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Bladder Paraganglioma: A Case Report

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Abstract

Bladder paragangliomas are extremely rare in clinical cases. Due to their location between the submucosa and muscle layers and their potential malignancy, they are often misdiagnosed as bladder urothelial carcinoma. This tumor originates from the chromaffin cells of the bladder. This article reports on a case of an asymptomatic 30-year-old female patient with bladder paraganglioma and reviews relevant literature to deepen understanding of this disease.

Keywords

Paraganglioma, Bladder tumors, Treatment.

1. Introduction

Bladder cancer ranks 11th in global cancer incidence rates [1]. However, the incidence of bladder cancer has been gradually increasing in recent years [2]. Bladder paragangliomas, also known as bladder chemoreceptor tumors, originate from chromaffin cells in the sympathetic nervous system that produce catecholamines. They were first reported in 1953. They account for less than 0.05% of all bladder tumors and less than 1% of all chromaffin cell tumors. However, the bladder remains the most common site affected in the genitourinary tract [3]. There are literature reports showing slight differences in the incidence of bladder paragangliomas between genders, generally higher in females than males, and mainly occurring between 30 and 40 years old [4]. Due to the rarity of this type of tumor, there are few reports in the relevant literature, and more diagnosis and treatment methods need to be further explored.

2. Case presentation

A 30- years-old female was admitted due to the discovery of a bladder mass on routine physical examination. She did not complain of any urinary symptoms or discomfort. Color Doppler ultrasound reported the mass located on the right posterior wall of the bladder, measuring approximately 18x11x19cm, protruding into the bladder. Color Doppler flow imaging showed slightly abundant blood flow signals within the hypoechoic mass. Pelvic MRI plain scan with contrast enhancement revealed a nodular abnormal signal in the posterior wall of the bladder, connected to the bladder wall at its base, measuring approximately 1.1x1.4x1.4cm (see Figure 1). The signal intensity on T1-weighted imaging (T1WI) and T2-weighted imaging (T2WI) was slightly higher than that of normal tissue, with no definite exclusion of malignant lesions, recommending a puncture biopsy. Physical examination: Vital signs stable, no obvious abnormalities found in the heart, lungs, abdomen, or external genitalia. After admission, transurethral cystoscopy under lumbar epidural anesthesia and electrocautery of the bladder mass was performed. Intraoperatively, the posterior bladder mass was lobulated papillary and similar in size to the imaging data, and bilateral ureters were normally visible.The mass and partial bladder wall were completely excised, reaching the submucosal layer. Postoperative

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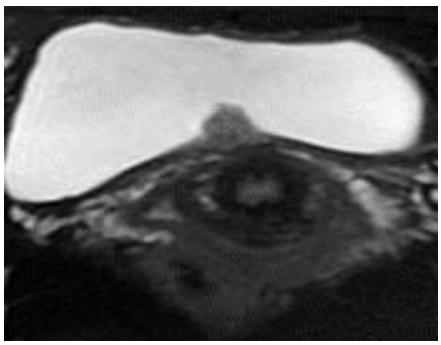
pathology reported a tumor cell nodular growth, with cells arranged in a sheet-like pattern, round in shape, rich in eosinophilic cytoplasm, round nuclei, prominent nucleoli, visible mitotic figures, and no obvious necrosis or vascular invasion (see Figure 2-3). Immunohistochemistry: S-100 (+)、Syn (+)、CD56 (+)、Ki67 (approximately 1%+)、CK-pan (-)、GATA3 (-)、GFAP (-)、CK7 (-)、CK20 (-)、P63 (-).The patient was followed up for 1 year after discharge, with no tumor recurrence detected.



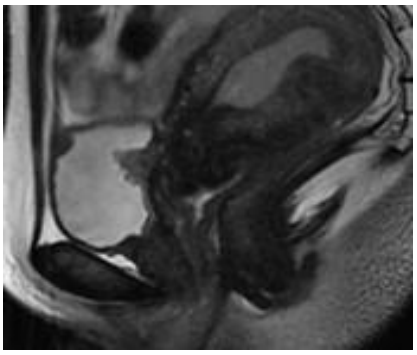
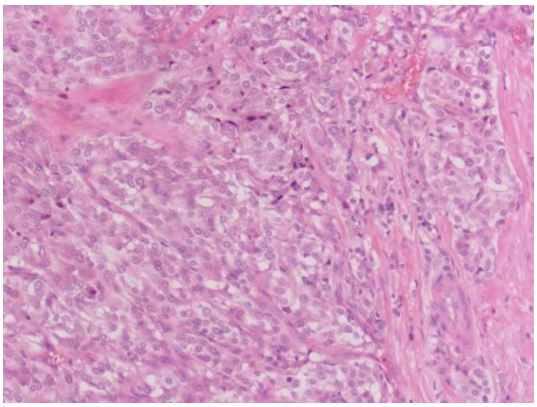


Fig. 1] (a) T2W transverse axis (b) T2W sagittal position.



