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Case Report .

Non-Functional Bladder Paraganglioma: A Case Report

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ABSTRACT

Paragangliomas of the urinary bladder are rare extra-adrenal tumours. A 45-year-old male complaint of single episode of painless gross hematuria without any micturitional attacks. Ultrasonography and Computed tomography scan revealed a well-defined solid mass in urinary bladder wall. Cystoscopy revealed a protruding lobulated brownish bladder mass with normal smooth mucosa for which trans urethral resection of bladder tumour (TURBT) was done. Biopsy and Immunohistochemistry confirmed the diagnosis of paraganglioma. The current report presents a non-functioning paraganglioma of the urinary bladder and emphasises on having high risk of suspicion on the abnormal cystoscopic finding for its diagnosis.

Key-words: Paraganglioma, Pheochromocytoma, Bladder, Diagnosis, Treatment.

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INTRODUCTION

Paragangliomas of the urinary bladder are rare extra-adrenal tumors of the autonomic nervous system which accounts for less than 0.06% of all bladder neoplasm and less than 1% of all pheochromocytoma.[1,2] Often they tend to be functional and typically cause micturitional attacks (hypertensive crisis during micturition). However, 27% of paragangliomas of the urinary bladder are hormonally non-functional.[2,3] These tumors typically lack the malignant features like mitoses and cellular dissociation, yet they are deemed malignant as they have the capacity to invade.[4] We report a case of paraganglioma of the urinary bladder that presented to us without any typical symptoms and thus was not suspected before the operation.

CASE HISTORY

A 45-year-old male came with the complaint of single episode of painless gross hematuria. He denied history of paroxysmal hypertension, postural hypotension or sudden-onset blurred vision. He had no abdominal symptoms, no previous medical problems, had been in good health and had no specific positive past medical, drug or family history. His vitals were in the normal range.

Routine haematological examination, biochemical tests and urine tests were within normal limits and physical examination showed no evidence of hypertensive disease. Ultrasonography revealed an abnormal mass on the right lateral wall of the urinary bladder and computed tomography (CT) showed a solitary well defined intensely enhancing soft tissue density lesion of size approximately 14 x 19 x 11 mm in right lateral wall region, away from ureterovesical junction protruding in lumen [fig 1]. No sign of any metastatic disease was found on ultrasound examination or CT scans. Cystoscopy revealed a protruding lobulated brownish bladder mass with normal mucosa for which TURBT was done. The procedure was uneventful with no occurrence of hypertension or massive bleeding. Postoperatively recovery was good and he did not suffer from paroxysmal hypertension during hospital stay.

Histological examination of the specimen elucidated the diagnosis of paraganglioma. Microscopically, tumour was submucosally situated and was covered with normal urothelium. There was no muscle invasion. The tumour cells comprising of round or polygonal cells were arranged in characteristic compact cell nests (Zellballen) pattern bounded by a delicate fibrovascular stroma. There was no cytoplasmatic inclusion or pleomorphism, Necrosis or mitosis was not seen. Immunostains were positive for chromogranin A and S-100 protein further supporting the diagnosis of paraganglioma. Patient is doing well, with complete resolution of his symptoms and is under follow up.

DISCUSSION

Extra-adrenal Paraganglioma originates from the chromaffin tissue of the sympathetic nervous system and accounts for less than 10% of all pheochromocytoma.[5] Majority of them are found in the retroperitoneum, thorax, and neck. The most common site in the genitourinary tract is urinary bladder followed by the urethra, pelvis and ureter. Bladder pheochromocytoma occurs mainly during the third-fourth decade of life and with slight preponderance in females as compared to males.[1,2]

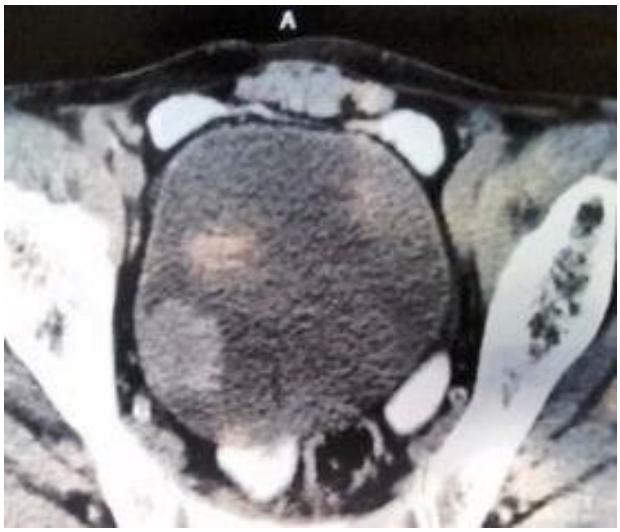

Fig 1: Computed tomography (CT) showed a solitary well defined lesion in right lateral wall region

