Non Functioning Paraganglioma in the Urinary Bladder: A Case Report

Shangren Wang[1], Aiqiao Zhang[1], Shiqiao Huang[2], Yong Ma[2],*, Yongjiao Yang[1], Xiaoqiang Liu[1] , Ludong Zhang[2]

Paragangliomas are tumors that arise from autonomic nervous system. Non-functioning bladder paraganglioma is rare and usually misdiagnosed. Here we describe a case of a 45-year-old man with primary urinary bladder paraganglioma. The patient had no active signs and symptoms, and histological and immunohistological examinations of a transurethral resection specimen confirmed correct diagnosis. After successful transurethral resection of the tumors, the patient showed no signs of recurrence at one-year follow-up.

Keywords: pheochromocytoma; non-functioning paraganglioma; urinary bladder

INTRODUCTION

Primary urinary bladder paraganglioma is a rare neoplasm that develops from the chromaffin tissue of the sympathetic nervous system in urinary bladder wall. The diagnosis and prognosis of bladder paraganglioma has not been well established. In addition, non-functioning paragangliomas of the urinary bladder have rarely been reported [(1-4)]. Here we present a rare case of an asymptomatic patient with non-functioning paraganglioma of the urinary bladder.

CASE REPORT

A tumor of the urinary bladder was found during a routine physical examination of a 45-year-old man. The patient had been in a good health without abdominal and urinary tract symptoms. Routine hematological and biochemical examinations showed no evidence of hypertension. Ultrasound revealed a mass (1.4*1.1 cm) on the left wall of the bladder. Color Doppler sonography showed the mass with abundant blood supply (**Figure 1 A, B**). Non-contrast and contrast CT of the abdomen showed a solitary tumor protruding into the bladder (**Figure 1 C, D**). Metastatic disease was not found in other abdominal organs. The patient was diagnosed with synchronous nonfunctional paraganglioma in retroperitoneum and urinary bladder, and underwent a transurethral resection of bladder tumor. The mass was well circumscribed (**Figure 2A**), and tumor cells were arranged in a nested pattern (**Figure 2B**).

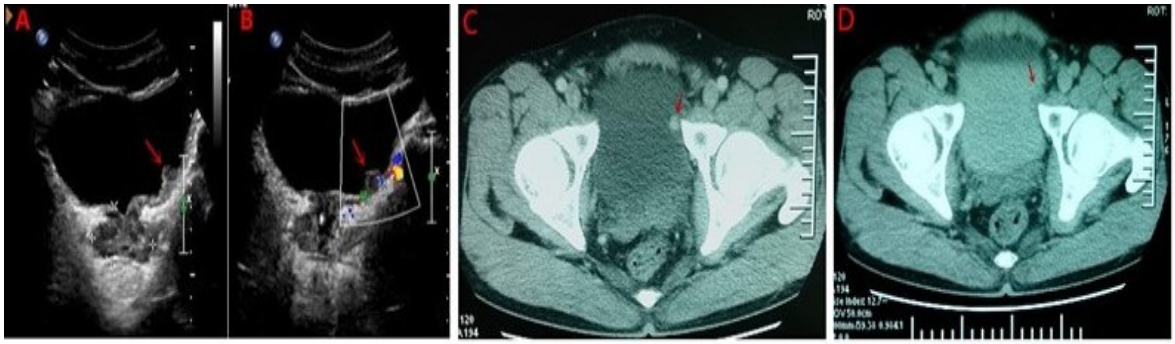

Figure 1. Ultrasound and computed tomography images. Ultrasound revealed a mass on the left wall of the bladder **(A, arrow)**, color Doppler sonography showed that the mass had abundant blood supply **(B, arrow)**, Non-contrast CT **(C)** and contrast-enhanced CT **(D)** showed a solitary tumor protruding into the bladder (arrow).

