

Urinary Bladder Paraganglioma: An Unusual Site of Presentation

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Case Report

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Abstract: Urinary bladder Paraganglioma are extremely rare tumors arising from the sympathetic innervations of the bladder. These tumors account for 0.06% of urinary bladder tumors and can occur in all age groups. Most common symptom is hematuria simulating often bladder tumors. Preoperative diagnosis is important, failure of which may lead to serious consequences intraoperatively. We report a case paraganglioma of the urinary bladder occurring in 48 years old woman.

Keywords: Urinary bladder, Paraganglioma, Bladder tumor.

INTRODUCTION

Paraganglioma of the urinary bladder arise from the chromaffin cells derived from embryonic rests. Tumors of chromaffin cells in the adrenal medulla are termed as pheochromocytoma. Tumors occurring at the extra adrenal sites from these cells are termed as paragangliomas. Extra adrenal paragangliomas account for 10% of tumors of chromaffin cells and occur intraabdominally along the sympathetic chain. These tumors account for 0.06% of all the urinary bladder neoplasms [1].

Paraganglioma of the urinary bladder are most commonly situated at the trigone or dome of the bladder [2]. 55-60% of the cases present with haematuria which is not specific for paraganglioma. Clinical presentation of functional tumors includes intermittent hematuria, paroxysmal hypertension and the attacks precipitated by micturition [3]. In our case patient presented with hematuria, but during operative procedure developed hypertensive crisis.

CASE HISTORY

A 48 years old woman presented with history of lower abdominal pain, hematuria & dysuria since 1 day. There were no other symptoms and no previous history of urological interventions. Physical examination & laboratory investigations were unremarkable. Ultrasound abdomen showed ill-defined heterogeneously hyperechoic lesion measuring 3.9x3.6x3.4cm arising from base of bladder with a calcified peripheral rim. Lumen of the urinary bladder was filled with hyperechoic foci suggestive of blood clots. Mass was showing significant vascularity on CDFI.

CECT abdomen after giving IV contrast revealed a well-defined lobulated oblong shaped lesion measuring 3.4x3.4x3cm in the right paramedian base of bladder with its broad base attached to the posterior well associated with large blood clots within the bladder. The lesion showed irregular incomplete

calcific rim with well-maintained fat planes with surrounding pelvic structure. Diagnosis of inflammatory pseudotumor was suggested on imaging. Cystoscopy revealed submucosal well vascularized mass on the base of the bladder. Patient underwent transurethral resection of bladder tumor (TURBT) and biopsy was sent to the pathology department for histopathological examination. During the procedure of TURBT patient developed tachycardia and hypertension.

Microscopic examination of TURBT specimen showed fibro collagenous tissue with round to polygonal tumor cells arranged in nesting pattern or Zellballen pattern separated by fibro vascular septa (Figure 1). Tumor cells have round to oval nuclei with clumped chromatin and with moderate to abundant eosinophilic granular cytoplasm (Figure 2). Immunohistochemistry showed chromogranin positivity of tumor cells (Figure 3).

Later total cystectomy was performed. Cystectomy specimen was measuring 8X7.5X5cms and tumor was measuring 3.2X3X3cm at the base of the bladder. Grossly tumor is yellow coloured showing

infiltration into the muscle layer. (Figure4). Microscopy showed polygonal tumor cells infiltrating the muscularis.

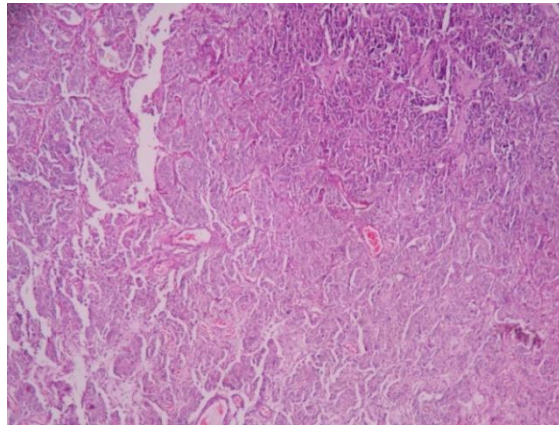


Fig-1: Tumor cells arranged in nesting (Zell ballen) pattern separated by their fibrous septa (H&E, X100)

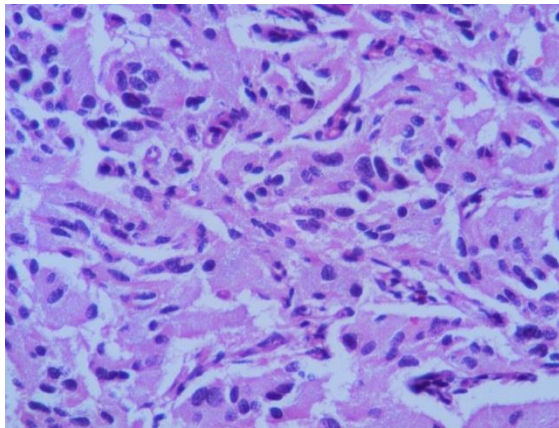


Fig-2: Tumor cells having abundant granular cytoplasm (H&E, X400)

