Case Report

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Non Functioning Paraganglioma of the Urinary Bladder Presenting as Micturition Syncope

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Abstract

Syncope is a clinical presentation which is a transient loss of consciousness for a short period of time. Micturition (or post-micturition) syncope is fainting during or immediately after micturition due to a severe drop in blood pressure. Micturition syncope is most common in elderly men and usually occurs at night after a deep sleep.

Paraganglioma of the urinary bladder are tumors of chromaffin tissue originating from the sympathetic innervations of the urinary bladder wall and are extremely rare. Paraganglioma of the urinary bladder is one of the causes of micturition syncope in early postmenopausal women who usually present with severe dysuria, hypertension and post-micturition syncope with elevated Plasma and Urinary metanephrine levels.

In our case it is a silent paraganglioma of the urinary bladder that presented only with severe dysuria, hypertension and post-micturition syncope with normal Plasma and urinary catecholamine levels.

In this report we analyse the clinical presentation, investigations and management of bladder pheochromocytoma.

Keywords: Micturition syncope; Bladder pheochromocytoma.

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Introduction

The micturition syncope occurs immediately after or during urination and was first reported by Rugg-Gunn in 1946. The main reason for this syndrome is the vasodilatation and decreased vascular resistance due to vaso-vagal reflex related to urination.

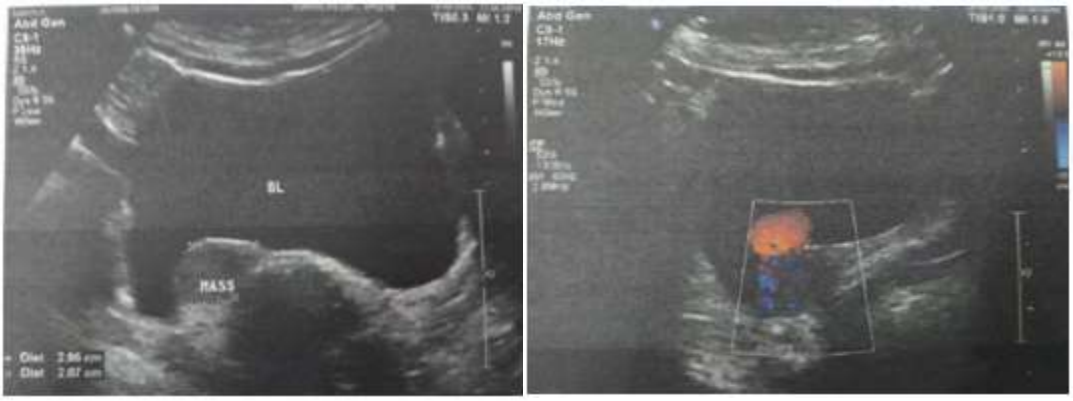
Paraganglioma is an extra adrenal site of pheochromocytoma. It is reported to account for 10% of pheochromocytomas, and 10% of cases occur in the bladder. Vesical Paraganglioma is a very rare disease and accounts for 0.06% of all bladder tumors.[1,2]

Paraganglioma of the urinary bladder are extremely rare and are usually functional and symptomatic. In our case it is a silent paraganglioma of the urinary bladder that presented only with severe dysuria, hypertension and post-micturition syncope with normal Plasma and urinary catecholamine levels.

Case Report

A 48-year old post menopausal woman presented with severe dysuria and recurrent post-micturition syncope for 2 months. Physical examination was unremarkable. Urine Analyzis showed plenty of red blood cells and few pus cells hence Urine culture was done which did not grow any microorganisms.

Ultrasound KUB demonstrated a homogenous mass in the Right lateral wall of the bladder, measuring 2.5 × 2.0 cms (Figs. 1,2).



Figs. 1,2: USG KUB.

MRI Pelvis was done which showed a 3 × 2 cms growth in the right lateral wall of the urinary bladder (Fig. 3).

She was on alpha and beta blockade for 14 days on an outpatient basis for hypertensive crisis. In view of post-micturition syncope, urinary catecholamine levels were estimated and were found to be within normal limits. Urine cytology was negative.

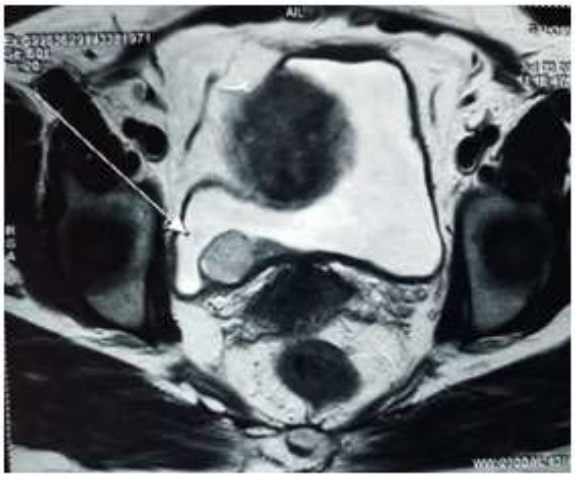


Fig. 3: MRI Pelvis.

