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# Paraganglioma of Urinary Bladder in a Pregnant Lady

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**Abstract** **Case Report**

Paraganglioma of the urinary bladder is extremely rare accounting for less than 0.05% of all bladder neoplasm. Zimmerman and coworkers noted the first instance of paraganglioma of the urinary bladder in 1952. Most patients are young. Age at detection can range from 10 to 70 years. Most common presentation of this tumour is painless haematuria, headache, palpitations and anxiety. Symptoms are paroxysmal and associated with micturation. Episodic hypertension is also encountered. In 2004 the WHO classified pheochromocytoma as endocrine tumours arising from chromaffin cells of adrenal medulla while extra-adrenal tumours are classified as paraganglioma. The distinction between pheochromocytoma and paraganglioma is important due to the risk of malignancy, associated neoplasm and implications on genetic testing. Paraganglioma mainly secrete norepinephrine and adrenal pheochromocytoma epinephrine.

**Keywords**: Paraganglioma, bladder, pregnant lady.



## INTRODUCTION

Neoplasms arising from chromaffin cells of the sympathetic nervous system are called pheochromocytomas. Approximately 70% of pheochromocytomas arise in the adrenal medulla. Extra-adrenal tumors identified as pheochromocytomas are referred to as extra-adrenal paragangliomas. These arise predominantly in the retroperitoneum, from the upper abdomen to the pelvic floor. The urinary bladder can also be a primary site of extra-adrenal paragangliomas.

9.8% of extra-adrenal paragangliomas arise from the urinary bladder, although paragangliomas account for only 0.06% of all tumors of the urinary bladder.

Tumors of chromaffin cells, derived from the embryonic neural crest, usually originate from the adrenal medulla and are designated as pheochromocytomas. However, 10% of these tumors occur at extra-adrenal sites and are known as paragangliomas[1]. Paraganglioma of the urinary bladder are extremely rare and are usually functional and symptomatic [2].

## CASE REPORT

A 29 years old pregnant lady of 28 weeks gestational age, presented with a single episode of painless hematuria 2 days back that resolved spontaneously. Her general physical examination was unremarkable. Complete urine examination showed plenty of red blood cells and few pus cells and urine culture was negative for any growth. Ultrasonography was done and shows a mass on the right lateral wall of the urinary bladder, measuring 1x1.5cm. Cystoscopy also shows a mass on the right lateral wall of the bladder. Patient also complains of excessive sweating,palpitations and her blood pressure was high upto 190/110mm Hg. Anti-hypertensives were given and blood pressure was controlled. Patient's serum and urinary catecholamines were elevated and diagnosis of Pheochromocytoma was made. Surgical excision was done and sent for histopathological examination. Grossly it was soft and grey yellowish in colour. Total specimen was submitted and examined under microscopy after the staining procedure. On microscopy section show nests (zellballen) of polygonal cells, with abundant granular cytoplasm and uniform round to ovoid nuclei, enclosed by trabeculae of fibrous tissue and elongated sustenticular cells. Based on the above findings it was diagnosed as Paraganglioma of the bladder.

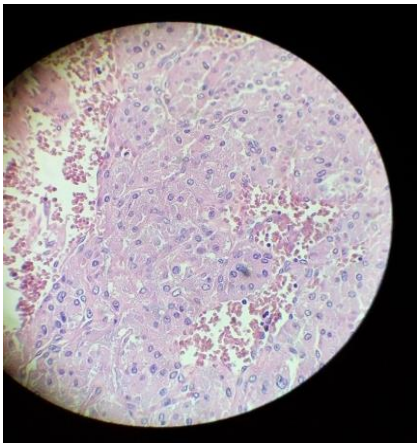

**Fig-1: Section showing zellballen pattern arrangement of tumour cells having ovoid nuclei separated by delicate fibrous septa [ H&E,X40]**

## DISCUSSION

Most of the pheochromocytoma tumours are benign, and are potentially curable. Approximately 10% of pheochromocytoma is malignant, while 25% of paraganglioma are malignant; especially those associated with succinate B dehydrogenase deficiency. Clinical, radiological and biochemical features cannot distinguish a benign from a malignant lesion. Malignant pheochromocytoma is clinically diagnosed by presence of distant metastases mainly to the liver, lymph nodes, lungs or bones. The malignant tumours are diagnosed when metastases are found at non-chromaffin sites.

Most of cases of pheochromocytoma are sporadic. However some are associated with hereditary or familial syndrome. At least 10 different genes have been implicated in pathogenesis of these tumours. Mutational analyses of ret-proto oncogene (RET), succinate dehydrogenase B (SHDB), succinate dehydrogenase D (SHDD) and VHL revealed mutation in 25% of sporadic cases suggesting that one-third of patients with pheochromocytoma have inherited the disease.

The patients with malignant pheochromocytoma should be offered testing for SDHB, VHL and SHDD mutations as about 40–50% of such patients will have a detectable mutation. Malignant and/or extra-adrenal pheochromocytoma, particularly in the abdomen, is strongly associated with SHDB mutation. It has been suggested that SDHB mutation analysis should always be performed in cases of familial chromaffin tumours.

The medical management is directed towards control of hypertension with α-blockage and β-blockage. The surgical treatment is debulking of the tumour mass. In some cases where surgical treatment is not possible, radiotherapy can be used. Malignant pheochromocytoma remains incurable. Palliative chemotherapy regimen commonly used is a combination of cyclophosphamide, vincristin and decarbazine—with improvement of symptoms and a remission rate of 50% observed in some of the patient [3,4]. This remission lasts 1–2 years.

Paraganglioma of the urinary bladder is very rare and account for 0.06% of all bladder tumors and 6% of extra-adrenal pheochromocytomas. However, in the genitourinary tract, the urinary bladder is the most common site (79.2%), followed by the urethra (12.7%), pelvis (4.9%), and ureter (3.2%).

These tumors originate from chromaffin tissue of the sympathetic nervous system associated with the urinary bladder wall and are most commonly situated at the dome or the trigone of the bladder and may be nonfunctional or functional. They remain usually benign, but 15–20% tumors may show malignant behavior. Bladder paraganglioma occur more frequently in women than in men, and clinical presentation occur mainly during the third decade of life[2].

In functional tumors, presenting symptoms are usually resulting from excessive catecholamine secretion. The patient typically suffers from hypertensive crises that may be accompanied by headache, palpitations, hot flushes, and sweating. About 17% of bladder paragangliomas are hormonally nonfunctional and can be asymptomatic [2]. Painless hematuria is a common presenting complaint in about 60% of reported cases, though it is nonspecific for paraganglioma and can be a presenting feature of any bladder tumor. In symptomatic patients, functional assessments of plasma and/or urine catecholamine levels are crucial in the initial workup and in the follow-up afterward. Plasma metanephrines are more sensitive and specific than urinary metanephrines for these lesions.

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