Case Report

Paraganglioma of urinary bladder: An unusual presentation. Pitfalls in diagnosis and treatment

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Abstract

Paragangliomas of urinary bladder are extremely rare which usually present with characteristic clinical picture in about half of the cases. We report a case where the presentation was unusual, leading to initial misdiagnosis corrected on histological review.

Keywords: Paraganglioma, Urinary bladder, Gross haemeturia, Immunohistochemistry.

Introduction

Paraganglioma of the urinary bladder is a rare tumour, accounting for less than 0.06% of all vesical tumours and less than 1% of all the pheochromocytomas.[1] About 98% of the paragangliomas are located in the abdomen, 90% of these are in the adrenal medulla with 10% extra-adrenal sites[2] extending from neck to the pelvis. Paraganglioma arise from the chromaffin tissue of the sympathetic nerves of the bladder

wall. As they are rare tumours there are pitfalls in their diagnosis and no standard treatment protocols are available.

Case Report

A 32-year-old man presented to a local hospital with painless gross haematuria of four months. He had no other irritative lower urinary tract symptoms. His medical history and examination was unremarkable as documented. Ultrasonogram of abdomen and pelvis performed revealed a broad based mass within the bladder. Transurethral resection of the bladder mass was undertaken. His intra- and post-operative course remained uneventful. Originally the histology of the resected mass was reported as transitional cell carcinoma (TCC) with muscle invasion. Patient was then referred to us for further management. An MRI of abdomen and pelvis was performed and there was no residual/recurrent local disease and no synchronous lesion was identified elsewhere. As per standard hospital procedures, we had a histological review of the mass specimen with immunohistochemical staining. This confirmed that it was a paraganglioma. A MIBG (meta-iodobenzylguanidine) scan was normal and no other lesion was identified. Serum catecholamines were reported within normal range and we attributed it to the fact that the tumour was inactive. The patient is on regular follow-up, free of any recurrence. Patient was followed-up every 3 months for the first 6 months and is now on a 6 monthly follow-up. He is symptoms free and cystoscopy and imaging findings are unremarkable.

Discussion

There are fewer than 200 reported cases of urinary bladder paragangliomas worldwide[3] and this is the first from Pakistan.

Paragangliomas of the urinary bladder often present with attacks of hypertension during micturation that are typically provoked by detrusor muscle activity along with haematuria.[4] The characteristic symptoms of paragangliomas include one or more of the triad of the symptoms of headache, sweating, or palpitations. Their absence virtually excludes the diagnosis.[5] Other symptoms like dysuria and supra-pubic pain may also be present. As in our case typical symptoms are absent because not all vesical paragangliomas are hormonally active.[6]

The histological differential diagnosis of paraganglioma includes nested variant of TCC, inverted papilloma and nephrogenic adenoma. Immunohistochemical analysis is useful in distinguishing paraganglioma from nested variant of TCC.[7] Nested variants of TCC express cytokeratin and other epithelial markers. Paragangliomas express neuroendocrine markers such as chromogranin (Figure-1) but are non-reactive for cytokeratin (Figure-2), as demonstrated in this reported case. Nephrogenic adenoma has a papillary component with prominent tubular growth pattern.[8] Inverted papilloma lacks a nested architecture.[9]

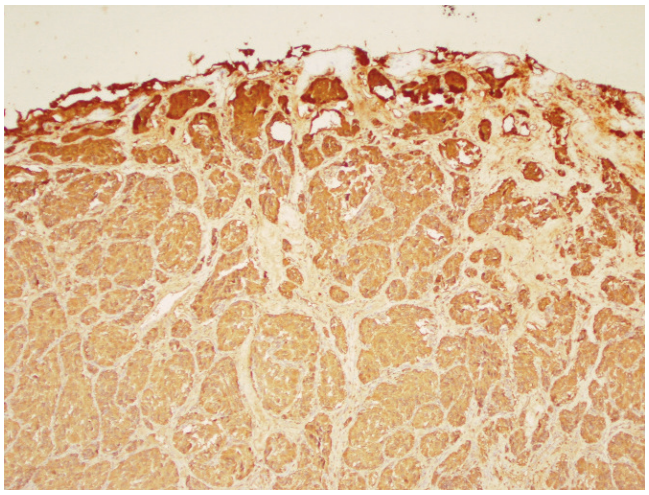

Figure-1: 40x Micrograph of the resected urinary bladder mass staining positive for chromogranin immunohistochemical stain.

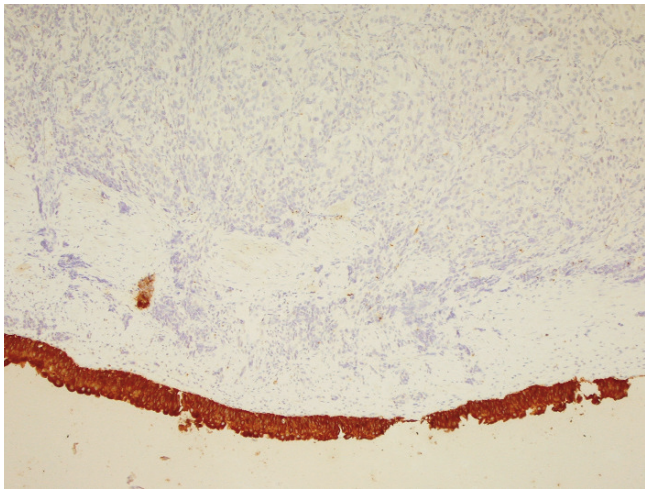

Figure-2: 40x Micrograph of the lesion showing negative cytokeratin immunohistochemical staining.

Most of the paragangliomas (~90%) are benign and can be cured by surgical excision alone. Extensive literature review shows that in most cases partial/segmental cystectomy was performed as primary surgical treatment of vesical paragangliomas[10] but there are also reported cases, though far fewer in number, in which transurethral resection was performed as the sole surgical procedure.[11] Owing to its rarity, it is difficult to ascertain the optimal treatment modality but partial cystectomy has been preferred over transurethral resection in cases where the diagnosis was confirmed preoperatively. Tumour cells infiltrate between muscle fibers makes it less amenable to transurethral resection for complete

eradication and increases the chances of local recurrence.

Precautions need to be taken to avoid complications during tumour manipulation as transient hypertensive crisis can occur, leading to serious consequences.

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