sheets and interlacing fascicles with eosinophilic cytoplasm. The neoplastic proliferation has an oval to elongated nuclei with no marked atypia. Histomorphological features are consistent with a perivascular epithelioid cell tumour (PEComa) (hematoxylin and eosin stain).

At the most recent follow-up visit, the patient was continent and reported partially preserved sexual function, with spontaneous nocturnal erections. Oncologic and functional surveillance was established, consisting of pelvic magnetic resonance imaging at six-month intervals, combined with computed tomography of the chest, abdomen, and pelvis every six months, aiming at early detection of local recurrence or metastatic disease.

3. DISCUSSION

Perivascular epithelioid cell tumors are exceptionally rare neoplasms, with a predilection for the kidney, gynecologic tract, and gastrointestinal tract, whereas primary prostatic involvement represents an extremely uncommon occurrence³. To date, only a limited number of prostatic PEComa cases have been reported, making each new case relevant for expanding clinical and surgical understanding of this entity^{4,5}.

From a clinical standpoint, prostatic PEComa tends to mimic far more prevalent urologic conditions. Lower urinary tract obstructive symptoms, urinary retention, and hematuria are indistinguishable from those observed in benign prostatic hyperplasia or epithelial neoplasms of the prostate and bladder, thereby limiting the possibility of an accurate preoperative diagnosis⁵.

Definitive diagnosis is fundamentally histopathological and relies on a characteristic immunohistochemical profile. The absence of epithelial markers, particularly cytokeratins and prostate-specific antigen (PSA), combined with positivity for melanocytic and smooth muscle markers, represents the diagnostic signature of PEComas^{1,2,8}. The present case clearly illustrates this diagnostic challenge, as only immunohistochemical analysis allowed exclusion of epithelial neoplasms and more common sarcomas of the genitourinary tract.

Prognostic stratification remains challenging. The criteria proposed by Folpe *et al.* continue to represent the main framework for assessing the malignant potential of PEComas, considering factors such as tumor size greater than 5 cm, necrosis, nuclear atypia, and increased mitotic activity⁷. In the present case, the large tumor size and the presence of extensive necrosis supported classification as a high-risk neoplasm, thereby justifying strict surveillance and a broader therapeutic discussion.

From a surgical perspective, complete resection remains the cornerstone of treatment for localized PEComas. Transurethral resection of the prostate plays a diagnostic and palliative role but should rarely be considered definitive in high-risk tumors. In selected cases, radical prostatectomy represents the strategy with the greatest potential for achieving durable local disease control⁹.

Although the majority of PEComas exhibit indolent behavior, a subset demonstrates malignant potential. When metastasis occurs, dissemination is predominantly hematogenous, most commonly involving the lungs, liver, and bones. Lymphatic spread appears to be uncommon but has been reported, particularly in tumors exhibiting adverse histopathological features such as large size, infiltrative growth pattern, high nuclear grade, increased mitotic activity, necrosis, and vascular invasion, as described by Folpe *et al.*⁷

In cases of advanced, recurrent, or metastatic disease, systemic therapy becomes necessary. The identification of dysregulation of the mTOR signaling pathway in PEComas has enabled the use of mTOR inhibitors, which have demonstrated consistent clinical activity and currently represent the main systemic therapeutic option for selected malignant tumors^{10,11}. Conventional cytotoxic chemotherapy has shown limited efficacy, and radiotherapy plays a restricted role, generally reserved for palliative purposes.

4. CONCLUSION

Primary prostatic PEComa is an exceptionally rare neoplasm that poses significant diagnostic challenges, as

its clinical and radiological presentation frequently mimics common urologic conditions, such as benign prostatic hyperplasia and epithelial neoplasms of the prostate and bladder. Recognition of this entity fundamentally relies on careful histopathological evaluation combined with a comprehensive immunohistochemical panel capable of excluding epithelial tumors and identifying the characteristic myomelanocytic profile. Accurate identification of PEComa is essential for appropriate risk stratification and therapeutic decision-making, as tumors meeting criteria for malignancy may exhibit aggressive behavior and a substantial risk of recurrence. Complete surgical resection remains the cornerstone of treatment, while targeted systemic therapies, including mTOR pathway inhibitors, represent relevant options in selected cases. Given the rarity and unpredictability of the biological behavior of these tumors, rigorous clinical follow-up and a multidisciplinary approach are crucial to optimize outcomes and expand current understanding of this uncommon entity.

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