CASE REPORTS

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Pheochromocytoma of the urinary bladder – A case report

Feohromocitom mokraćne bešike

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Abstract

Introduction. Pheochromocytoma of the urinary bladder is a rare tumor and presents less than 0.06% of all urinary bladder tumors. **Case report.** We presented a 49-year-old female patient with a history of daily paroxysmal hypertension accompanied with flushing of the face and upper chest, palpitations and excessive sweating prior to micturition. Ultrasonography reported a 3 cm bladder wall tumor. The ^{131}I-metaiodobenzylguanidine (^{131}I-MIBG) scan showed a pathological isotope accumulation in the projection of the bladder. The patient underwent a partial cystectomy. One year following the operation the patient was normotensive and without recurrence. **Conclusion.** The most efficient treatment option for bladder pheochromocytoma is surgical resection. The most important fact in the diagnostics is suspicion on this rare condition.

Key words:
pneochromocytoma; urinary bladder neoplasms; ultrasonography; radionuclide imaging; urologic surgical procedures.

Apstrakt

Uvod. Feohromocitom mokraćne bešike je veoma redak i javlja se kod manje od 0,06% svih tumora mokraćne bešike. **Prikaz bolesnika.** Prikazali smo bolesnicu, staru 49 godina, sa svakodnevnom paroksizmalnom hipertenzijom praćenom crvenilom lica i gornjeg dela grudnog koša, lupanjem srca i preznojavanjem koji su prethodili aktu mokrenja. Ultrasonografski, viđen je tumor mokraćne bešike, promera 3 cm. Metajod-benzil-guanidin (MIBG) sken pokazao je patološku akumulaciju izotopa u projekciji mokraćne bešike. Bolesnici je učinjena parcijalna cistektomija. Godinu dana nakon operacije, bolesnica je bila normotenzivna, bez recidiva tumora u mokraćnoj bešici. **Zaključak.** Najefikasniji vid lečenja feohromocitoma mokraćne bešike je njegovo hirurško uklanjanje. Najvažnije u postavljanju dijagnoze je to da lekar posumnja na ovo retko oboljenje.

Ključne reči:
feohromocitom; mokraćna bešika, neoplazme; ultrasonografija; scintigrafija; hirurgija, urološka, procedure.

Introduction

Pheochromocytoma of the urinary bladder is a rare tumor which presents less than 0.06% of all urinary bladder tumors and less than 1% of all pheochromocytomas [1, 2]. They are tumors of the chromaffin tissue of the sympathetic nervous system of the urinary bladder wall [3]. They are most frequently derived from the adrenal medulla and in approximately 10% of cases are extraadrenal [4]. In the genitourinary tract the most frequent localization is the bladder (79.2%), urethra (12.7%), renal pelvis (4.9%) and the ureter (3.2%) [2]. Aside from the localization features the diagnosis and treatment of these tumors is not much different than pheochromocytomas of other localizations.

Case report

We presented a 49-year-old female patient with a history of daily paroxysmal hypertension (from 220/130 mmHg), accompanied with flushing of the face and upper chest, palpitations and excessive sweating. The paroxysmal hypertension was preceded by morning urination. Ten minutes following the attack there was a spontaneous decrease of the blood pressure. Similar lower intensity episodes which the patient subjectively better tolerated were reported occasionally during the day after voiding. The concentration of vanillylmandelic acid (VMA) in the 24 hour urine was in two occasions in normal range. Also, normal levels of epinephrine and norepinephrine were recorded.

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Ultrasonography of the lower pelvis reported a 3.2 × 2.8 × 1.8 cm well vascularized, clearly defined tumor localized on the edge of the left lateral and anterior wall of the bladder (Figure 1). Computed tomography (CT) of the chest, abdomen and lower pelvis showed a round mass approximately 1 cm behind and under the main bronchi that can be by differential diagnosis a ganglion or lymph node. Also there was an oval well vascularized mass, 2.3 × 3.4 cm in diameter on the margin of the anterior and left lateral wall (Figure 2). The ^{131}I-metaiodobenzylguanidine (MIBG) scan showed a pathological isotope accumulation in the projection of the bladder (Figure 3). The patient underwent partial cystectomy. The tumor was identified in the bladder wall covered by intact mucosa (Figure 4). During the operation a few hypertensive episodes occurred that were revolved. No intraoperative or postoperative complications were registered.

