

## Case Report

# Case report on a very rare tumour of the urinary bladder: paraganglioma

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## ABSTRACT

Urinary bladder tumours are usually of three types; urothelial carcinoma, squamous cell carcinoma and adenocarcinoma. Urothelial carcinoma is the most common type of bladder carcinoma and accounts for almost 90% of all bladder carcinoma. It is also known as transitional cell carcinoma. Squamous cell carcinoma accounts for 4% and adenocarcinomas 2% of all bladder carcinomas. Urinary bladder paragangliomas are a very rare variety of bladder tumours which account for less than 0.06% of all bladder tumours. They are clusters of neuroendocrine cells associated with sympathetic and parasympathetic nervous system. Paragangliomas are tumours arising from these chromaffin cells. They are most commonly seen in the head and neck and account for about 70% of all paragangliomas. The same type of tumour when found in the adrenal glands is referred to as a pheochromocytoma. Paraganglioma is usually seen in the age group of 30 and 50 years. The tumour is often slow growing and benign. These tumours may be functional or non-functional. Functional ones present with symptoms of catecholamine excess, which include hypertension, palpitations, diaphoresis, headache, and post-micturition syncope.

**Keywords:** Haematuria, Hypertensive crisis, Micturition, Paraganglioma, Urinary bladder

## INTRODUCTION

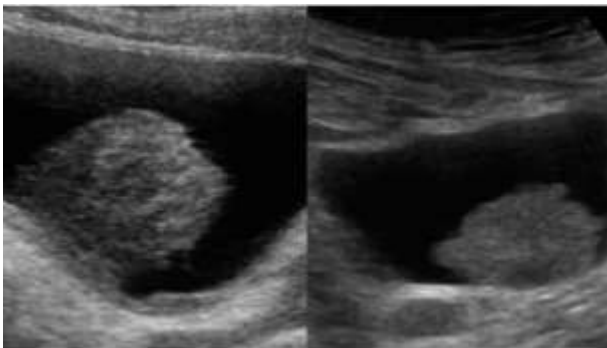
Urinary bladder tumours are usually of three types; urothelial carcinoma, squamous cell carcinoma and adenocarcinoma. Urothelial carcinoma, also known as transitional cell carcinoma is the most common type of bladder carcinoma and accounts for almost 90% of all bladder carcinoma. Squamous cell carcinoma accounts for 4% and adenocarcinomas 2% of all bladder carcinomas. Urinary bladder paragangliomas are a very rare variety of bladder tumours and account for less than 0.06% of all bladder tumours. A paraganglioma is a rare neuroendocrine neoplasm that may develop at various body sites like the head, neck, thorax and abdomen.

Seventy percent of the extra-adrenal paragangliomas arise from the head and neck region.<sup>1</sup> When the same type of tumour is found in the adrenal gland, they are referred to as pheochromocytoma. Paragangliomas of the urinary bladder originate from chromaffin tissue of the sympathetic nervous system associated with the urinary bladder. These tumours may be functional or non-functional. Zimmerman et al in 1953 reported the earliest case of paraganglioma of the urinary bladder.<sup>2</sup> Functional ones present with symptoms of catecholamine excess, which include hypertension, palpitations, diaphoresis, headache, and post-micturition syncope.<sup>3</sup> Paraganglioma is usually seen in individuals between the ages of 30 and 50. The tumour is often slow growing and benign. The

five year survival in the setting of metastatic disease is 90% to 100%. Conservative treatment is advocated to control hypertension and palpitation. Surgery to remove the tumour is usually the treatment of choice for a paraganglioma. It can result in severe or life-threatening complications if left untreated.

## CASE REPORT

A 38 years old female patient presented with the history of frequency, urgency, occasional headache, dull lower abdominal pain since 3 months and an episode of gross haematuria 7 days back. She was hypertensive, on calcium channel blocker and well controlled. There was no history of diabetes, smoking or any previous surgery. She was also a divorcee with no history of recent sexual contact. She was worked up with complete blood count, urine analysis with culture sensitivity (sterile urine), coagulation profile (normal), urine for pregnancy test (negative), liver function test, renal function test (normal), electrolytes, fasting and post-prandial blood sugar (normal) and blood pressure measurements (110/78 mmHg). She was subjected to ultrasonography of the whole abdomen which revealed a growth measuring approx. 20 mm x 18 mm at the base of the urinary bladder with good vascularity and normal upper tract (Figure 1). She was taken up for cystoscopy which confirmed the growth at the base of the bladder away from the ureteric orifices (normal efflux). No active bleeding was noted (Figure 2).



**Figure 1:** USG showed an approximately 20 mm x 18 mm sized adherent echogenic lesion at the base of the urinary bladder with good vascularity.



