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Urology

Rare Case Report of Urinary Bladder Paraganglioma: Lessons for Clinicians

Dr Omar Jendouzi[1]*, Dr Lamghari Aziz[1], Dr Aya Sobhi[1]

[1]Urology Department- Military Teaching Hospital Agadir
[2]Néphrology Department- Military Teaching Hospital Agadir

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***Corresponding author:** Dr Omar Jendouzi
Urology department- Military Teaching Hospital Agadir

Abstract **Case Report**

Background: Paraganglioma of the urinary bladder is a rare neuroendocrine tumor arising from chromaffin cells within the bladder wall. When functional, it may secrete catecholamines and present with characteristic symptoms such as hypertension, headache, palpitations, and sweating, which can be triggered by micturition or bladder distension. ***Case Presentation:*** Our patient, a 45-year-old male presented to the urology clinic with a 6-month history of intermittent episodes of severe headaches, palpitations, and profuse sweating immediately following micturition. Urinanalysis revealed a microscopic hematuria and ultrasound demonstrated a mass on the left lateral wall of the bladder. Elevated levels of 24-hour urinary fractionated metanephrines and normetanephrines confirmed the suspicion of a catecholamine-secreting tumor; A premedication with alpha-adrenergic blockade was started, then the patient was subjected to transurethral resection with complete excision of the mass. Histopathological examination came out to be paraganglioma. ***Conclusion:*** Paraganglioma of the urinary bladder, though rare, must be considered in the differential diagnosis of paroxysmal hypertension, especially when symptoms are related to micturition. Successful management requires a multidisciplinary approach involving urology, endocrinology, and anesthesiology, with rigorous preoperative alpha-blockade being the most critical step for a safe surgical outcome. Our patient remains disease-free and normotensive at 12-month follow-up.

Keywords: Paraganglioma, Urinary bladder, Case report.

INTRODUCTION

Paraganglioma of the urinary bladder (PUG) is an extremely rare neuroendocrine tumor originating from chromaffin cells within the bladder wall. It accounts for less than 0.06% of all bladder tumors. When functional, these tumors can secrete catecholamines, leading to characteristic symptoms such as paroxysmal hypertension, headache, palpitations, and sweating, often triggered by micturition or bladder distension [1].

Our patient, a 45-year-old male who presented with chef complaints of severe headaches, palpitations, and profuse sweating immediately following micturition with no urinary tract symptom. Ultrasound revealed a urinary bladder mass

A complete transurethral resection was done. Findings of histopathological analysis were consistent for Paraganglioma of the urinary bladder.

This case is reported due to its rarity and highlights the typical clinical presentation, diagnostic pathway, and successful surgical management of a functional PUG.

This case report has been reported in line with the CARE Criteria.

CASE PRESENTATION

A 45-year-old male presented to the urology clinic with a 6-month history of intermittent episodes of severe throbbing headaches, palpitations, and profuse sweating immediately following micturition. His blood pressure was significantly elevated during these episodes (up to 220/110 mmHg) but normalized between attacks. No reports of hematuria nor other lower urinary tract symptoms. Physical examination was unremarkable

Initial investigations revealed markedly elevated levels of 24-hour urinary fractionated

metanephrines (3500 nmol/day; reference range: < 1900 nmol/day) and normetanephrines (7800 nmol/day; reference range: < 3900 nmol/day), confirming the suspicion of a catecholamine-secreting tumor.

Urinalysis revealed the presence of red blood cells, while urine culture yielded no bacterial growth. Urinary system ultrasound was in favour of a left lateral mass in the bladder wall, measuring 1.19 cm × 1.69 cmx 1,63 cm [Figure 1] Metastatic evaluation was unremarkable, and urine cytology showed no malignant cells. One month postoperatively, the patient's plasma metanephrines and urinary vanillylmandelic acid (VMA) were within normal limits. Contrast-enhanced CT of the kidneys, ureters, and bladder showed no residual tumor, local invasion, or evidence of metastasis. The patient is being followed every three months with history-taking, physical examination, plasma and urinary metanephrines and cystoscopy. No tumor recurrence was noted up to 2 years.