The most common presenting symptoms of bladder paraganglioma are hypertensive attacks and hematuria accompanied by headache, palpitations, seating and hot flushes which usually occurs due to increased catecholamine release during detrusor contraction and are mainly provoked by activities like micturition, overdistension of the bladder, sexual activity, ejaculation, defecation or bladder instrumentation.[1,2,4] However, 27% of bladder paragangliomas are hormonally non-functional.[2,3] Elevated serum and urinary catecholamine levels confirms the diagnosis of a functionally active paraganglioma. However, not all patients have typical test results if blood or urine samples are not collected at the occurrence of typical symptoms.[5] As there was no history of hypertension, headache or flushing in this patient, we did not considered endocrine screening for paraganglioma in his management.

Imaging studies are used to determine the size, location, and local extent of the tumor. MRI is better than CT in the evaluation of the paragangliomas.[5] I^{131} Metaiodobenzylguanidine (MIBG) is the imaging study of choice for diagnosing and localizing extra-adrenal paraganglioma with 90% sensitivity and about 100% specificity. However, Positron emission tomography (PET) due to its higher spatial resolution is even more accurate than MIBG.

The tumors in cystoscopy appear as protruding globular submucosal masses with an intact surface and continuous mucosa.[3,5] Biopsy is not considered as a diagnostic method due to low positive rate, risk of bleeding and may provoke a hypertensive crisis. Histological features are similar to adrenal pheochromocytomas. The tumor cells usually show nested, zellballen pattern surrounded by delicate fibrovascular stroma.[4] Immunohistochemical staining is mandatory for a definitive diagnosis. Chromogranin, synaptophysin and NSE helps to identify neuroendocrine cells and neural tissue.[2,4] The differential diagnosis for paraganglioma includes urothelial carcinoma, malignant melanoma, metastatic renal cell carcinoma, prostate carcinoma, carcinoid tumors, and granular cell tumour.[4] Treatment modalities include transurethral resection and cystectomy (partial or complete).[1,2] Laparoscopic partial cystectomy has become the treatment of choice but due to the multilayer involvement of the bladder wall, open partial cystectomy is still recommended. TURBT can be a viable minimally invasive procedure for paraganglioma of the bladder if the tumour is small, localised and non-invasive with well controlled preoperative blood pressure and absence of "micturition attacks". Approximately 10% of bladder pheochromocytomas are malignant. Therefore, life-long follow-up is necessary to diagnose any localized recurrences and metachronous metastases.[5] Prognosis of bladder paraganglioma is similar to that of adrenal pheochromocytoma.[1]

CONCLUSION

Diagnosis of a bladder paraganglioma is difficult and non-functioning variety is even rarer and not easily recognized however it should be considered in patients with unusual presentations and unusual appearances on cystoscopy. Cystoscopic finding of "a yellowish brown lobulated protruding mass with normal and intact mucosa" appearance should raise the suspicion of a paraganglioma adenoma. This will facilitate prompt diagnosis and treatment and especially prevent intraoperative complications due to pheochromocytoma which may be life threatening. The results are usually satisfactory after transurethral resection.

REFERENCES

1. J. B. Thrasher, R. R. Rajan, L. M. Perez, P. A. Humphrey, E. E. Anderson. Pheochromocytoma of urinary bladder: contemporary methods of diagnosis and treatment options, Urology 1993, 41(5), pp. 435–439.

2. Beilan et al. Pheochromocytoma of the urinary bladder: a systematic review of the contemporary literature. BMC Urology 2013; 13:22.

3. Naqiyah I, Rohaizak M, Meah FA, Nazri MJ, Sundram M and Amram AR. Phaeochromocy-toma of the urinary bladder. Singapore Med J 2005; 46: 344-346.

4. Zhou M, Epstein JI, Young RH. Paraganglioma of the urinary bladder: A lesion that may be misdiagnosed as urothelial carcinoma in transurethral resection specimens. Am J Surg Pathol 2004;28:94-100.

5. Whalen RK, Althausen AF, Daniels GH. Extra-adrenal pheochromocytoma. J Urol. 1992;147:1–10.

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