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Bladder Paraganglioma: A Rare Tumor of the Urinary Tract

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SUMMARY: Paragangliomas are rare tumors of urinary tract and most common localization of urinary tract involvement is bladder. Malignant type is not frequent and complete recovery is possible with local excision in most of the patients. The diagnosis can be made with typical symptoms, such as painless gross hematuria, paroxysmal hypertension, and micturition attacks including headache, palpitations, diaphoresis and blurred vision during or after voiding. Imaging procedures are supportive to ensure the diagnosis. In this case we report a 68 years old woman with bladder paraganglioma who gained complete recovery with partial cystectomy.

KEY WORDS**:** Paraganglioma, bladder, surgery

ÖZET: Paragangliyomlar, üriner sistemin nadir görülen tümörleri olmakla beraber üriner sistemde en sık mesaneyi tutarlar. Malign tipi pek görülmez ve genelde bir çok hastada lokal eksizyon ile tam iyileşme sağlanabilir. Tanıya yardımcı tipik semptomlar arasında; ağrısız gros hematüri, paroksismal hipertansiyon ve işeme esnasında veya sonrasında oluşan başağrısı, çarpıntı, aşırı terleme ve bulanık görme atakları sayılabilir. Tanıyı kesinleştirmek için görüntüleme yöntemlerinden faydalanılır. Bu makalede, parsiyel sistektomi ile tam iyileşme sağlanan mesane paragangliyomlu 68 yaşında bir kadın olgunun sunumu yapılacaktır.

ANAHTAR KELİMELER: Paragangliyom, mesane, cerrahi

1. Introduction

Paraganglioma of the bladder is a rare entity and it accounts for less than %0.5 of all bladder tumors [1]. They usually derive from orbit, nose, ear, mediastinum, carotid area, duodenum and urinary tract. Bladder is involved most frequently among genitourinary structures (%79.2). Other common sites for genitourinary paragangliomas are urethra (%12,7), pelvis (%4,9) and ureter (%3,2) [2]. Malignancy rates are estimated to be approximately %10-15 but the histopathologic criteria for evaluating the malign transformation is lacking [1].

Bladder paragangliomas affect predominantly women and are usually located intramurally in the lateral and posterior wall and the trigone [3]. Derived from chromaffin cells, these tumors possess the potential to synthesize and secrete catecholamines, leading to some clinical symptoms, such as painless gross hematuria, paroxysmal hypertension, and micturition attacks including headache, palpitations, diaphoresis and blurred vision during or after voiding [4]. In this case we present a 68 years old woman with bladder paraganglioma successfully treated with partial cystectomy.

Case Report

A 68 years old woman was examined in our clinic suffering palpitation and headache during and after micturation. No gross hematuria was existed and 8 months before her visit she was operated for breast cancer and received chemo and radiotherapy subsequently.Ultrasound

imaging revealed a 3,2 x 2,2 cm bladder mass located superolaterally that protruded into bladder lumen. Cystoscopy was performed and no luminal pathology was seen but it was possible to see the bulge of a mass submucosally. Magnetic resonance imaging (MRI) showed a 2,2 cm mass located in the dome of the bladder without any intraluminal extension. Partial cystectomy was performed to the patient and the mass was completely excised. The pathologic examination was reported as "ganglioneuroma"(Figure1).

Histopathology revealed nests of cells with an infiltrative growth pattern in the bladder wall, which were composed of polyhedral cells with acidophilic to amphophilic cytoplasm and mostly centrally located nuclei. These nests were seperated by a thin delicate plexiform vascular network. No atypia or mitosis was identified. Neoplastic cells were positive for neuron-specific enolase (NSE), synaptophysin and chromogranin. Immunostain for S-100 showed sustentacular cells surrounding Zellballen nests. After operation, complaints of palpitation and headache were completely disappeared. The patient has been followed for 5 years in our clinic with no evidence of disease (Figure 2).

