URINARY BLADDER PARAGANGLIOMA – CASE REPORT AND REVIEW OF THE LITERATURE

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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

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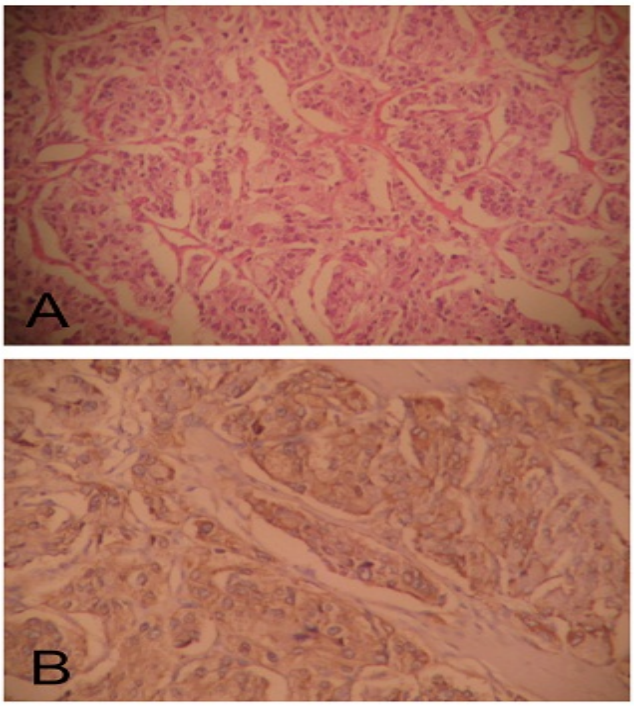

Figure 1 A: The tumor cells grow in a nested, zellballen pattern surrounded by a fibrous network that is rich in blood vessels. B: The tumor cells are strongly positive for synaptophysin on immunohistochemistry.

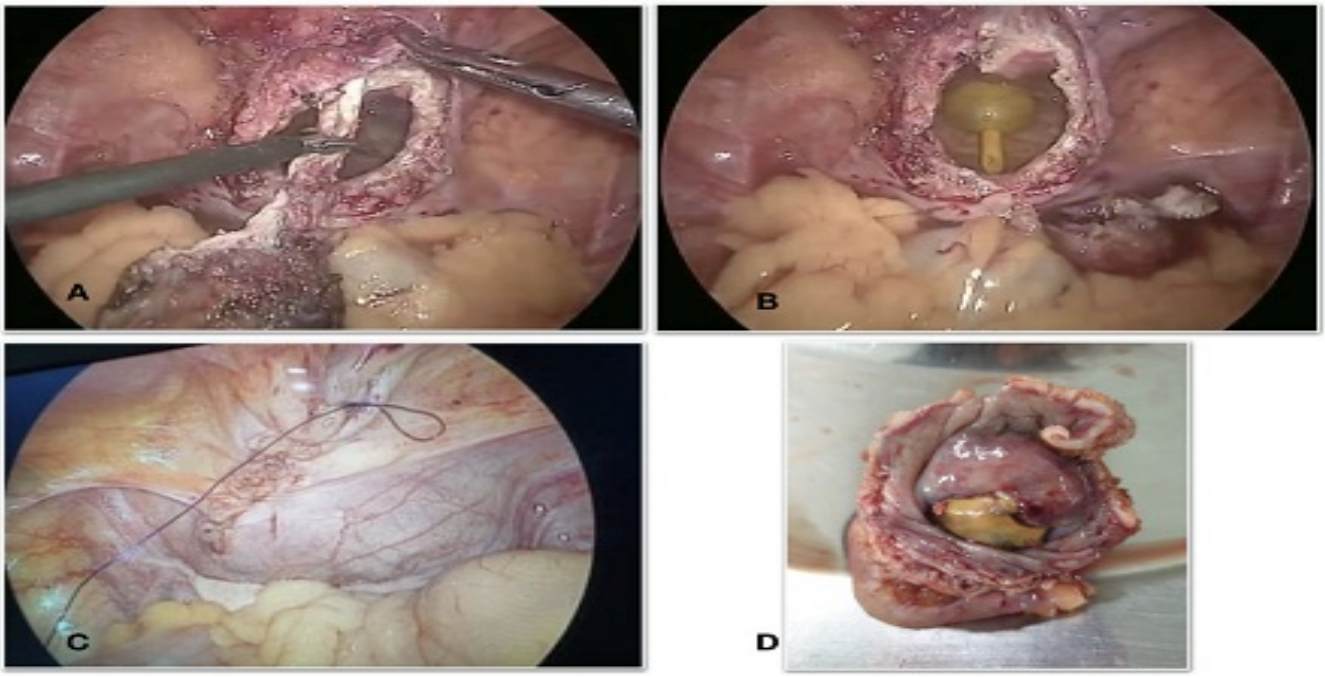

Figure 2 A: Laparoscopic view of wide local excision of the tumor B: Laparoscopic view of partial cystectomy with wide local excision of the tumor C: Remnant urinary bladder closed in two layers D: Gross tumor specimen

small hypoechogenic 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 examination (HPE) the cells with vesicular nuclei were arranged in sheets and were strongly positive for chromogranin and synaptophysin on immunohistochemistry (IHC), confirming a urinary bladder paraganglioma. (Figure 1) She was posted for a laparoscopy partial cystectomy with wide local excision of the tumor. (Figure 2) The tumor involved the inner half of the muscularis mucosa, and the surgical margins were free of the tumor on pathological evaluation.

Discussion

Bladder paraganglioma is a rare entity with a female preponderance (3:1) during the second to fourth decade of life.1 Catecholamine secreting paragangliomas may mimic a hyperfunctioning 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 release 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 bladder 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 localise the tumour. In contrast to an adrenal pheochromocytoma, bladder paragangliomas are likely to be homogenous on a T2 weighted MRI signal. I131-methyliodobenzylguanidine (I131-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 controversial for the fear that it may provoke a hypertensive crisis. However, in centres without nuclear imaging access, cystoscopy after adequate 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 Zellballen 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 cytokeratin, 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 partial 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 preoperative 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 liberal 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 fluctuations 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.

References

1. Xu D-F, Chen M, Liu Y-S, Gao Y, Cui X-G. Non-Functional Paraganglioma of the Urinary Bladder: A Case Report. Vol 4.; 2010. http://www.jmedicalcasereports.com/content/4/1/216.
2. Ranaweera M. Bladder paraganglioma: A report of case series and critical review of current literature. World Journal of Clinical Cases. 2014;2(10):591. doi:10.12998/wjcc.v2.i10.591
3. Dwethi G, Vikas C, Ramesh T. Urinary bladder paraganglioma. Applied Radiology. 2019;48(3):48A-48C.
4. Palla AR, Hogan T, Singh S. Malignant Paraganglioma of the Urinary Bladder in a 45-Year-Old Woman. Clinical Advances in Hematology & Oncology. 2012;12(10):836-841.
5. Menon S, Goyal P, Suryawanshi P, et al. Paraganglioma of the urinary bladder: A clinicopathologic spectrum of a series of 14 cases emphasizing diagnostic dilemmas. Indian Journal of Pathology and Microbiology. 2014;57(1):19-23. doi:10.4103/0377-4929.130873