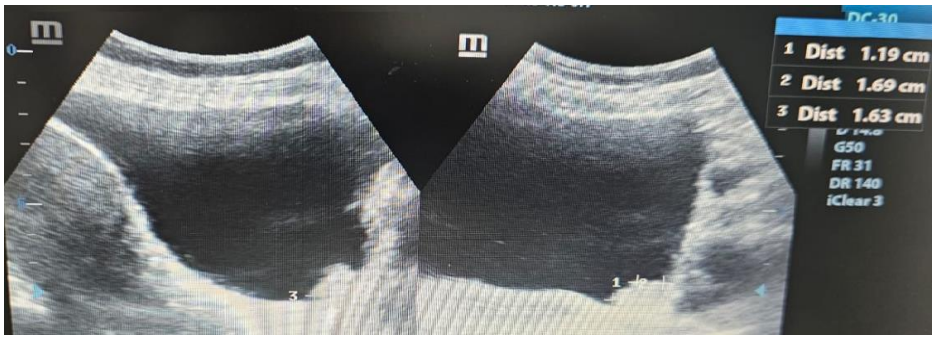

Fig. 1: Ultrasound scan demonstrated a mass on the left lateral wall of the bladder, measuring 1.19 cm × 1.69 cmx 1,63 cm

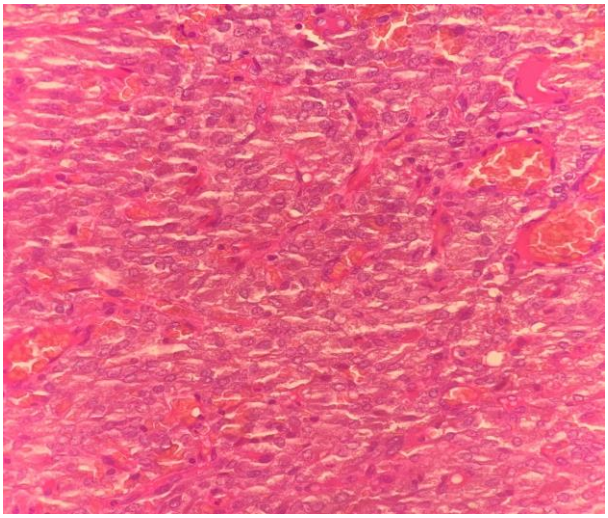

Fig. 2: Proliferation of cord-like architecture composed of epithelioid cells without major atypia or mitotic figures. (hematoxylin and eosin (H&E) stain, 20x)

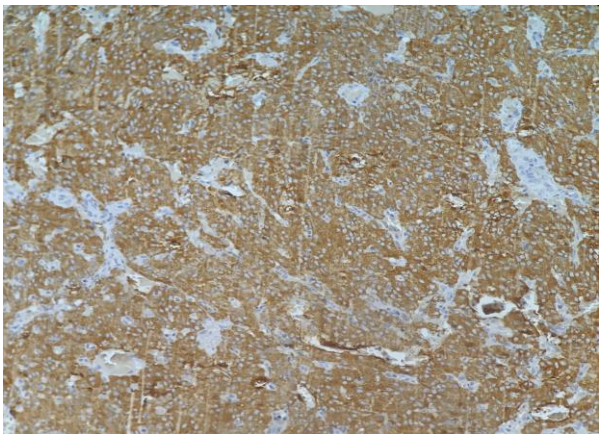

Fig. 3: Cytoplasmic expression of chromogranin

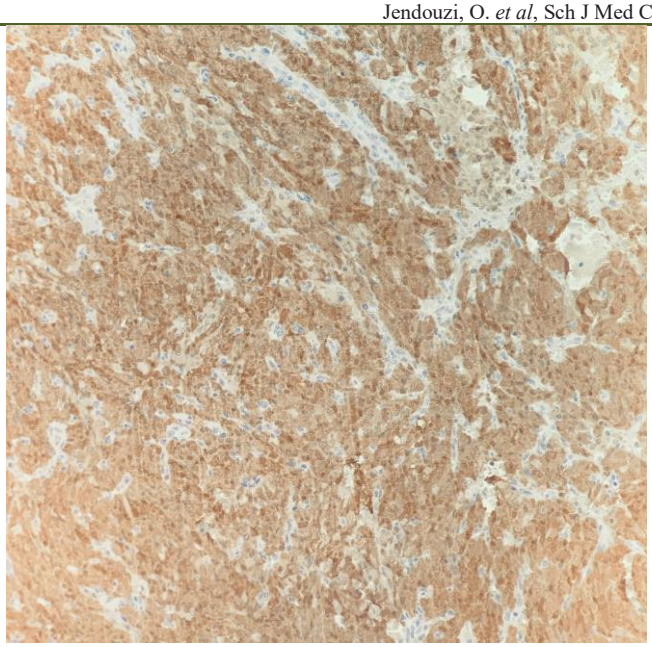

Fig. 4: Cytoplasmic expression of Synaptophysine

DISCUSSION

Paraganglioma of the urinary bladder is a neuroendocrine non epithelial neoplasm, extremely rare, accounting for approximately 0.06% of all bladder tumors and 6% of extra-adrenal pheochromocytomas [1]. Within the genitourinary tract, the bladder is the most frequently affected site (79.2%), followed by the urethra, pelvis, and ureter [2]. These tumors arise from chromaffin tissue of the sympathetic nervous system within the bladder wall and are most commonly situated at the dome or trigone of the bladder. They can be either nonfunctional or functional, secreting catecholamines [2,3]. Although most bladder paragangliomas are benign, 15-20% may exhibit malignant behavior, with the potential for local invasion, lymph node dissemination, and distant spread [2,3]. These tumors appear more frequently in women and generally present clinically in the third decade of life [4].

