CASE REPORT

Extra adrenal pheochromocytoma of urinary bladder

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Abstract

Pheochromocytoma of urinary bladder is rare disease causing a hypertension, palpitation, throbbing headache & hemorrhage induced by voiding. This case is reported because of rarity of this disease in urinary bladder.

Keywords Pheochromocytoma of urinary bladder · Bladder Tumors · Extra adrenal Pheochromocytoma.

Introduction

Pheochromocytoma is an uncommon entity, which requires effective means of detection, localization and management. It derives from neural crest and accounts for less than 1 % of hypertension. It also occurs spontaneously in familial disease known as multiple endocrinal neoplasia type 2, Neurofibromatosis and Von Hippel-Lindau disease, which have an autosomal dominant trail. In up to 5% of patients Pheochromocytoma occurs as part of this pluriglandar syndrome including medullary carcinoma of the thyroid, hyperparathyroidism (adenoma or hyperplasia) and oral mucosal neuromas with neuroectodermal dysplasia. Although pheochromocytomas are generally benign, about 10% of these tumors are malignant [1]

Case report

A 58-year-old female patient presented herself with giddiness during urination, syncope off and on during initiation of urination, dysuria, and haematuria with passage of clots in urine off and on and weakness since 5 years. She was under regular treatment of a physician since 5 years and was receiving symptomatic treatment for the above problems. Her blood pressure was 170/100 mm of Hg and pulse rate was 114/minute and ECG report was within normal limits.

Initially she was markedly anemic with 4 gm% Hb that was raised to 11 gm% after 6–7 units of blood transfusion. Blood Urea, Serum Creatinine and Blood Sugar were normal in the patient. Urine examination revealed significant pus cells and red blood cells [RBCs]. Exfoliative cytology of urine was negative for transitional cell carcinoma.

Ultrasonography (USG) of patient showed $3.4 \times 3.3 \times 2.7$ cm size bladder mass arising from anterior wall and right lateral wall of bladder (Fig. 1). Kidney, ureter and

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Fig. 1 USG-Bladder showing solid tumor arising from anterior and right lateral wall of urinary bladder.

bladder were normal on USG. There was no significant enlargement of pelvic lymph nodes.

On cystoscopy there was solid oval shape tumor arising from anterior wall and roof of the bladder. Mucosa over tumor has shown old signs of hemorrhage. Biopsy was taken for histopathological examination of tumor. Histopathological report was confirmative of extra adrenal Pheochromocytoma or benign extra adrenal ganglioneuroma of urinary bladder.

After confirmation of extra-adrenal Pheochromocytoma of urinary bladder the antihypertensive drugs of patient was changed with B-Blocker Atenolol, alpha-blocker Doxazocin and Nifedine. After adequate improvement of general condition and stabilization of blood pressure the patient was taken for surgery after 15 days (Fig. 2).

Partial cystectomy was done through lower mid line extra peritoneal incision & Pheochromocytoma removed. Pelvic lymph nodes [Internal iliac, external iliac and common iliac] were examined to rule out secondaries in lymph nodes. Biopsies of pelvic lymph nodes were taken for HPE on both sides. Biopsy was negative on histopathological examination. Post cystectomy specimen of bladder was again confirmative of extra adrenal Pheochromocytoma with minimal invasion in bladder muscle in a few places (Fig. 3).

On the 10th day blood pressure came down to normal and the patient was discharged without antihypertensive drug on the 15th post-operative day.

Discussion and review of literature

Pheochromocytoma is a rare disease and often grows in the adrenal gland or Para-aortic ganglion. The most common presentation is hypertension, which may be sustained or paroxysmal. Other symptoms include severe headache, excessive sweating, palpitation, tachycardia anxiety, constipation, and an orthostatic hypotension. When Pheochro-

Fig. 2 Photomicrography showing nests of polyhedral and spindle shaped chromaffin cells with finely granular eosinophilic cytoplasm

