Introduction:

Urinary bladder Pheochromocytomas (bladder paraganglioma) are exceedingly rare tumors accounting for less than 1% of extra adrenal pheochromocytomas and less than 0.05% of all bladder tumours. It is a catecholamine-secreting tumour of chromaffin cells and can arise anywhere in the genitourinary tract with the urinary bladder is the most common site (1).

Although it could be totally asymptomatic, patients often present with headache, hypertension, palpitation, sweating, fainting or blurring of vision immediately after voiding(1,2). Painless hematuria also is common(2).

Most bladder Pheochromocytomas are benign. Only 10% of Pheochromocytomas are malignant and diagnosed according to the clinical behaviour ; the presence of local recurrence or metastatic disease.

Treatment is usually by open partial cystectomy. TURT is possible but has a high risk of hypertensive crisis due to catecholamine over secretion during resection.

In this case report we will discuss a case of pheochromocytoma presented with obstructive urinary symptoms and treated successfully by TURT.

Case report.

A 62 year old male smoker not known to have any medical illness was referred to our urology clinic at prince Hamza hospital (PHH) complaining of obstructive urinary symptoms mainly poor stream, hesitancy and straining. Physical exam was

unremarkable, PSA total was 2.5 mg\dl, his urea and creatinine were normal. Urinary tract ultrasound was done and showed a large bladder mass originating from the anterior wall of the urinary bladder measuring 4.3 *3.5 cm with post void residual of

120cc. Uroflowmetrey Q max was 9 ml/sec. CT scan and MRI showed multilobulated mass originating from anterior bladder wall(figure 1).

During cystoscopy, a large bladder mass was seen originating from the anterior wall of the bladder protruding toward the bladder neck which explains the patient's symptoms. Pre operatively and during the diagnostic cystoscopy his blood pressure was within normal values around 120\80. Decision was made to go for transurethral resection of tumor (TURT). Early during the resection his blood pressure started to rise up to 220/120 so the procedure was held. Post operatively his blood pressure was observed for 24 hours and was normal. Nephrology consult was requested regarding the rise in blood pressure. He had no abnormal readings during post operative period. On discharge, patient was asked to monitor his blood pressure regularly. No High readings were recorded.

Histopathology report mistakenly showed transitional cell carcinoma of the bladder. Second stage TURT was planned 4 weeks later after proper cardiology consultation. During the second TURT hypertensive crisis happened again shortly after starting resection. The procedure was held.

Histopathology this time revealed pheochromocytoma (paraganglioma). Biochemical workup was done which showed increased level of urinary metanephrine .TURT was planned for the third time. In order to complete the resection, preoperative preparation

with alpha and beta antagonists 2 weeks before TURT were prescribed (doxazocin 4 mg once daily and bisoprolol 2.5 mg once daily). On the 3rd session of TURT, complete resection was done as shown in (figure 2) without any rise in his BP intraoperatively.

In the Postoperative period the patient was doing well, all of his symptoms improved dramatically, and has no rise in his blood pressure.
Urinary and plasma metanephrine level were done 2 weeks ,6 months , and 1 year after complete resection and were normal. Follow up biphasic CT scan at 9 months showed completely normal bladder with no recurrence (figure 3). Cystoscopy 1 year after resection showed normal bladder walls with no recurrence .

Discussion and review of literature:

Correct preoperative identification of bladder Pheochromocytomas is important. Unsuspected bladder Pheochromocytomas may result in intraoperative hypertensive crises and greatly increase the perioperative mortality forcing the surgeon to terminate cystoscopic tumor resection(3). preoperative stabilization of hypertension strategy is necessary as in other pheochromocytomas with α-blockade using phenoxybenzamine. β-Blockade may be added to counteract the rebound tachycardia. Certain patients may require calcium channel blockers such as nifedipine to maintain adequate control (1).

However, due to its rarity compared to urothelial carcinoma. Urologists usually do not put it in differential diagnoses when dealing with bladder mass. Ultrasonography (USG), Computed tomography (CT) magnetic resonance imaging MRI and meta-iodobenzylguanidine (MIBG) scintigraphy are imaging modalities used for the diagnosis

of urinary bladder Pheochromocytomas. while urothelial carcinoma is a hypovascular lesion, bladder Pheochromocytomas should always be considered when a hypervascular lesions seen in the bladder by enhanced CT scan (4).

Sudden increase of catecholamine release during micturition is responsible for characteristic symptoms of sharp headache, hypertension, palpitation, sweating, fainting or blurring of vision immediately after voiding (1). About one forth of urinary bladder Pheochromocytomas are nonfunctional. Painless hematuria has been seen in 50-60% (3). Our patient has completely different presentation of obstructive urinary symptoms without hematuria or adreno-sympathetic symptoms during micturation.

Patients cure best achieved by surgery, the most common surgical procedure performed for bladder Pheochromocytomas is Partial cystectomy. Laparoscopic excision also reported for many cases with bladder Pheochromocytomas. Radical cystectomy with pelvic lymph nodal dissection is the procedure of choice for malignant disease(1).About 20% of published cases that where localized or locally advanced treated by TURT.

Pathologist may misdiagnose bladder Pheochromocytomas as urothelial cancer, The major histologic features that led to misdiagnosis included a diffuse growth pattern, focal clear cells, necrosis, and frequent involvement of the muscularis propria, with significant cautery artifact compounding the diagnostic problems, some pathologists fail to include Pheochromocytomas in their differential diagnosis when evaluating a bladder tumor (5).

Since bladder pheochromocytomas may be malignant, patients should receive long-term follow up after initial surgery. Life-long follow up with annual determination of catecholamine production is required because of late endocrinal manifestations and metastasis in this tumor(5).

Conclusion:

Single or multiple stages TURT is feasible option for treatment of bladder pheochromocytoma. Sudden rise of blood pressure during TURT for bladder mass should raise the suspicion of bladder pheochromocytoma. Preoperative alpha and B blocker 2 weeks before the surgery is mandatory to prevent intraoperative hypertensive crisis during resection. Long term follow up after complete resection is advised.

Keywords:
Bladder pheochromocytoma; Paraganglioma; Extra-adrenal

References:

1. Lacefield, E., Raheem, O. A., Cox, R., Davis, R., & Kamel, M. H. (2014). Composite Paraganglioma and Neuroblastoma of the Urinary Bladder: A Rare Histopathological Entity. Current Urology, 8(4), 208–211. doi:10.1159/000365718

2. Vyas S, Kalra N, Singh SK, Agarwal MM, Mandal AK, Khandelwal N. Pheochromocytoma of urinary bladder. *Indian J Nephrol*. 2011;21(3):198–200. doi:

10.4103/0971-4065.78072

3. Li S, Lui S, Li F, Yue Q, Huang X, Gong Q. Unsuspected paraganglioma of the urinary bladder with intraoperative hypertensive crises: A case report. *Exp Ther Med*. 2013;6(4): 1067–1069. doi:10.3892/etm.2013.1242

4. Zhou M, Epstein JI, Young RH. Paraganglioma of the urinary bladder: a lesion that may be misdiagnosed as urothelial carcinoma in transurethral resection specimens. Am J Surg Pathol. 2004 Jan;28(1):94-100

5. Shikha Goyal, Tarun Puri, Gowthaman Gunabushanam, Anup Kumar Das, Pramod Kumar Julka. Skeletal metastases from recurrent paraganglioma of the urinary bladder.Indian J Urol 2005;21:122-24.