

## Pheochromocytoma of the Urinary Bladder – A Rare Case Report

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**Abstract:** Pheochromocytoma of the urinary bladder is a rare neoplasm of the chromaffin tissue of the sympathetic nervous system that occurs within the layers of the bladder wall. The diagnosis is based largely on the presence of clinical symptoms related to catecholamine hypersecretion, although the differential diagnosis of carcinoma of bladder must be excluded. A young male patient of 26 year age presented with haematuria, paroxysmal attacks of hypertension causing palpitation, dizziness, blurring of vision and headache while micturition for last 03 months. Clinical suspicion of pheochromocytoma was confirmed by 24 hour urinary catecholamine level, transabdominal USG and CT scan of abdomen. After having two weeks of preoperative preparation with phenoxybenzamine and propranolol, partial cystectomy was done. Peroperative fluctuation of BP was well managed by IV fluid overload, intravenous phentolamine, intravenous esmolol and intravenous ephedrine. Postoperative recovery was uneventful and BP regains to normal range from 1st postoperative day. Pheochromocytoma is a rare cause of hypertension. If the diagnosis of pheochromocytoma is overlooked, the consequences could be disastrous, even fatal; however, if a pheochromocytoma is identified, it is potentially curable, as being one of the cause of surgically correctable hypertension.

**Keywords:** Pheochromocytoma, catecholamine, hypertension, alpha blocker, urinary bladder

### I. Introduction

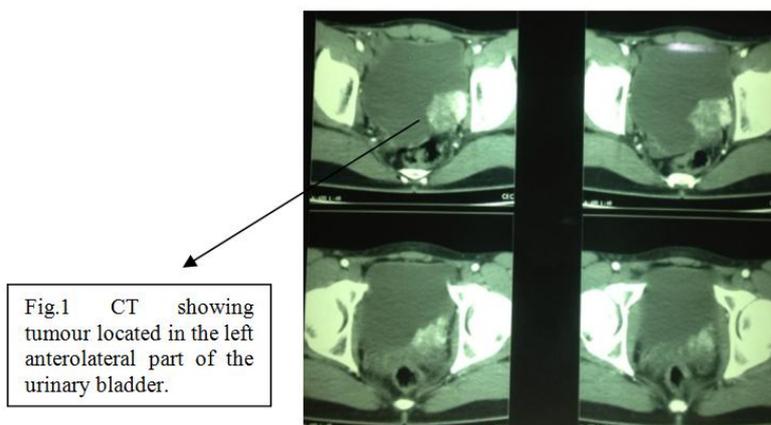
Pheochromocytoma is a catecholamine-producing tumor, which arises from chromaffin cells which are derived from the ectodermic neural system/neuroendocrine system. Most pheochromocytomas are situated within the adrenal medulla [1] and pheochromocytoma of the urinary bladder is rare [2]. The tumor can secrete epinephrine, norepinephrine, and dopamine, which are responsible for its major clinical manifestations: sustained or paroxysmal hypertension, headache, palpitations, and profuse sweating. Although the prevalence is low (about 0.1% of hypertensive population), pheochromocytoma is of great clinical significance because, if left untreated, it can result in devastating consequences. When pheochromocytoma is suspected clinically, the diagnosis is based on measurements of urinary or plasma catecholamines and metabolites, followed by radiological or scintigraphic studies for tumor localization. Surgical removal of the tumor is the mainstay treatment option. I herein present a 26-year-old male with hematuria and occasional episodes of hypertension.

### II. Case Report

A 26-year-old man presented with a history of hematuria, occasional paroxysmal attacks of hypertension causing palpitation, dizziness, blurring of vision and headache while micturition for last 03 months. The headaches and palpitations started approximately 2 minutes after urination, and continued for approximately 3 minutes. He was asymptomatic in between these episodes.

His medical, surgical, and family histories were not significant. On examination he was thin, with a pulse of 86 beats per minute and a blood pressure of 124/76 mmHg.

Ultrasonography showed a revealed a vascular, nearly homogenous mass in the left anterolateral wall of the urinary bladder wall which was confirmed on computed tomography (CT) scan (Fig. 1).



CT Scan showed markedly enhancing soft tissue lesion of size 49×30×47 mm (AP×TR×CC) in pelvis on left side with large extra vesical component and small intramural and intra vesical component and left lower uretric encasement and occlusion causing gross chronic hydronephrosis of left kidney with renal parenchymal thinning. Left kidney shows gross hydronephrosis and is enlarged in size with paper thin parenchyma(4-6mm) left ureter dilated in its course with abrupt narrowing and encasement at the level of left pelvic mass. This scan also showed that his adrenal glands were completely normal.

The results of routine laboratory examinations (full blood count, blood chemistry, coagulation studies, and urinalysis) were within normal limits. His plasma metanephrine was 0.20 nmol/L (normal range at our hospital laboratory, 0.00–0.49 nmol/L) and plasma normetanephrine 39.9. His chest radiograph and electrocardiogram were normal.

He was diagnosed with a urinary bladder pheochromocytoma and underwent partial cystectomy. During the surgery, his arterial blood pressure was raised. An extended partial cystectomy was performed because the tumor was involving the whole thickness of the bladder wall and the peritoneum covering the bladder wall at the site where the tumor was located.

