URINARY BLADDER PARAGANGLIOMA – CASE REPORT AND REVIEW OF THE LITERATURE

Rajesh P Shrivastava*, Jignesh Ashwin Gandhi** and Amay Manish Banker*

*Consultant Advanced Laparoscopic and Bariatric Surgeon, Shreeji Hospital Deheli, Bhilad Sanjan Road, Valsad, Bilad, Gujarat 396105, **Head of Unit and Associate Professor Department of General Surgery Seth G.S. Medical College KEM Hospital Parel, Mumbai, India. J Junior Resident, Department of General Surgery Seth G.S. Medical College KEM Hospital Parel, Mumbai, India.

ABSTRACT
Paragangliomas of the urinary bladder constitute a small part of all bladder tumors. Faced with a wide variety of presenting symptoms, they propose a diagnostic dilemma and lack of a definite preoperative diagnosis may result in inadequate pre-operative preparation with disastrous consequences. We present a case of urinary bladder paraganglioma wherein we could achieve definite diagnosis after a cystoscopy guided biopsy of the lesion. This enabled us to prepare the patient for a laparoscopic partial cystectomy adequately. The patient was discharged after an uneventful postoperative course.

KEYWORDS
Paraganglioma; urinary bladder; laparoscopic partial cystectomy; extra-adrenal pheochromocytoma; zellballen; hypertensive crisis.

Introduction
Paragangliomas or extra-adrenal pheochromocytomas arise from chromaffin tissue of the sympathetic nervous system in locations outside the adrenal gland. Paragangliomas of the urinary bladder account for only 6% of all extra-adrenal pheochromocytomas. [1] When asymptomatic, these tumours present a diagnostic challenge and lack of a definitive preoperative diagnosis may result in insufficient preparation, leading to an unexpected intra-operative hypertensive crisis. Here we present a 55-year-old female who presented with non-specific urinary complaints. The patient was diagnosed with a non-secretory bladder paraganglioma for which we successfully performed a laparoscopic partial cystectomy.

Case Report
A 55-year-old lady complaining of dysuria and lower abdominal pain for two months was admitted to our hospital. She had an unremarkable family history and no previous medical problems. Her vitals were stable; she was soft per abdomen and had no hematuria. Routine blood and urine tests were normal. Ultrasonography (US) of the urinary bladder showed a

Figure 1 A: The tumor cells grow in a nested, zellballen pattern surrounded by a fibrous network that is rich in blood vessels. B: The tumor cells are strongly positive for synaptophysin on immunohistochemistry.

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Associate Editor: Ivan Inkov (BG);
*Department of General Surgery Seth G.S. Medical College & KEM Hospital Parel, Mumbai, India. Email: amaybanker@gmail.com

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small hypoechoic mass from the anterior wall. A computed
tomography (CT) confirmed a rounded mass lesion 2.9 x 2.9 x
2.5 cm without any metastatic disease. Our patient underwent
cystoscopy and biopsy of the lesion. On histopathological exam-
ination (HPE) the cells with vesicular nuclei were arranged in
sheets and were strongly positive for chromogranin and synap-
tophysin on immunohistochemistry (IHC), confirming a urinary
bladder paraganglioma. (Figure 1) She was posted for a la-
paroscopy partial cystectomy with wide local excision of the
tumor. (Figure 2) The tumor involved the inner half of the mus-
cularis mucosa, and the surgical margins were free of the tumor
on pathological evaluation.

Discussion
Bladder paraganglioma is a rare entity with a female pre-
ponderance (3:1) during the second to fourth decade of life.1
Catecholamine secreting paragangliomas may mimic a hyper-
functioning adrenal pheochromocytoma. Their position within
the bladder results in a characteristic symptom complex of
headache, dizziness, sweating and palpitation after micturition
or overdistension of the bladder. Systemic catecholamine re-
lease which occurs due to the increased pressure secondary to
bladder contraction may explain these sympathomimetic attacks.
[2]Non-functional tumours may be asymptomatic, making them
a diagnostic challenge. So, when a paraganglioma of the blad-
ner is suspected, catecholamine level and its metabolites such
as metanephrine and vanillylmandelic acid (VMA) secretion
in either the blood or urine is measured. In a non-functional
tumour, the levels of these metabolites may be normal. Since
our patient was asymptomatic, the authors felt these laboratory
investigations to be costly and unnecessary.

US, CT or a magnetic resonance imaging (MRI) may help lo-
calise the tumour. In contrast to an adrenal pheochromocytoma,
bladder paragangliomas are likely to be homogenous on a T2
weighted MRI signal. 1131-methylidobenzylguanidine (1131-
MIBG) and positron emission tomography (PET)-CT help to
evaluate the functional status of the disease and look for distant
metastasis. [1] The role of cystoscopy and biopsy is controversi-

al for the fear that it may provoke a hypertensive crisis. However,
in centres without nuclear imaging access, cystoscopy after ade-
quate hydration and alpha neurogenic blockade may be required
to confirm the diagnosis.

The tumour is well circumscribed and turns black when
placed in a Zenker’s fixative, indicating a positive chromaffin
reaction. On HPE, paraganglioma cells are arranged in a Zell-
ballen pattern and are surrounded by a fibrous network rich
in blood vessels (Figure 1). On IHC analysis, the chief cells are
positive for neuroendocrine markers like chromogranin, and
synaptophysin and the sustentacular cells are negative for cy-
tokeratin, which differentiates it from urothelial carcinomas. [3]
Metastasis to distant organs remains the only widely accepted
proof of malignancy.

Treatment modalities include a transurethral resection, a par-
tial or total cystectomy with pelvic lymph node dissection in
the presence of proven metastasis. [3-5] Since a hypertensive
crisis may be precipitated during surgery; appropriate preopera-
tive preparation is of paramount importance. An α-adrenergic
blockade should be ensured before the β-adrenergic blockade,
to prevent the risk of unopposed adrenergic stimulation. A lib-
eral salt diet and adequate hydration are advised to expand the
contracted blood volume.

In our case, the large size of the tumour prompted us to
perform a laparoscopic partial cystectomy. There were no fluctua-
tions in the intra-operative blood pressure, and the resection
margins were tumour free. The urinary bladder was closed in
two layers, and the patient was discharged after an uneventful
postoperative course.
Conclusion
Urinary bladder paragangliomas are a rare entity which may pose a diagnostic and therapeutic challenge when the classical signs of a sympathomimetic attack are absent. Awareness of this entity amongst the urologists and pathologists is of paramount importance, to ensure adequate preoperative correction of the catecholamine excess and prevent surgical misadventures.

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Conflict of interest
The authors declared that this project was done independently without any conflict of interest.

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