Cystoscopy revealed a smooth, well-vascularized mass on the Right lateral wall of the bladder, close to the ureteric orifice. In view of the tumour located near to the right ureteric orifice patient underwent Partial Cystectomy with complete excision of the vesical mass (negative frozen section) and Right Ureteric re-implantation in view of its proximity to the Right ureteric orifice. During handling of the bladder mass intraoperatively, the patient became severely hypertensive. Her blood pressure raised up to 280/180 mm Hg and pulse rate dropped to 38/ min. Intraoperatively the Hypertensive crisis was managed with intravenous beta blockers and atropine. Postoperative recovery was uneventful and she was weaned off the antihypertensive medications.

Histopathology showed bladder wall lined by attenuated transitional epithelium overlying a well circumscribed, encapsulated neoplasm composed of cells arranged in organoid nests and alveolar pattern. The cells were round to oval with vesicular nuclei, mild pleomorphism, fine chromatin and inconspicuous nucleoli (Fig. 4). Many of the cells showed fine granular eosinophilic to clear cytoplasm (Fig. 5). The thin stroma in between the cells showed thick walled capillaries and mild inflammation. HPE report was consistent with bladder pheochromoctoma with deeper tissues showing no evidence of tumor.

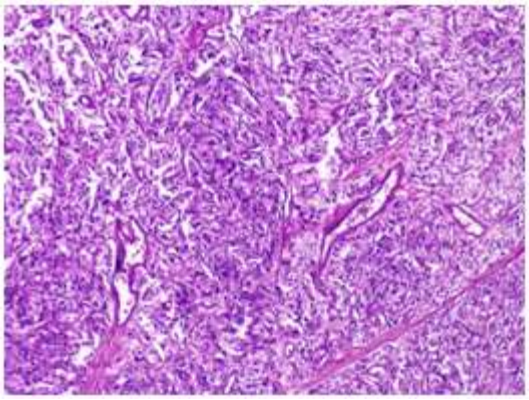


Fig. 4: Bladder wall with organoid nests of tumor cells (H&E 40x).

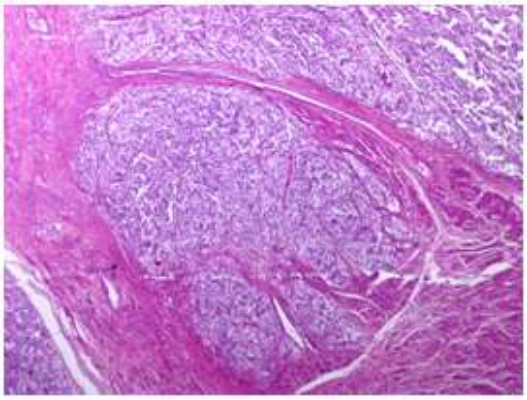


Fig. 5: Tumor cells with abundant granular cytoplasm, mild pleomorphism and fine chromatin (H&E 40x).

Immunohistochemistry Report

- The neoplastic cells expressed strong cytoplasmic staining for Synaptophysin and NSE.
- Most of the cells were S100 positive.

Patient is on regular follow-up with physical examination, plasma and urinary metanephrine levels, Ultrasonography and Cystoscopy. For the past 12 months, on follow-up, there is no tumor recurrence and no antihypertensive medications.

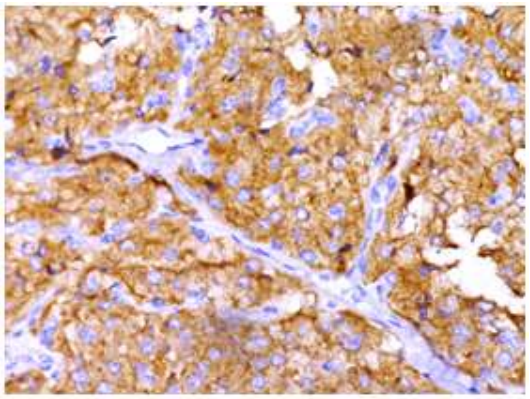


Fig. 6: Neoplastic cells expressing strong cytoplasmic staining with synaptophysin (IHC: 40x).