Functional tumors often present with catecholamine related symptoms. Patients may experience hypertensive crises accompanied by headache, palpitations, sweating, and flushing. Postmicturition hypotension and syncope are also commonly observed. These crises can be triggered by micturition, bladder overdistension, defecation, sexual activity, ejaculation, or bladder instrumentation [2,5,6]. Approximately 17% of bladder paragangliomas are hormonally nonfunctional and may remain asymptomatic [2]. Painless hematuria, although nonspecific, is reported in about 60% of cases [2,5,6].

Imaging plays a crucial role in diagnosis and staging. CT and MRI are effective in localizing primary tumors and detecting metastases, while 131I-MIBG scanning demonstrates high sensitivity and specificity for pheochromocytomas [4]. Functional imaging targeting catecholamine synthesis, storage, and secretion is particularly useful in monitoring recurrence or metastatic disease [2,4]. Biochemical evaluation of plasma and urine catecholamines is essential both preoperatively and during follow-up, with plasma metanephrines being more sensitive than urinary measurements [2,3,5].

Cystoscopic evaluation typically reveals a yellow, submucosal tumor, raising suspicion for paraganglioma [2]. Manipulation of the tumor can release catecholamines, potentially causing severe transient hypertension. Thus, biopsy or resection should be performed only under appropriate alpha-adrenergic blockade and preoperative volume expansion to minimize perioperative morbidity and mortality [2,3,6]. In our case, intraoperative hypertensive crises were avoided thanks to the preoperative diagnosis that was established.

Histopathologically, bladder paragangliomas are composed of large polygonal cells with abundant granular cytoplasm arranged in a Zellballen pattern, surrounded by a fibrous vascular network [1,6,7]. They can be misdiagnosed as urothelial carcinoma, particularly the nested variant, or other tumors such as granular cell tumors, metastatic large cell neuroendocrine carcinoma, or malignant melanoma [6,7]. Immunohistochemical staining is helpful in differentiation, with tumor cells positive for neuroendocrine markers (chromogranin, synaptophysin, neuron-specific enolase) and negative for urothelial markers (cytokeratin) [6,7]. Malignant behavior is difficult to ascertain histologically and is usually confirmed clinically by the presence of metastases [2,4,6].

Complete Surgical excision remains the mainstay of treatment. However, Premedication with an alpha-blocker is necessary to prevent hypertensive crises and arrhythmias during tumor manipulation. Partial cystectomy is preferred over transurethral resection when feasible, as most tumors involve the deep detrusor muscle [2,3,5]. In our case report, the tumor size was small, and did not invade the muscle layer transurethral resection was a safer approach. Recurrence is relatively common and does not necessarily indicate malignancy. Total cystectomy is reserved for large tumors or cases with lymph node involvement [2,3,5]. Bladder paragangliomas are generally resistant to chemotherapy and radiotherapy, although these modalities have been used in selected cases [8,9,10].

Given the risk of recurrence and metastasis, lifelong follow-up is recommended, including periodic history, measurement of plasma and urinary catecholamines, cystoscopy, and imaging (CT or 123I-MIBG scintigraphy) when symptoms or biochemical markers suggest recurrence [2]. but no consensus is clearly established because of the rarity of the tumour.

This case report highlight the need to consider bladder paraganglioma as a differential diagnosis in patients presenting with post-micturition symptoms and paroxysmal hypertension, even without typical symptoms like hematuria. Biochemical, imaging, and histopathologic correlation, avoid misdiagnosis and reduce morbidity related to an unexpected intraoperative hypertensive crisis.

Limitation of this case include the relatively short follow up period explaining the for more prolonged monitoring, especially with a conservative approach.

CONCLUSION

Bladder paraganglioma is a rare neuroendocrine tumor with variable clinical presentation, ranging from asymptomatic lesions to life-threatening catecholamine crises. Accurate diagnosis relies on biochemical tests, imaging, and histopathology. Surgical excision is the mainstay of treatment; transurethral resection may represent a safe approach.

Lifelong follow-up is essential due to the risk of recurrence and metastasis. Early recognition and multidisciplinary management are key to optimal patient outcomes.

Ethics Approval and Consent to Participate: Ethical approval was not required for this case report in accordance with local or institutional guidelines. Written informed consent was obtained from the patient for participation.

Consent for Publication: Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Availability of Data and Material: All data generated or analysed during this case report are included in this published article.

Competing Interests: The authors declare that they have no competing interests.

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