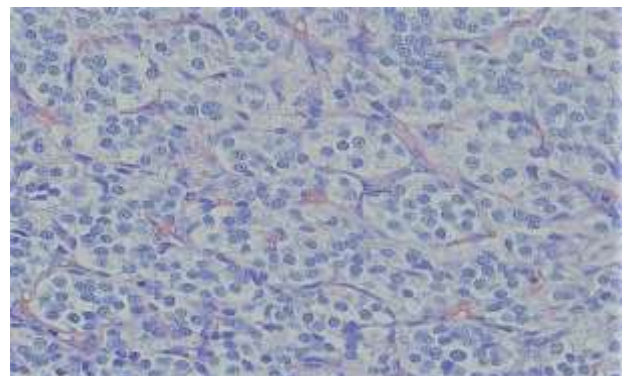
**Figure 2:** Cystoscopy revealed a sessile solitary growth at the base of the urinary bladder away from the ureteric orifices.

## Management

After pre-anesthetic check up and fitness for surgery, the patient was taken up for TURBT (transurethral resection of bladder tumor) under spinal anesthesia with bilateral obturator block.

Intraoperative findings suggested a very vascular and sessile tumor. The tumor was completely resected and the margins coagulated. Apart from the tissue from the growth, the deep tissue from the base of the growth was also sent separately for histopathology (protocol followed in our institution). After proper hemostasis, the patient was catheterized with tri-way large bore Foley catheter and bladder irrigation was done with normal saline. Blood pressure and other vitals were well maintained throughout the procedure. Operative time was approximately 45 minutes.

Patient had an uneventful postoperative period. Catheter was removed 24 hours after surgery and the patient was discharged, only with an oral antibiotic after 48 hours. On histopathology, tumour cells were round to oval with eosinophilic cytoplasm exhibiting pleomorphism and hyperchromasia (salt and pepper nuclei). Cells were arranged in occasional nests with trabecular pattern (Zellballen) with prominent vascular network. Occasional mitotic figures were present (Figure 3). Immunohistochemistry study was not done.



**Figure 3:** On histology, tumour cells were round to oval with eosinophilic cytoplasm exhibiting pleomorphism and hyperchromasia (salt and pepper nuclei). Cells were arranged in occasional nests with trabecular pattern (Zellballen) with prominent vascular network. Occasional mitotic figures were present.

The patient is being followed up every three months with ultrasonography of abdomen, urine analysis and blood pressure recordings. She is asymptomatic and compliant with the follow up schedule. We subjected her to cystoscopy 6 months after surgery which was absolutely normal. We plan to follow her up at 6 month intervals for the next 2 years and then once a year for 5 years.

## DISCUSSION

Paragangliomas of the urinary bladder are very rare tumours that originate from chromaffin tissue of the sympathetic nervous system associated with the urinary bladder. Seventy percent of the extra-adrenal paraganglioma arise from the head and neck region.<sup>1</sup> These tumours may be functional or non-functional. Zimmerman et al in 1953 reported the earliest case of paraganglioma of the urinary bladder.<sup>2</sup> Functional ones present with symptoms of catecholamine excess, which include hypertension, palpitations, diaphoresis, headache, and post-micturition syncope.<sup>3</sup>

Urinary bladder paragangliomas account for less than 0.06% of all tumours. The urinary bladder is the most common site for paragangliomas (79.2%) in the genitourinary tract, followed by the urethra (12.7%), pelvis (4.9%) and ureter (3.2%).<sup>2,4</sup>

They are three times more predominant in females and occur primarily between 30 and 50 years of age.<sup>5</sup> Eighty five percent of the extra-adrenal tumours are below the diaphragm along the sympathetic chain at the origin of inferior mesenteric artery or bifurcation of aorta.<sup>6</sup>

Urinary bladder paragangliomas may be found anywhere in the bladder as well-circumscribed nodules or nodular aggregates, red-pink to brown. Microscopically composed of nests (Zellballen) of round to oval chief cells (neuroectodermal in origin) that is surrounded by delicate vascular septae. The tumour cells contain abundant clear or granular eosinophilic cytoplasm. Nucleus may be uniform, round to ovoid or sometimes vesicular.<sup>1</sup>

Cytoplasm may be pleomorphic with occasional mitosis. Chief cells stain strongly for chromogranin, synaptophysin, neuron specific enolase, CD 56 and CD 57. Supporting network of spindle shaped stromal cells (sustentacular cells) test positive for S100 protein. Urothelial cancers are positive for cytokeratin.

Ultrasonography usually identifies the growth within the bladder with Doppler picking up increased vascularity. They present as heterogeneous hyper enhancing mass on CT with angiography depicting intense tumour blush with enlarged feeding arteries. MRI can also be done though it is only slightly better than CT to denote soft tissue extension.

Complete surgical resection is the treatment of choice with appropriate measures in the perioperative period to tackle hypertensive crisis. The reported options of surgical treatment for localized or advanced disease of

the urinary bladder include radical cystectomy, partial cystectomy, and transurethral resection.<sup>7</sup>

## CONCLUSION

In a high output centre like ours, it is very difficult to preoperatively diagnose paraganglioma. However in presence of features suggestive of episodic hypertension, palpitations, headache and post micturition syncope, paraganglioma should be considered as a possible differential diagnosis and should be evaluated to prevent catastrophic perioperative complications. This is rarest of the rare UB growth, considering the massive number of patients of bladder space occupying lesions encountered in our daily practising lives.

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