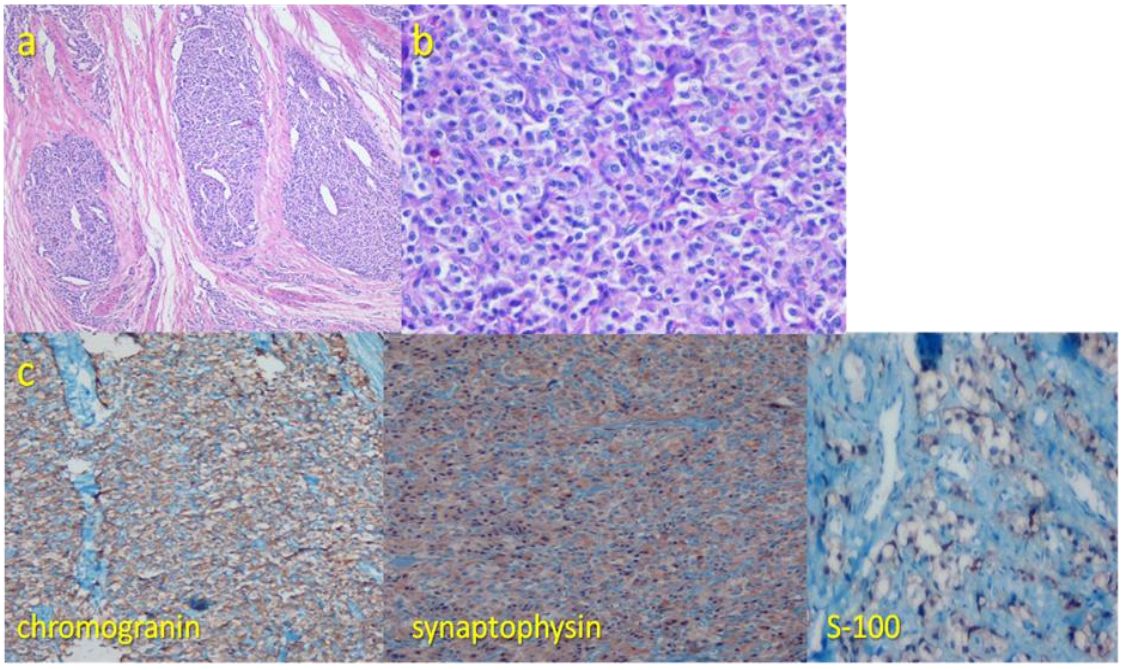

Figure 1. *Bladder paraganglioma with an infiltrative growth pattern in the wall of the bladder, H&E x100 (a). Classical zellballen pattern of a bladder paraganglioma, H&E x200 (b). Immunohistochemistry showed diffuse cytoplasmic staining of the cells with chromogranin and synaptophysin which were surrounded by S100 (+) sustentacular cells (c).*

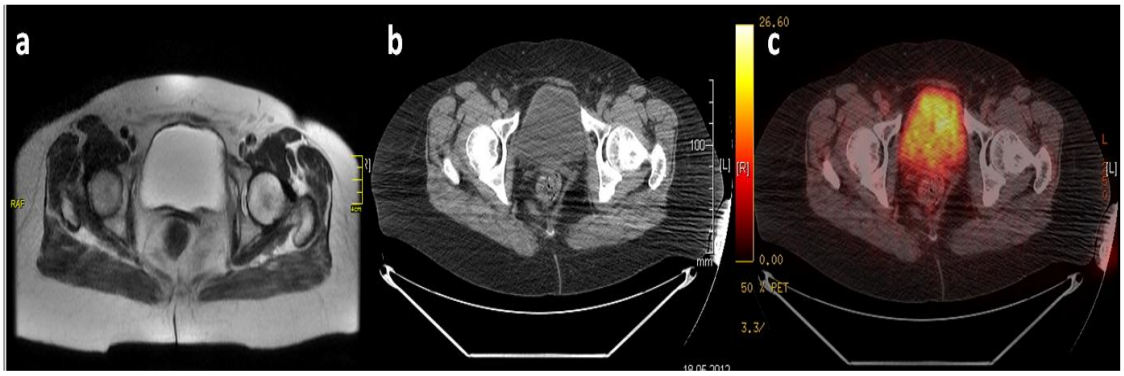

Figure 2. *MRI (a), computerized tomography (b) and 18 FDG positron emission tomography (PET) (c) images showing no evidence of recurrent disease 5 years after operation.*