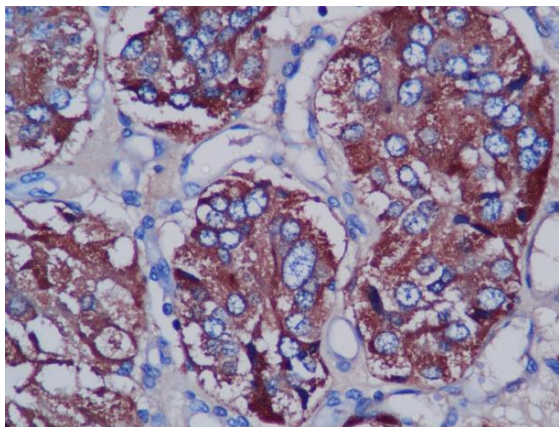


Fig-3: Tumor cells showing Chromagranin A positivity (Chromogranin, X400)

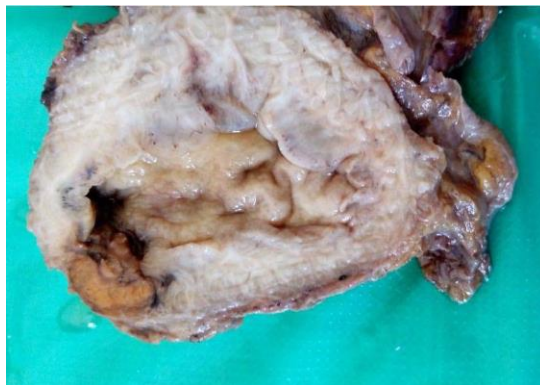


Fig-4: Bladder showing yellow colored tumor at the base of the bladder

DISCUSSION

Paraganglioma of the urinary bladder is the very rare tumor and account for 6% of extra adrenal pheochromocytomas [4]. These tumors arise from chromaffin cells of the sympathetic nervous system in the urinary bladder wall. Most common location of the tumor is trigone of the bladder near the ureteral orifices. Other locations are the dome of the bladder & lateral walls. They occur in the third decade of life with female predominance [5].

The most common site in the genitourinary tract is the urinary bladder (79.2%). Other sites in the genitourinary tract involved in the decreasing order of frequency are the urethra (12.7%), pelvis (4.9%) & ureter (3.2%) [6].

Paraganglioma occurring in the bladder may be functional or nonfunctional. These tumors are usually benign, but malignant behavior is seen in 15-20% of these tumors [7]. Patients with nonfunctional tumors (17%) are mostly asymptomatic or many present with haematuria which is the feature of other bladder tumors. Functional tumors may have the symptoms presenting hypertensive crisis like headache, hot flushes and palpitations and sweating. These symptoms result from excessive secretion of catecholamines which may be provoked by over distension of the bladder, micturition, defecation, ejaculation, sexual activity or bladder instrumentation [8].

Magnetic resonance imaging (MRI) & computer tomography (CT) scanning are useful for localization of the primary tumour & metastatic deposit. However high sensitivity & specificity for detection of Pheochromocytoma has been noted by scanning with ¹³¹Iodine metaiodine benzylguanidine[9]. Assessment of catecholamine levels in urine and /or plasma is important in symptomatic patients as initial work up and in follow up of patients.

On cystoscopy tumor appears as yellowish submucosal mass. During operative procedures catecholamines can be released on tumor manipulation and can lead to intra operative hypertensive crisis. In

our case also there was hypertensive crisis while performing TURBT.

Histopathological examination reveals tumor with polygonal cells having abundant granular cytoplasm. Tumor cells are arranged in nesting pattern and Zellballen pattern surrounded by delicate fibrovascular septa. Other bladder tumors which histologically resemble paraganglioma are granular cell tumors of bladder, urothelial carcinoma nested variant and malignant melanoma. These tumors can be differentiated by immunohistochemical analysis. Paraganglioma are immunohistochemically positive for synaptophysin, chromogranin & Neuron Specific Enolase but negative for cytokeratin which is urothelial marker [10].

Treatment of the choice is surgical excision. Partial cystectomy is preferred if the tumor is diagnosed pre operatively. However TURBT is not sufficient if the tumor extends into the deeper muscle layer. Total cystectomy is treatment of choice when there is distant metastasis to lymphnode or bladder preservation is not possible. Bladder paraganglioma are radio resistant and chemoresistant tumors with high recurrence rate. Hence lifelong follow up is required with yearly measurement of urinary / plasma catecholamines & cystoscopy.

CONCLUSION

Paraganglioma of urinary bladder is a rare tumor which requires preoperative diagnosis to avoid serious intraoperative consequences of hypertensive crisis. These tumors simulate bladder carcinoma on clinical presentation. Histopathologically paragangliomas can be diagnosed by using immunohistochemical markers. Patient follow up is required due to high recurrence rate.

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