[1]Department of Urology, Tianjin Medical University General Hospital, Tianjin, 300052, China.
[2]Department of Urology, Shanxian Central Hospital, Heze 274300, China.
*Correspondence: Department of Urology, Tianjin Medical University General Hospital, Tianjin, 300052, China.
E-mail: xiaoqiangliu1920@126.com
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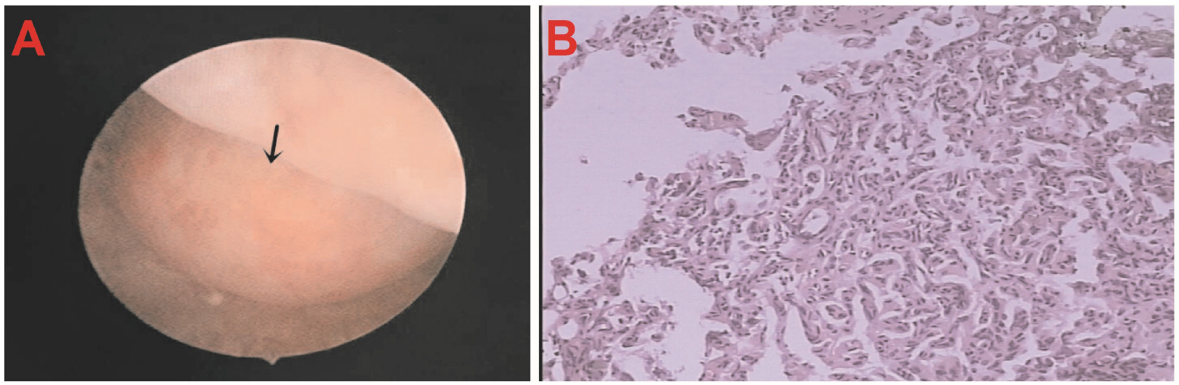

Figure 2. Cystoscopy and histological analysis. Bladder mass on left vesical wall with spherical shape and well circumscribed **(A, arrow)**. The tumor cells were arranged in a nested pattern (HE staining ×200) **(B)**.

Histopathological examination showed positive staining for NSE, Syn and CgA, and Ki-67 staining revealed a proliferation index of < 1%. On a recent follow-up after one year, ultrasound examination and cystoscopy examination showed no signs of recurrence.

DISCUSSION

Paraganglioma of the urinary bladder is extremely rare, accounting for less than 0.05% of all bladder neoplasm. In particular, 10-15% of such tumors are non-functioning and the patients had no history of hypertension, headache or flushing that would suggest a diagnosis of paraganglioma[5]. Thus non-functioning paragangliomas are more difficult to diagnose.

Imaging examinations are essential for preoperative localization of paragangliomas. Ultrasound showed the tumor as a submucosal homogeneous mass with continuous mucosa and abundant blood supply. Scintigraphy with I-123 labelled MIBG offered superior specificity than CT and MRI[6]. PET has an increased accuracy compared with MIBG scans for the localization of paragangliomas due to the higher spatial resolution of PET scanning[7]. The tumors in cystoscopy appeared as globular submucosal masses protruding into the bladder. However, the significance of diagnostic cystoscopy and biopsy is limited since it has a low positive rate, risk of bleeding and may provoke a hypertensive crisis if the tumor is functional. Therefore, histological and immunohistochemical diagnosis is important. The tumors show histological features similar to adrenal pheochromocytomas and the cells usually grow in a characteristic nested Zellballen pattern. Chromogranin, synaptophysin and NSE may aid the identification of neural tissue and neuroendocrine cells. A positive staining with synaptophysin, NSE, CgA and S-100 was observed in present case, which was compatible with paraganglioma[8]. Therefore, differential diagnosis of paraganglioma depends on histological and immunohistochemical aspects.

The most effective treatment of paraganglioma is surgery, including transurethral resection, partial or csytectomy and laparoscopy partial cystectomy. However, the optimal management mode is still uncertain. For patients with paroxysmal hypertension it is necessary to stabilize hypertension before the operation by using alpha-blocking drugs, similar to treatment for other pheochromocytomas[9]. In this case, although the patient was asymptomatic, we successfully performed a transurethral resection of the tumor to prevent its progression and metastasis, and blood pressure was stable during the operation. We chose conventional loop resection because the tumor was relatively small and this resection has little impact on the patient's quality of life. In addition, this technique has very low risk of bladder perforation.

Primary non-functioning bladder paraganglioma is easily misdiagnosed[10]. This case is unique because of two rare synchronous nonfunctional paraganglioma in retroperitoneum and urinary bladder without any clinical symptoms. If CT or cystoscopy reveals a mass well-defined that is located in the submucosa with an intact surface and there is no symptom of hematuria, a diagnosis of bladder paraganglionma must be considered. A definitive diagnosis may be reached by histological analysis.

CONFLICT OF INTEREST

The authors report no conflict of interest.

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