2. Discussion

Bladder pheochromocytoma occurs more frequently in women than in men, and mainly during the third decade of life. It has been estimated that only 17% of bladder pheochromocytoma patients are hormonally nonfunctional and it is called as paraganglioma. The tumor is usually benign, only 15–20% being malignant [5]. The patient typically suffers from hypertensive crises that may be accompanied by headache, palpitations, hot flushes and sweating. These crises are mainly provoked by micturition, overdistention of the bladder, defecation, sexual activity, ejaculation, or bladder instrumentation. Haematuria is the presenting complaint in about 60% of reported cases [6]. Presenting symptoms of paraganglioma are mainly due to catecholamine overproduction. Some specific symptoms may also be present depending on the mass localization. In fact, patients with bladder paraganglioma may present with hematuria, post-micturition syncope and upper urinary tract dilatation when the tumor develops near the ureteral orifice [7]. Our patient showed palpitation and headache only during and early after the micturition episodes. Measurement of catecholamines and their metabolites (metanephrine, normetanephrine, vanillylmandelic acid, homovanillic acid and dopamine) in blood and urinary samples is useful but its negativity does not exclude the diagnosis of paraganglioma. In our patient none of these measurements were positive. After biochemical confirmation, additional radiological imaging should be performed to locate the tumor. The most useful imaging technique to localize primary and metastatic paragangliomas of the urinary bladder are computed tomography or magnetic resonance imaging [8].Surgical resection is the most common treatment of choice and to avoid hypertensive crisis during the mass manipulation, preoperative treatment with alpha and beta-blocking agents is required, starting 1–3 weeks before surgery [7]. The tumor usually involves multipl layers of bladder wall and for better distinction of tumor margins, partial cystectomy should be made by open surgery. If the lesion is adjacent to any ureteral orifice, removal of the distal part of the ureter is also necessary to obtain a radical excision of the tumor. In the presence of proven metastasis, radical cystectomy with pelvic lymphadenectomy is recommended [7]. Dilbaz et al. [9] first reported a case treated with the laparoscopic approach and in the literature, there samples of cases treated successfully with laparoscopy. In our case, we successfully performed an open partial cystectomy.

Intra-operative blood pressure was stable during the operation, and the margins were negative for tumor.Most of bladder paragangliomas grow slowly and have a good prognosis. All these patients have to be followed up regularly for the bladder function after partial cystectomy as well as the surveillance for recurrence. The blood pressure should be monitored regularly and periodic endocrine and radiological evaluation like I-MIBG and MRI is essential. Life-long follow up is recommended for these patients as recurrence or metastasis cannot be predicted on the histology alone. When recurrent or metastatic pheochromocytoma is demonstrated, surgical removal or debulking is the treatment of choice. Radiotherapy and chemotherapy are of limited effectiveness in the treatment of locally recurrent and metastatic pheochromocytoma [10].

3. Conclusion

Paragangliomas are rare tumors of the bladder. There are typical symptoms due to catecholamine secretion especially associated with micturation periods. They are rarely malignant and most of the tumors are treated successfully with partial cystectomy. In selected patients minimally invasive treatment methods can be performed like trans-urethral resection and laparoscopic excision. We treated a patient with bladder paraganglioma by open partial nephrectomy and no disease progression was reported after 5 years.

REFERENCES

1. Hanji, A.M. Rohan, V.S. Patel, J.J. Tankshali, A.(2012). Pheochromocytoma of the urinary bladder: a rare cause of severe hypertension. *Saudi J Kidney Dis Transpl*, 23(4), 813-6.
2. Dahm, P. Gschwend, J. E. (2003). Malignant non-urothelial neoplasms of the urinary bladder: a review. *Eur. Urol*. 44(6), 672-81.
3. Schaefer, I. M. Gunawan, B. Füzesi, L. et al. (2010). Chromosomal imbalances in urinary bladder paraganglioma. *Cancer Genet. Cytogenet*. 203(2), 341-4.
4. Chen, C. H. Boag, A. H. Beiko, D. T. et al. (2009). Composite paraganglioma - ganglioneuroma of the urinary bladder: a rare neoplasm causing hemodynamic crisis at tumour resection. *Can Urol Assoc J*. 3(5), E45-48.
5. Al - Zahrani, A. A. (2010). Recurrent urinary bladder paraganglioma. *Adv Urol*. 912125.
6. Das, S. Bulusu, N.V. Lowe, P. (1983). Primary vesical pheochromocytoma. *Urology*. 21(1), 20-5.
7. De Pasquale, V. Magistrelli, A. Diomedi Camassei, F. Caione, P. (2011). Unusual bladder mass in a 14-year-old boy: bladder paraganglioma. *Urol. Int.* 86(1),117-20.
8. Kappers, M.H.W. Van den Meiracker, A.H. Alwani, R.A. et al. (2008). Paraganglioma of the urinary bladder. *Neth J Med*. 66(4), 163-5.
9. Dilbaz, B. Bayoglu, Y. Oral, S. et al. (2006). Laparoscopic resection of urinary bladder paraganglioma: a case report. *Surg Laparosc Endosc Percutan Tech*. 16(1), 58-61.
10. Yadav, R. Das, A.K. Kumar, R. (2007). Malignant non-functional paraganglioma of the bladder presenting with azotemia. *Int Urol Nephrol*. 39(2), 449-51.