Case Report

A case report of bladder paraganglioma and literature review

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ABSTRACT

Bladder paraganglioma is a rare tumor originated from the chromaffin cells of the bladder. We discuss a case of a 49-year-old male patient with bladder paraganglioma, including the clinical and ultrasonographic features, the histopathological and immunohistochemical manifestations, the treatment and prognosis, and the differential diagnosis of this disease. The combination of ultrasonic examination and clinical manifestations may help to make the accurate diagnosis of bladder paraganglioma, and pathological examination should be used to confirm the diagnosis.

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Introduction

Bladder paraganglioma is a rare tumor originated from the chromaffin cells of the bladder, accounting for about 0.06% of bladder tumors [1,2]. A case of bladder paraganglioma is reported in this paper. In order to improve the understanding of this disease, the clinical and ultrasonographic features, the histopathological and immunohistochemical manifestations, and the treatment and prognosis of the tumor are reviewed.

Case report

During routine physical examination in another hospital, the right bladder wall lesion was found through ultrasonic examination in a 49-year-old male patient. For further diagnosis and treatment, the patient came to The First Affiliated Hospital of Wannan Medical College (Wuhu, China). The physical and special examination findings of this patient were normal. Two-dimensional ultrasonic examination showed an oval hypoechoic nodule measured 18mm × 11mm with clear boundary in the muscularis of right bladder wall (Fig. 1). Color
Doppler flow imaging showed moderate blood flow signal in this nodule. And a suspicion of a solid lesion of bladder wall was given (Fig. 2). Multi-tumor markers showed that the tumor was likely to be benign. Laparoscopic partial cystectomy was conducted under general anesthesia. During the operation, the right bladder wall was found to be locally uplifted, which was considered as the location of the tumor. It was found that the tumor was a muscular mass about 1 cm × 1 cm in size. The tumor and the bladder wall around the tumor were resected and taken out of the body. One right pelvic drainage tube was placed, and the incision was sutured in turn. The operation went well. The family members of the patient reviewed the specimens because of their request, which is the usual procedure at our facility. And then the specimens were sent for pathology by the circulating nurse. The postoperative pathological diagnosis was bladder paraganglioma (Fig. 3). After operation, anti-infection, hemostasis, gastric protection, and fluid infusion were taken. The patient recovered well and was discharged on postoperative day 4, and is currently under regular follow-up. The patient has maintained a good condition so far and is satisfied with the treatment. The timeline was presented (Table 1). Informed written consent was obtained from the patient for publication of this case report.

Discussion

Zimmerman described the first bladder paraganglioma in 1953 [3]. The age of onset ranged from 20 to 40 years. The incidence rate of bladder paraganglioma in female is higher than that of male [4]. Bladder paraganglioma can be functional or nonfunctional. The main symptoms of functional bladder paraganglioma include hypertension, headache, palpitation, and intermittent painless gross hematuria. However, the clinical symptoms of nonfunctional bladder paraganglioma are not obvious.

Although cystoscopy is an important method for the diagnosis of bladder paraganglioma, it is generally not recommended as it may induce severe blood pressure fluctuations or bleeding [5].

Ultrasound imaging is helpful for the location of the tumor [6–8]. The ultrasonographic features of the bladder paraganglioma present as a mixed echo mass with abundant blood flow. Compared with transabdominal ultrasound, transvaginal ultrasound could more accurately detect the extent of tumor invasion. Through transvaginal ultrasonography, Bialek et al. revealed that the tumor had invaded a vaginal wall. However, by transabdominal ultrasound, the tumor infiltrating through

Fig. 1 – Two-dimensional ultrasonic image of bladder paraganglioma. An oval hypoechoic nodule measured 18 mm × 11 mm (arrow) was found on the right wall of the bladder.
a bladder wall and adjacent to a cervix had only been detected [9]. In our case, on the transabdominal ultrasonic examination, a tumor measured 18 mm × 11 mm in the muscular layer of the bladder was revealed. On color Doppler, a signal of moderate blood flow was revealed.

The ultrasonic features of bladder paraganglioma should be differentiated from angioma and bladder cancer. Angioma often occurs in young people. The typical ultrasonographic features of angioma present as a hypoechoic mass with small anechoic areas, which likes a "sieve hole" structure. The blood flow signal is moderate in some cases of angioma. Bladder cancer often occurs in the bladder triangle. Hematuria is the main symptom. The ultrasonographic features of the bladder cancer present as an irregular mass in the bladder with mixed

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**Fig. 2** – Color Doppler flow imaging of bladder paraganglioma. The blood flow in bladder paraganglioma was moderate.

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**Fig. 3** – Microscopic appearance of bladder paraganglioma. (A and B) H&E-stained slides (200×, ×400, respectively) reveal the round or polygonal tumor cells are distributed in the shape of cords and nests. The nucleolus is small and clear and the cytoplasm is transparent.
Like, the chemical glioma. The melanoma. Differently, the tumor was likely to be benign. The postoperative pathological diagnosis was bladder paraganglioma. The laparoscopic partial cystectomy was conducted on this patient. The patient recovered well and was discharged.

### Table 1 – The time elapsed from presentation of bladder paraganglioma.

| Time       | Items                                                                 |
|------------|----------------------------------------------------------------------|
| Day 1      | The physical and special examination findings of this patient were normal. |
| Day 2      | Ultrasonic examination gave a suspicion of a solid space occupying lesion of bladder wall. |
| Day 3      | Multi-tumor markers showed that the tumor was likely to be benign.       |
| Day 4      | The postoperative pathological diagnosis was bladder paraganglioma.      |
| Postoperative days 1-3 | Anti-infection, hemostasis, gastric protection, and fluid infusion were taken. |
| Postoperative day 4 | The patient recovered well and was discharged.                            |

Echo and abundant blood flow. The clinical symptoms of this case were not obvious, and we were not sure whether this lesion was angiosarcoma or bladder paraganglioma.