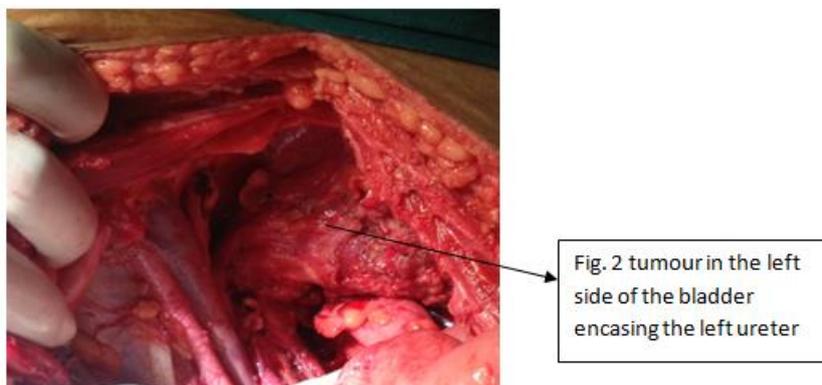




Fig. 3 Excised specimen of the tumour with distal end of left ureter

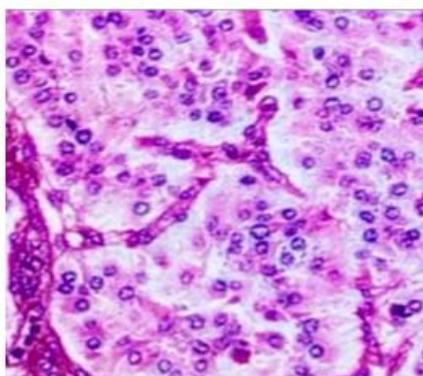


Fig. 4 Histopathological findings were suggestive of pheochromocytoma, the tumor cells were arranged in a nested pattern.

### III. Discussion And Literature Review

Pheochromocytoma of the urinary bladder was first reported in 1953 by Zimermana Zimmerman [3] and to date about 300 cases have been reported in the literature. Pheochromocytoma of the urinary bladder accounts for less than 1% of pheochromocytomas in humans and less than 0.06% of bladder tumors [4,5].

The symptoms of bladder pheochromocytoma, such as headaches, palpitations, dizziness, and sweating, are similar to those of adrenal pheochromocytoma. However, the symptoms are usually associated with micturition or defecation. Gross or microscopic hematuria was noted in 60% of patients with bladder pheochromocytoma. The hypertensive crises result from excessive catecholamine secretion, which usually accompanies voiding [6,7]. Other symptoms such as dysuria or suprapubic pain are rare. It has been reported that 17% of bladder pheochromocytomas are hormonally nonfunctional and asymptomatic [8,9].

Our patient presented with paroxysmal hypertension and symptoms related to excessive catecholamine secretion only when he got up to urinate at midnight. This might be explained by the following: (i) the low probability that this was a functional tumor or the small size of the tumor; (ii) his sympathetic nervous system being more excitable when he woke up at midnight; or (iii) the peritoneum being closely attached to the bladder tumor, so that the tumor was easily stimulated by stretching when he got up to urinate at midnight.

The diagnosis of pheochromocytoma is generally established by measurement of catecholamines in plasma, and catecholamine metabolites (metanephrine and normetanephrine) in a 24-hour urine collection.

On ultrasonography, pheochromocytomas appear as solid masses or contain foci of hemorrhage and necrosis. CT can detect larger bladder tumors, but its sensitivity is just 82%. Magnetic resonance imaging (MRI) is superior to CT in locating tumors and differentiating them from surrounding structures. In 2010, Wang reported the use of MRI in the diagnosis of a bladder paraganglioma [10]. He found that homogeneous T1 hyperintensity was a diagnostic characteristic of bladder paraganglioma on MR imaging, and that necrosis, oval shape, and lower apparent diffusion coefficient (ADC) values provided further support for the diagnosis of bladder paraganglioma.

Surgical removal of the tumor was the most effective treatment for bladder pheochromocytoma. In the report of Das et al. with 100 cases of bladder pheochromocytoma, partial cystectomy accounted for 84% and total cystectomy 7% of the treatments performed [11]. Transurethral resection was performed in about 7% of cases as an alternative for small and well-defined lesions. It was unclear if there was any advantage for total cystectomy over partial cystectomy in terms of disease control. As the majority of tumors (94%) involve the muscularis propria of the bladder wall, it has been proposed that transurethral resection is insufficient and that partial cystectomy remains the first-choice treatment for bladder pheochromocytomas [12-14]. We believe that

extended partial cystectomy is sufficient to control the disease in patients with lesions that involve the whole thickness of the bladder wall, and that total cystectomy, which impairs patient quality of life is not necessary.

#### **IV. Conclusion**

A case of bladder pheochromocytoma with symptoms related to increased catecholamine release after urination was successfully treated with extended partial cystectomy. When pheochromocytoma is suspected clinically, the diagnosis is based on measurements of urinary or plasma catecholamines and metabolites, followed by radiological or scintigraphic studies for tumor localization. Surgical removal of the tumor is the mainstay treatment option.

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