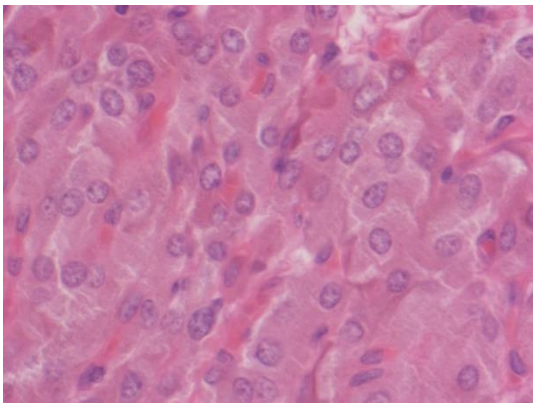


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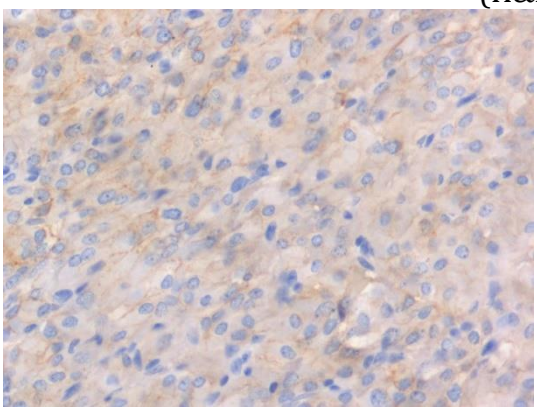
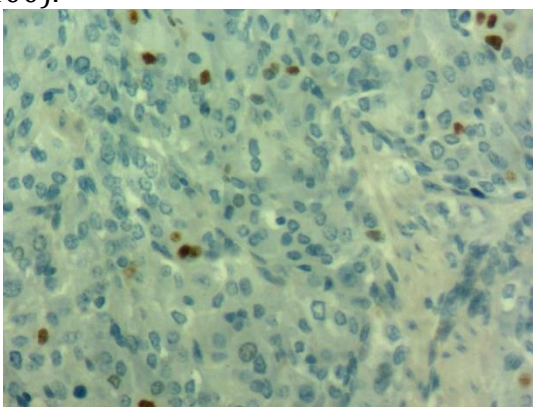
Fig. 2] Histopathological examination (hematoxylin-eosin staining): (a) (H&E, ×100), (b) (H&E, ×400).





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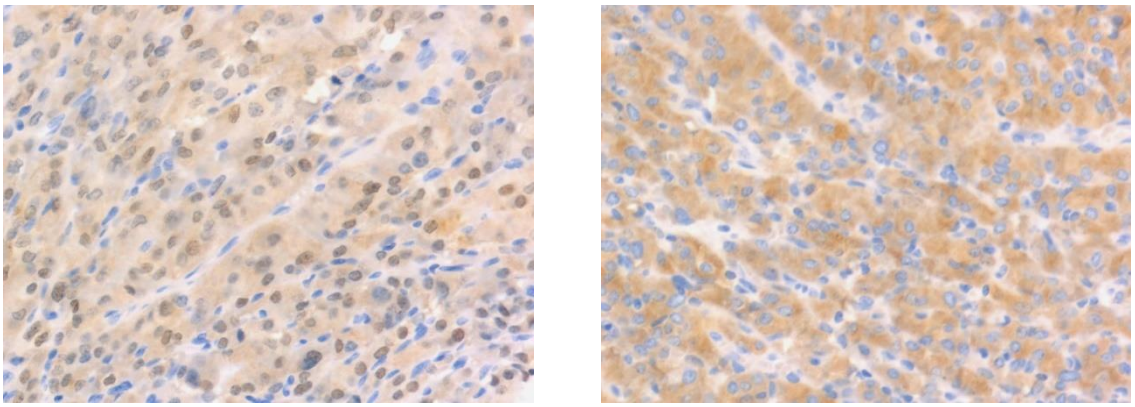

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Fig. 3] (a) Immunohistochemical staining of CD56 (x200), (b) Immunohistochemical staining of Ki67 (x200), (c) Immunohistochemical staining of S-100 (x200), (d) Immunohistochemical staining of Syn (x200).

3. Discussion

Bladder paraganglioma is a very rare type of tumor. Tumors are mostly located between the submucosa and muscle layers in the bladder dome or trigone, and they can be functional or non-functional. They are usually benign but have potential malignancy, with about 20% of paragangliomas exhibiting malignant behavior [5]. Patients may present with a wide range of symptoms, including no symptoms at all, detection of bladder mass only during physical examination, recurrent headaches, hypertension, palpitations, syncope during urination, or even painless gross hematuria as the main complaint [6]. Thus, sometimes requiring differentiation from bladder urothelial carcinoma.

Treatment options for bladder paragangliomas are diverse, including surgery, chemotherapy, and radiotherapy. However, the treatment approach should be individualized considering the tumor's progression. For localized or locally advanced tumors without metastasis, surgery is usually the preferred treatment, but adequate preoperative preparation is necessary to address potential complications such as hypertensive crisis, postoperative heart failure, and ARDS [7]. There are also literature reports indicating that the tumor has rich blood supply, and the patient's blood pressure may fluctuate due to repeated stimulation of the tumor during surgery, and it is easy to leave tumor residues. Therefore, transurethral resection of bladder tumor is not recommended [8]. Radioisotope therapy is recommended for patients who cannot undergo surgery or have multiple metastases, with 131I-MIBG being the most commonly used drug.

The diagnosis of this disease relies on pathological examination. Tumor cells typically express neuroendocrine markers such as CD56, Syn, CgA, NSE, and some cases may also express S-100 protein positively. Postoperative patients should undergo long-term follow-up, including ultrasound, CT scans, and long-term blood pressure monitoring [9]. Therefore, before treating patients with such tumors, it is necessary to conduct sufficient catecholamine-related hormone tests and imaging examinations to clarify the disease type, avoid misdiagnosis, formulate the best individualized treatment plan, and adhere to long-term follow-up.

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