Case report

Micturition syncope secondary to urinary bladder paraganglioma

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SUMMARY

Paraganglioma of the bladder is a rare tumour accounting for less than 0.06% of all urinary bladder tumours and has varied presentations. It may present with clinical symptoms of phaeochromocytoma, may be non-functioning and asymptomatic or may present with haematuria. Hence, paragangliomas are occasionally misdiagnosed, and this results in unanticipated intraoperative hypertensive crisis. We present the case of a 44-year-old woman with urinary bladder paraganglioma who presented with young onset hypertension, recurrent micturition syncope with prior history of coronary artery disease and stroke. She was stabilised preoperatively with alpha blocking agents and subsequently underwent successful transurethral resection of the same.

BACKGROUND

Bladder paraganglioma is a rare clinical entity and may not present with the typical triad of headache, palpitations and sweating. This case highlights the classical presentation of bladder paraganglioma in a middle-aged woman who presented with recurrent episodes of micturition syncope.

CASE PRESENTATION

A 44-year-old woman presented to our centre for the evaluation of recurrent episodes of syncope since past 10–12 years. On further inquiry, she recalled that these episodes were usually precipitated by micturition. She had been diagnosed with hypertension at the age of 28 years and her clinical course thus far was complicated by one episode of cerebrovascular accident (exact medical details of which were not available) and a recent myocardial infarction. A detailed evaluation at her hometown had also revealed the presence of a bladder tumour for which she had undergone transurethral resection followed by chemotherapy and radiotherapy. There was history of hypertensive crisis with flash pulmonary oedema during the bladder tumour excision. There was no history of haematuria. There was no history suggestive of proximal myopathy or quadriparesis. There was no history of thyroid disease, bone pain, fractures or recurrent renal calculi. Histopathological examination of the excised tumour was reported elsewhere to be a transitional cell carcinoma of the urinary bladder.

At presentation, her blood pressure was 126/80 mm Hg on two antihypertensive medications. Ophthalmoscopic examination revealed grade 3 hypertensive retinopathy bilaterally. Her breast and thyroid examination were normal. Cardiovascular examination revealed a third heart sound. There was no clinical evidence of Cushing's syndrome or features suggestive of acromegaly. Per-vaginal examination revealed a nodular, indurated area on the anterior vaginal wall.

INVESTIGATIONS

Her haemoglobin level was 117 g/L (N: 120–150 g/L). ECG showed T wave inversions in aVL and in leads V3–V6. Echocardiography showed left ventricular systolic dysfunction, regional wall motion abnormalities and an ejection fraction of 37%. Cystoscopy showed a 2×1 cm lesion with necrotic area at 5 o'clock position just proximal to the bladder neck. On CT scan of the abdomen and pelvis, an enhancing lobulated soft tissue density lesion was seen in the base of the bladder measuring 32×33×30 mm involving the left vesicoureteric junction (figure 1). Iodine-131 meta-iodo benzyl guanidine (MIBG) scan was suggestive of a functioning neuroendocrine tumour of the urinary bladder (figure 2). Urine normetanephrine level was elevated at 1386 µg/24 hours (N: <600 µg).

DIFFERENTIAL DIAGNOSIS

Recurrent episodes of micturition syncope in a young individual with chronic hypertension and end organ damage in the presence of a bladder tumour suggested the possibility of a paraganglioma. This was further established by the MIBG scan and the elevated levels of 24 hours urine metanephrine.

TREATMENT

As her clinical presentation and imaging features were suggestive of functioning bladder paraganglioma, she was started on an alpha adrenergic receptor blocking agent with prazosin, the dose of which was gradually increased to ensure sufficient alpha blockade. Betablockers were continued. Thereafter, she underwent an excision biopsy of the recurrent bladder tumour, which was consistent with paraganglioma.

OUTCOME AND FOLLOW-UP

After the procedure, she was kept under regular follow-up. She did not have further episodes of syncope and her blood pressure was well controlled. She is being continued on antiplatelets, statins, carvedilol and diuretics for her cardiac dysfunction.

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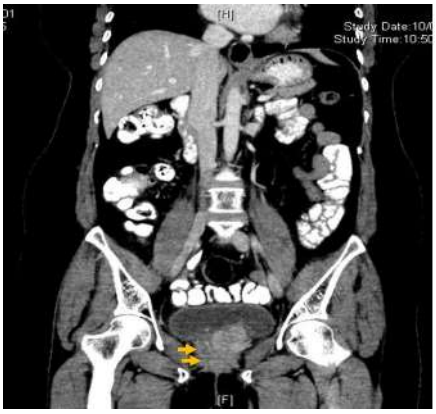

Figure 1 CT scan of abdomen and pelvis showing a mass in the urinary bladder.

DISCUSSION

Paraganglioma, which is also referred to as extra-adrenal phaeochromocytoma, is an uncommon type of neuroendocrine neoplasm and it accounts for about 10% of all pheochromocytoma.[1] The most common site for paraganglioma is the organ of Zuckerkandl at the distal aorta or aortic bifurcation. Less commonly, they are observed in the head, neck, thorax and bladder.[2] In the genitourinary tract, urinary bladder is the most common site for paragangliomas (79.2%). Other sites involved are the urethra (12.7%), pelvis (4.9%) and ureter (3.2%).[3] The first case of a paraganglioma of the bladder was described by Fernandez *et al*.[4] Bladder paragangliomas account for less than 0.06% of all urinary bladder tumours and 6% of all extra-adrenal phaeochromocytomas.[5] The presentation of paragangliomas of the bladder may be varied. It may present with the clinical symptoms of phaeochromocytoma such as headaches, palpitations and fainting as a result of the induction of catecholamine release from functional bladder paragangliomas when urinating or can present with the clinical symptoms of bladder tumours, such as haematuria.[6] However, among the urinary bladder paragangliomas, 10%–15% are non-functioning and a further 10% have hormonal activities that do not manifest clinically.[7] Hence, these paragangliomas are occasionally misdiagnosed, and surgery performed to remove the paragangliomas may result in an intraoperative hypertensive crisis.

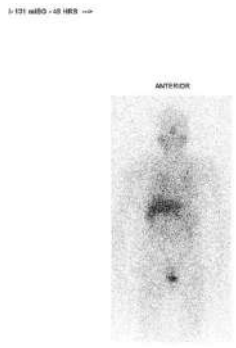

Figure 2 MIBG scan showing a persistent uptake in the urinary bladder region. MIBG, meta-iodo benzyl guanidine

Although the abovementioned patient was diagnosed with young onset hypertension with history of multiple episodes of micturition syncope, these symptoms were overlooked at her native place, and was managed as a case of probable transitional cell carcinoma of the bladder.This resulted in the hypertensive emergency causing flash pulomary oedema during her previous surgery. In patients who are suspected to have paraganglioma, it is necessary to stabilise hypertension prior to surgical treatment with alpha blocking agents. In the present case, partial cystectomy was performed with adequate alpha-receptor blockade and hence was uneventful.

Learning points

- Bladder paraganglioma is a rare tumour, but needs to be considered as an important differential diagnosis of micturition syncope.
- Not all bladder paragangliomas may present with classical clinical symptoms and hence a high index of suspicion needs to be maintained in identifying these lesions.
- Preoperative preparation with alpha blockers is necessary to prevent hypertensive crisis during surgery.

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