Paragangliomas are within 1 cm in size, and some of them are larger and may be multiple nodules. Histologically, the tumor is composed of polygonal or round epithelial tumor cells, which are arranged in strips, nests, and clusters. Most of the cells are consistent, with few mitotic figures. The stroma is rich in thin-walled vessels, and some of them are sinusoidal. Immunohistochemistry showed the characteristics of neuroendocrine, and the main manifestation was positive staining of chromaffin. In this case, the pathological examination showed that the round or polygonal tumor cells were distributed in the shape of cords and nests, and the nuclei were small and clear, and the cytoplasm was transparent. Immunohistochemical results were AE1/AE3(+), EMA(+), Syn(+), CD56(+), CgA(+), S-100(a few Sertoli cells +), and ki-67(+) < 1%.

Pathologically, bladder paragangliomas should be differentiated from the following diseases. The first is malignant melanoma. Malignant melanoma may occasionally occur in the bladder. Similar to bladder paraganglioma, in malignant melanoma, some cells could be presented as transparent or balloon-like. However, the HMB45 and S100 in malignant melanoma are often positive, while negative in paraganglioma. The second is clear cell adenocarcinoma of bladder. In clear cell adenocarcinoma of bladder, because of the large amount of glycogen, the cells are transparent, and can also be presented as nested and tubular, which are similar to the histological features of paraganglioma. However, immunohistochemical neuroendocrine markers are often positive in paraganglioma and negative in clear cell carcinoma. The third is bladder urothelium cancer. Just like bladder paraganglioma, the histological morphologies of the cells in bladder urothelium cancer are often nest-like. The bladder urothelium cancer is invasive with no clear boundary, and the cells of bladder urothelium cancer showed obviously malignancy. However, the boundary of bladder paragangliomas is often well-circumscribed and the tumor cells are arranged in nests or organs like, in addition, the nucleus of tumor cells are with less atypia. Immunohistochemistry also helps, neuroendocrine markers are often negative in bladder urothelium cancer.

Surgical resection is the main treatment for benign cases of bladder paraganglioma, and the postoperative results are good. Malignant cases are invasive, and had poor prognosis after treatment. For those who are ineffective in chemotherapy and radiotherapy, clinical symptoms should be controlled, including stabilizing blood pressure and correcting anemia caused by repeated hematuria. Patients with bladder paraganglioma should be followed up for a long time including ultrasonic examination, CT examination, long-term blood pressure monitoring and so on.

Enhanced computed tomography (CT) and magnetic resonance imaging (MRI) scanning are helpful in the diagnosis of bladder paraganglioma. A limitation in our study is that the patient hasn’t undergone CT and MRI examinations. In future studies, CT and MRI examinations will be carried out if possible.

### Conclusion

Sonographers should be aware of bladder paraganglioma since the ultrasonographic features of this entity resemble some other kinds of bladder tumors. The combination of ultrasonic examination and clinical manifestations may help to make an accurate diagnosis. However, in non-functional bladder paraganglioma just as our case, pathological examination should be used to confirm the diagnosis. Surgical resection is the main treatment for benign bladder paraganglioma.

### Ownership

All figures submitted are owned solely by the authors.

### Ethics approval

This study was approved by the local Ethics Committee. The study protocol was approved by the ethics committee and institutional review board of the First Affiliated Hospital of Wannan Medical College ([2019] Ethics research No. 87). The participant have consented to and placed no restrictions on the publication of his photographs.

### Availability of data and material

The datasets generated during and/or analyzed during the current study are available from the corresponding author on reasonable request.
Patient consent

All data published here are under the consent for publication. Written informed consent was obtained from the participant included in the study.

REFERENCES

[1] Persec Z, Buković D, Persec J, Sović T, Ljubanović D, Lambasa S, et al. Paraganglioma of the urinary bladder–clinicopathological, immunohistochemical and electron microscopy analysis—a case report. Coll Antropol 2012;36:1041–3.

[2] Siatelis A, Konstantinidis C, Volanis D, Leontara V, Thoma-Tsaglis E, Delakas D. Pheochromocytoma of the urinary bladder: report of 2 cases and review of literature. Minerva Urol Nefrol 2008;60:137–40.

[3] Zimmerman JJ, Biron RE, Macmahon HE. Pheochromocytoma of the urinary bladder. N Engl J Med 1953;249:25–6.

[4] Wang H, Ye H, Guo A, Wei Z, Zhang X, Zhong Y, et al. Bladder paragangioma in adults: MR appearance in four patients. Eur J Radiol 2011;80:e217–20.

[5] Zhang DX, Wang WY, Tian Y, Lv WC, Guo YW, Du LD. Diagnosis and treatment of paragangioma of the urinary bladder. J Clin Urol 2009;24:606–610.

[6] Xu DF, Chen M, Liu YS, Gao Y, Cui XG. Non-functional paraganglioma of the urinary bladder: a case report. J Med Case Rep 2010;4:216.

[7] Hermi A, Ichoua I, Kacem A, Hedhli H, Gargouri F, Khiai R, et al. Functional bladder paragangioma treated by partial cystectomy. Case Rep Urol 2019:2019:4549790.

[8] Priyadarshi V, Pal DK. Paraganglioma of urinary bladder. Urol Ann 2015;7:402–4.

[9] Białek W, Kawecki P, Dyndor K, Wronecki I, Rudzki S. Intraoperative decision to change the course of management based on an ultrasonographic image of urinary bladder paraganglioma—a case study. J Ultrason 2019;19:165–70.