Case Report

Unraveling the Mystery: Paraganglioma in the Urinary Bladder. A Case Report

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Abstract

The bladder paraganglioma, which makes up 0.06% of all bladder tumors, is a highly uncommon condition. A 30-year-old female patient who was diagnosed with bladder paragangliomas at our hospital. The study covers the patient's journey from the first clinical presentation to the follow-up after surgery, providing insight into this uncommon illness. Urinary bladder tumour shown on CT KUB. The case was established by a transurethral resection biopsy. In order to completely remove the tumour, the patient had a radical cystectomy along with a hysterectomy and a bilateral salpingo-oophorectomy. A tumour mostly seen in the detrusor muscle that is organised in zellbellen pattern and is bordered by pushing fibrovascular stroma. Upon CT follow-up at three and six months, the results were generally unremarkable in terms of margins, metastasis, and local recurrence. Urinary tract hyperplasia is the most typical sign in our instance of bladder paraganglioma, a very uncommon tumour. A tumour called a polypoid in the bladder's posterior wall suggests cancer. More investigation into this uncommon ailment is necessary, as surgical extraction is still the only treatment available to these people.

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Keywords | Bladder tumor; paragangliomas.

Introduction

Only 0.5% of bladder tumours and fewer than 1% of all neoplasms that secrete catecholamines are bladder paragangliomas. Often affecting young women in their second or fourth decade of life, they are believed to develop from chromaffin cells that are embryonic remnants within the bladder wall. The adrenal gland's pheochromocytoma has an additional location known as a paraganglioma. 10% of cases of pheochromocytomas are said to be caused by bladder-specific paragangliomas, which account for 18% of all pheochromocytomas.[1,2] Paroxysmal hypertension, palpitations, headaches, hematuria, and hematuria with distinctive micturition episodes—wherein paroxysmal hypertension, palpitations, and micturition syncope[3] are caused by catecholamine secretion—are the most common symptoms of bladder paraganglioma. Ten percent of these tumours are able to penetrate and are consequently categorised as malignant, even though the majority are benign and do not exhibit the mitoses and cellular dissociation commonly observed in malignant tumours. Nevertheless, malignant paraganglioma is only detectable when the main tumour grows in organs including the liver, spleen, lungs, brain, bones, and lymph nodes that do not have any remnant ganglia, or when the tumour infiltrates the non-nervous system locally.[4] In this instance, the patient herself gave her written, informed consent.

Case Report

A referred patient, 30-year-old female housewife who had a BMI of 26.37 - 157 cm / 65 kg and complained

Production and Hosting by KEMU
https://doi.org/10.21649/akemu.v30i1.5422
2079-7192/