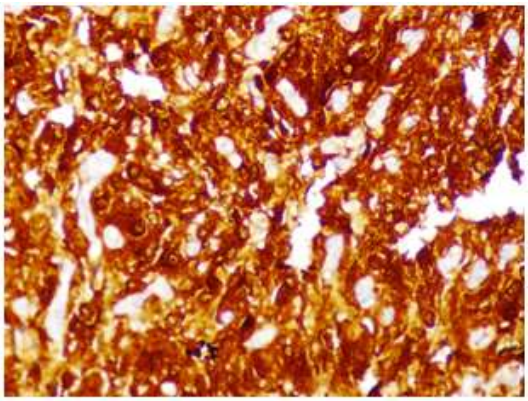


Fig. 7: Neoplastic cells expressing strong cytoplasmic staining with NSE (IHC: 40x).

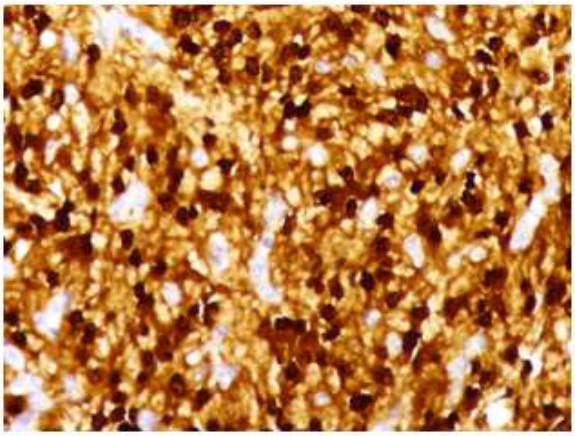


Fig. 8: Neoplastic cells expressing strong cytoplasmic staining with S100 (IHC: 40x).

Discussion

Vesical Paraganglioma are tumors of chromaffin tissue originating from the sympathetic innervations of the urinary bladder wall which are extremely rare and are most commonly situated at the trigone of the bladder-may be nonfunctional or functional. Vesical Paraganglioma is usually a benign tumor, but 5–10% of Vesical Paraganglioma tumors may have malignant changes. 10% of Vesical Paragangliomas are nonfunctional tumour and can be asymptomatic.

Usually patients will present with hypertensive crisis, headache, micturition syncope and palpitation. Micturition syncope are aggravated by digital rectal examination, defecation, ejaculation and urethral instrumentation. Vesical Paraganglioma is one of the causes of post micturitional syncope in post menopause patients. Elevated Urinary and Plasma metanephrines are more sensitive and specific for these lesions. 24-hours urinary and serum concentrations of vanillylmandelic acid (VMA), epinephrine, nor-epinephrine, and metanephrine are functional assessments of plasma and urine catecholamine levels in the initial workup and after surgical excision for follow-up. Serum metanephrines are more sensitive and specific than urinary metanephrines for these lesions.

The Vesical Paraganglioma is usually sub mucosal or intramural with intact vesical epithelium, the most common location being trigone. Most commonly seen in 4th decade with female to male ratio of 3:1. Paraganglioma in the bladder can be benign or malignant-histopathology showing atypical nuclei, increased mitotic activity, increased pleomorphism, vascular invasion, and infiltrating growth pattern.

Micturition syncope is a situational type of neurally mediated syncope syndrome.[3] Emptying of a full bladder stimulates the mechanoreceptors in the bladder wall. The afferent stimulus passes to brainstem through the vagus nerve, parasympathetic activity is triggered and bradycardia occurs. The inhibition of sympathetic activity results in arterial dilation and hypotension. Voiding in full bladder stimulates the mechanoreceptors in the bladder wall. The afferent stimulus passes to brainstem through the vagus nerve, parasympathetic activity is triggered and bradycardia develops. The inhibition of sympathetic activity results in arterial dilatation and hypotension.[4,5]

UItrasonography, CECT Urogram, Magnetic Resonance image of the pelvis are useful noninvasive imaging modalities. For the detection and spread of the Paraganglioma of the bladder tumor Metaiodinebenzylguinidine (MIBG) scan has shown to have a very high sensitivity and specificity.

Partial cystectomy or wide local excision of the bladder tumour is the better option over Trans-urethral resection of bladder tumor as majority of these tumors extend in the deep layers of the detrussor muscle.[2,3,5] Chance of recurrence is high following deep resection. Following complete surgical excision, frozen biopsy will show negative margin

Conclusion

Paraganglioma of the urinary bladder is a rare tumor that may be misdiagnosed as urothelial cancer, but a high index of suspicion, and search for the characteristic histological features and supportive immunohistochemical studies should lead to a correct diagnosis.

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