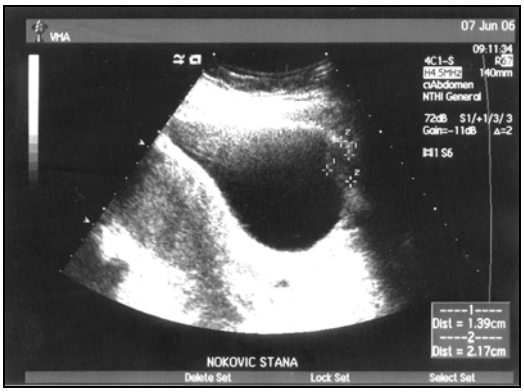


Fig. 1 – Ultrasonography shows a tumor mass at the edge of the bladder wall.

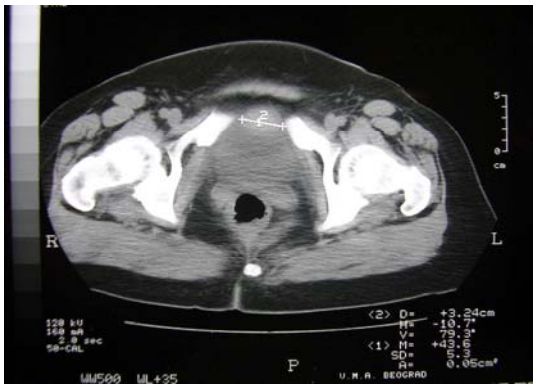


Fig. 2 – Computed tomography scan shows a tumor mass at the edge of the anterior and the left lateral bladder wall 2.5 × 3.5 cm in diameter.

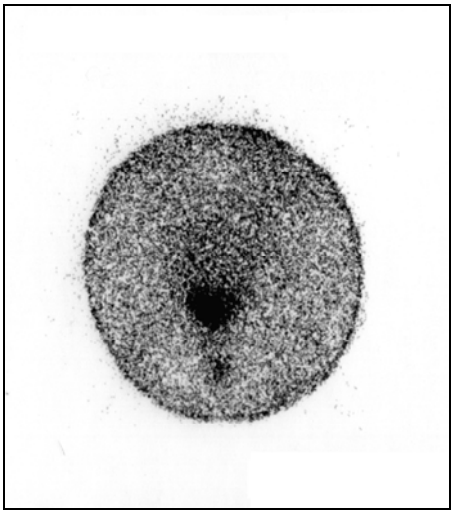
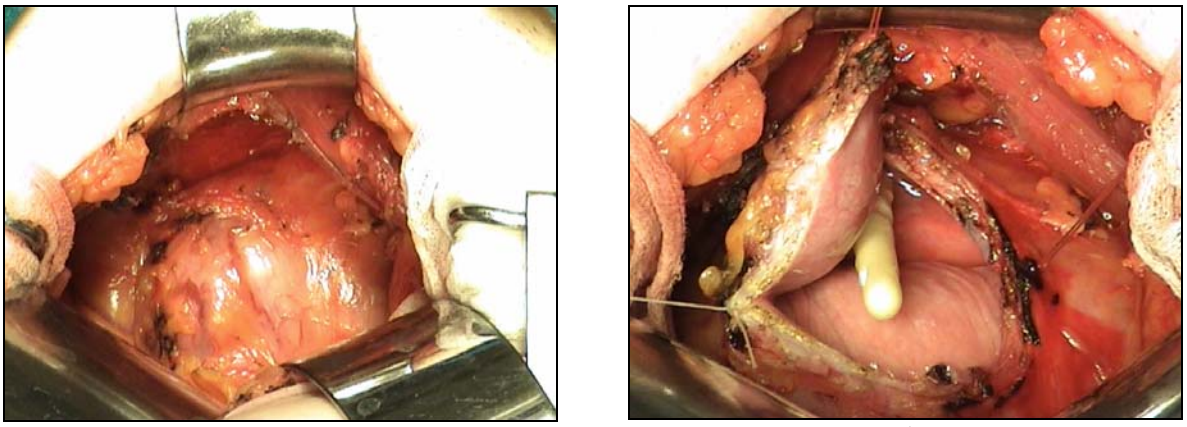


Fig. 3 – The ^{131}I-metaiodobenzylguanidine (MIBG) scan shows pathological isotope acumulation in bladder.