of dizziness, hypertension, palpitation, intermittent hematuria with clots for two weeks along with burning micturition, intermittency, and weight loss was seen in the urology outpatient department. There was no other noteworthy medical history, such as a fever, dysuria, or abdominal pain. Previous surgical and medical histories were unimportant. There was no notable family history of the condition. Urine D/R test (urinalysis) showed turbid appearance ((Reference range) Rr: Clear)), color of urine as dark yellow (Rr:0-2RBC/HPF) 25-30 white blood cells (Rr:0-5WBC/HPF), occasional epithelial cells (Rr:Nil/ HPF), 3+ leukocyte esterase enzyme (Rr: Negative), With the exception of minor hematuria (9–12 RBC/high power field), laboratory results including routine microscopy and microbiological investigation of urine were mostly normal. Except for somewhat low hemoglobin (9.0 g), the other hematological and biochemical tests were within normal range. An uneventful physical examination revealed stable vital signs. Ultrasound of kidney and bladder showed mass like thickening of urinary bladder at right VUJ causing right sided mild hydronephrosis. Radiological Computed tomography (CT) Kidney Ureter bladder showed mass at right lateral wall of urinary bladder involving adnexal region and right distal ureter (separate from uterus). After three months her MRI pelvis was done that showed lobulated enhancing mass lesion at the right hemi pelvis predominantly along the right lateral wall and base of urinary bladder and right anxeal regionl. Her TURBT was done, and biopsy proved the case of paraganglinoma of urinary bladder wall lined by benign urothelial lining. Patient underwent radical cystectomy with hysterectomy and bilateral salpingo-oophorectomy for the complete resection of tumor. Polypoid Tumor in posterior wall of urinary bladder was seen as gross. Postoperatively, the tumor was diagnosed as paraganglioma from microscopic histopathological results that showed urinary bladder wall lined by benign urothelial lining. The underlying bladder wall revealed a neoplasm arising in the detrusor muscle predominantly arranged in nests/ clusters (Zellbellen pattern) separated by fibrovascular stroma with pushing borders (paraganglioma of bladder) in figure 1. The tumor was composed of neoplastic cells with amphophillic cytoplasm, round nuclei with stippled salt and pepper chromatin and some mitoses. There were no areas of marked nuclear pleomorphism or necrosis. The tumor was limited to detrusor muscle and was not infiltrating the underlying fat. Tumor was limited to the muscularis propria. Benign urothelial lining epithilium was seen. Both right and left ureters, uretheral resection margin, six reactive lymph nodes, cervix, endometrium (proliferative pattern), myometrium, right and left ovaries, right and left fallopian tubes were unremarkable and free of tumor. Immunohistochemistry results showed Chromogranin as diffusely positive in tumor cells, S100 positive in sustentacular cells surrounding the tumor nests (figure 1), Marker of Proliferation Ki-67: 15-20%, synaptophysin negative in tumor cells, transcription factor GATA3 negative in tumor cells, cytokeratin tumor marker CKAE1/AE3 negative in tumor cells, cytokeratin CK-7 negative in tumor cells. With reference to WHO Classification of Tumors of the Urinary System and Male Genital Organs (2014) tumor was pT2, that was limited to the detrusor muscle (pT2) behave in a benign fashion, so no radio or chemotherapy was required. After procedure patient was under local surveillance. On follow up after three- and six-months CT was done to check for margins, metastasis and local recurrence, which showed essentially unremarkable results. At the moment, the patient is free of complaints and recurrences.

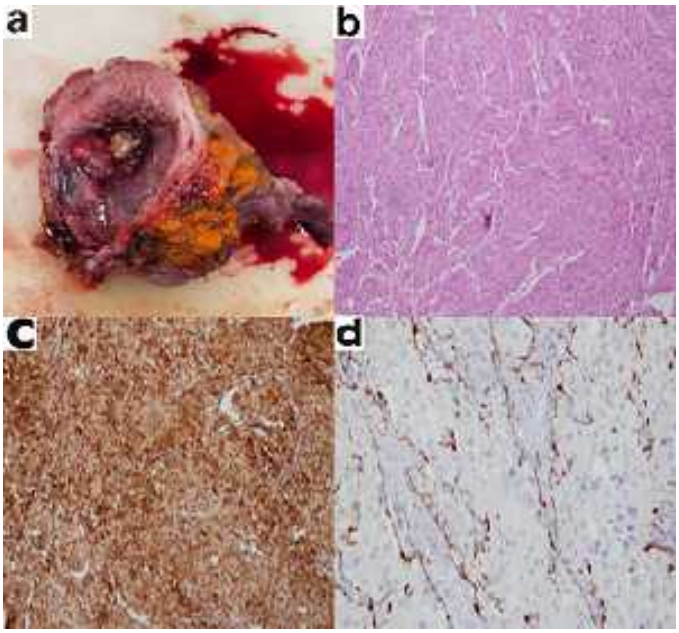

Figure 1: *Histopathological and Immunohistochemistry results: (a) Polypoid Tumor in posterior wall of urinary bladder, (b) Zellbellen pattern, (c) Chromogranin positive 100x_446 (d) S100 staining sustentacular cells 200x_446*