Fig. 3 Partial cystectomy specimen showing pheochromocytoma

mocytoma is located in the urinary bladder, the symptom may occur after micturation. However some patients are asymptomatic. The incidence of Bladder Pheochromocytoma is less than 0.06% of all bladder tumors and less than one percent of all Pheochromocytoma [2, 3]. They occur in patients at any age, but the age of peak incidence is between the third and fifth decade [4]. They arise from paraganglionic cells within bladder wall, usually in the region of trigone [5]. Extradrenal Pheochromocytoma may arise in any position of the paraganglionic system and accounts for less than 10% of all Pheochromocytoma. Extra adrenal Pheochromocytoma probably represents in, at least 15% of adults and 30% of childhood. Bladder Pheochromocytoma is rare but give a clinical syndrome of paroxysmal hypertension after relieving an over distended bladder and headache: Fifty percent of patients may have painful haematuria. Cystoscopy examination may reveal a normal bladder because the lesions can be small and present in the muscular layer even if they appear as a sub mucosal

nodule covered by intact urothelium. Histologically nets of polyhedral cells with eosinophilic cytoplasm compose the tumor. Histological features of benign Pheochromocytoma cannot be distinguished from its malignant counterpart [5]. There is no age and sex predilection for bladder Pheochromocytoma but peak incidence of this tumor is seen in the second to fourth decades of life [4].

Several case reports of bladder Pheochromocytoma were reviewed, and most of them had symptoms of hypertension, and a drop in blood pressure occurred during the operation. In those patients, the bladder Pheochromocytoma was larger than 2 cm in diameter and all of them received a partial cystectomy [6–9]. Other reports have demonstrated that metastasis is possible even after surgery. Two case reports revealed that patients treated with transurethral resection of the bladder tumour did not experience a hypertensive crisis during the operation, but a drop in the systolic pressure to 50 mm Hg was noted in one case during the operation [10, 11].

Some reports have suggested that bladder pheochromocytomas may be malignant, and patients should be aggressively treated and should receive long-term follow-up after initial surgery. In this patient's pathology reported a well-circumferential tumor without muscle invasion, however the patient still requires regular follow-up. About 10% of bladder pheochromocytomas are malignant and have the capacity to metastasize to the regional lymph nodes or distant sites [14]. Malignancy is determined more by the clinical behavior than histological features of the tumor. Partial cystectomy is the treatment of choice in bladder tumor but TUR-BT [13, 15] is also recommended in some study without any complication. CT scan should evaluate regional lymph nodes preoperatively, operatively by gross inspection and biopsy of suspicious lymph nodes. Lymph node involvement is marked by hypo vascular-lesion on arteriography. Pelvic lymphodectomy is indicated in case of lymph node involvement by tumor.

CT is currently the initial imaging procedure of choice but Ultrasonography is a good option in bladder Pheochromocytoma when facility of CT scan and ¹³¹MIBG scan is not available. CT scan is not useful for small or extra-adrenal tumours. T2-weighted or gadolinium-enhanced magnetic resonance imaging (MRI) would be more helpful because the signal intensity of the Pheochromocytoma would be higher than the signal intensity of an adrenal adenoma. An iodine-131-metaiodobenzylguanidine (MIBG) scan has high sensitivity for pheochromocytomas, especially small lesions, extra-adrenal lesions, and metastatic lesions of malignant pheochromocytomas [14]. One study reported that CT has high sensitivity for pheochromocytomas in the adrenal gland but lower sensitivity for detecting extra-adrenal pheochromocytomas. The MIBG scan has high sensitivity (77–90%) and high specificity (88–99%) for localizing Pheochromocytomas, and it can provide information complementary to CT and

MRI [12]. One study reported that the tests of choice to diagnose pheochromocytomas are urinary nor metanephrine and platelet nor epinephrine estimation. In combination with an MIBG scan, the diagnostic sensitivity can be improved. When a Pheochromocytoma is suspected and catecholamine measurements are within a normal range, an MIBG scan should be done [13]. So, an MIBG scan may be necessary during follow-up if the tumor recurs or there is any residual tumour.

Hence preoperative pharmacological preparation by administering Alpha and Beta receptors blocking agents, attentive intraoperative monitoring and aggressive surgical therapy has an important role in achieving the safest and most successful outcome.

Life-long follow up is required because of late endocrinal manifestations and metastasis in this tumor. Extra adrenal Pheochromocytomas are more likely to recur and metastasize than their adrenal counterparts making lifetime follow up with annual determination of catecholamine production essential. This case is reported because of rarity of this disease in urinary bladder [15].

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