A) **B)**

Fig. 4 – A) Urinary bladder; B) Resection of the urinary paraganglioma.

The patient was discharged on the day 9 after surgery. Pheochromocytoma of the urinary bladder was confirmed by pathological and immunochemical examination. One year following the operation the patient was normotensive and without recurrence. The MIBG scan showed no pathological accumulation of the isotope.

Discussion

The first case of pheochromocytoma of the bladder was published by Zimmerman et al. [5] in 1953. and from that time about 200 cases have been published. Pheochromocytoma of the bladder can be diagnosed in children and the elderly, at the age of 10 to 80 years [6, 7]. It is more common in the female population [8–10]. The localization of these tumors differs in various studies. By some authors they are solitary tumors that most frequently arise on the dome or trigone of the bladder, while by other authors they are more often on the lateral bladder walls [9, 11]. Approximately 10% of urinary bladder pheochromocytomas are malignant [2]. Some authors consider invasion into surrounding organs and tissues or lymph node involvement as a criteria for malignancy [12, 13]. The presence of necrosis, angiolymphatic invasion, increase of mitosis and DNA ploidia indicate the malignant potential of the pheochromocytoma [1, 5, 14–19]. The current literature reported clinical signs range from the micturition attacks of headaches, palpitations, visual disturbances, tachycardia, sustained or paroxysmal hypertension, hematuria, dysuria, suprapubic pain or sweating after voiding [20, 21]. In around 17% of cases tumors are hormone inactive with practically no symptoms or can be presented only by pain or hematuria [13, 19, 20]. Only 40% of cases are preoperatively diagnosed, and the majority got adequate diagnosis after bladder surgery [21]. The suspicion on bladder pheochromocytoma with the presence of the characteristic symptoms should be investigated using imaging techniques (ultrasonography, CT) measurement of hormonal metabolites, and visualization *via* cystoscopy [21–23]. MIBG scan is highly recommended in determining the localization and diagnosis of pheochromocytoma. Its specificity is very high (nearly 100%) [24]. Biochemical measurements of urine/plasma catecholamine's and their metabolites are mandatory, such as measurement of norepinephrine 3 hours prior and after voiding [15, 21]. The most efficient treatment option for pheochromocytomas is surgical resection. The preoperatively confirmed diagnosis and adequate preparation facilitate the safety of surgical treatment [25]. Partial cystectomy is the first choice option [8, 9, 21, 24]. There are studies where tumors were treated by transurethral resection with or without hypertension attacks and recurrence, especially tumors that were not preoperatively diagnosed as pheochromocytomas [26, 27]. Nowadays, these tumors are treated by laparascopy or by robot assisted partial cystectomy with pelvic lymph node resection in the centers of excellences [28–30].

In the follow-up for benign tumors it is recommended to do the biochemical measurements of urine/plasma catecholamine's and their metabolites every 3–6 months and ^{131}I-MIBG and CT scan annually [15]. Patients with malignant tumors should have a monthly check of catheholamine levels and imaging studies twice a year [31]. Long term follow-ups are necessary because there are published cases with local recurrence and development of metastasis decades following surgical treatment [32].

Conclusion

Paraganglioma of the urinary bladder is a very rare tumor. When suspected the diagnosis can be reliably confirmed or excluded almost in all of the cases. Surgical resection is the standard treatment modality and requires the same preparation as in pheochromocytomas of different localizations. Almost all of the patients with benign tumors can be cured by surgery. The surgical mortality for all of the pheochromocytomas is around 2–3% with an experienced anesthesiologist and the surgeon. The histological confirmation of the tumor must be completed with immunohistochemical examination. Because of frequent recurrence, especially in malignant pheochromocytomas there must be a long term follow-up of these patients. The optimal treatment of these patients must be reserved for highly specialized medical centers.

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