Discussion

Pheochromocytomas, which release catecholamines, are produced by sympathetic nervous system and adrenal glands. Paragangliomas, or pheochromocytomas outside the adrenal gland, make up around 15% of all pheochromocytomas.[5] Paragangliomas[6] have been linked to the B, C, and D subunit-coding genes of mitochondrial Complex II succinate dehydrogenase. Within the category of nonurothelial bladder tumours, paraganglioma is an uncommon tumour of the urinary bladder. Zimmerman first recognized it in 1953. Silent macroscopic hematuria (60 percent of recorded cases), paroxysmal hypertension, as well as "urinary attacks" are the traditional trio of clinical signs. Because of the bladder's specific position, these tumours produce a distinct set of symptoms that are frequently associated with micturition or severe bladder distension, which releases catecholamines. They are most typically found at the bladder's trigone or dome. Pre-operative diagnosis is frequently thought to be the cause based on the prevalent clinical symptoms and indicators. By quantifying biogenic amines in both urine and plasma, this suspicion is subsequently confirmed.[7] The location of the original tumor and any metastases can be determined using both CT scanning and MRI. However, phaeochromocytoma identification using iodine metaiodinebenzylguinidine (MIBG) scanning is exceedingly sensitive and specific. Considering the rarity of malignant bladder paragangliomas, many individuals may benefit from full surgical removal of tumour.[8] Many experts recommend total surgical removal as the standard of care for paragangliomas, including partial cystectomy or radical cystectomy as in our case, because they originate from the intramural area of the bladder wall (and are therefore difficult to completely remove with TURBT alone). Nevertheless, some surgeons still use TURBT and check their patients frequently for tiny cancers in certain body areas. For the treatment of locally recurrent and metastatic paraganglioma, radiation therapy and chemotherapy have not shown to be very effective. Due to its rarity, it has a tendency to be mistaken for urothelial carcinoma; however, distinctive histomorphological and immunohistochemical traits help in the accurate identification. Because it involves the muscularis propria (detrusor muscle) layer, its morphology is similar to that of urothelial carcinoma, especially in transurethral resection specimens; it is also misdiagnosed because it is not included in the histological differential diagnosis when evaluating a bladder tumour, and only a small percentage of patients have catecholamine-associated symptoms that migrate. On the basis of histology, benign from malignant tumors cannot be consistently distinguished by any specific traits. The "Zellballen" pattern, which is the most well-known pathological outcome, is characterised by immunohistochemical staining for synaptophysin and chromogranin that is commonly positive, with S-100 being predominant in sustentacular cells as in our case. The only reliable indicator of malignancy, according to a review of research by Pattarino et al. that considered additional parameters (such as biochemical and genetic markers) for this purpose, is the spread of metastatic disease.[9,10]

Conclusion

Paraganglioma in the urinary bladder is unusual. While characteristics like vascular invasion, highly invasive growth patterns, and recurrence usually indicate a bad prognosis, young age, severe local illness, and micturition attacks are risk factors for cancer. Further study into this uncommon illness is necessary because there is currently no treatment other than surgical excision for these patients. It is important to do a thorough family screening and look for potential neoplastic associations. This case illustrates a clinical presentation of bladder paraganglioma and provides a therapeutic management strategy for this rare condition. We would like to contribute our experience to the few cases that have already been reported because of its rarity.

Conflict of Interest: The authors declare no conflict of interest.

Funding Source: None

Authors' Contribution:

SI: Conception and design, or acquisition of data, Drafting the article or revising it critically for important intellectual content. Final approval of the version to be published.

NBN: conception and design, or acquisition of data, Drafting the article or revising it critically for important intellectual content. Final approval of the version to be published

NZ: conception and design, or acquisition of data,

Drafting the article or revising it critically for important intellectual content. Final approval of the version to be published.

AR: conception and design, or acquisition of data, Drafting the article or revising it critically for important intellectual content. Final approval of the version to be published.

SI: Drafting the article or revising it critically for important intellectual content. Final approval of the version to be published. Agreement to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

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