

Paraganglioma of Urinary Bladder

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INTRODUCTION

Paraganglioma of the urinary bladder is rarely encountered and its biological behavior is uncertain. It represents less than 1% of the bladder tumors and is usually benign. The tumor usually develops in young adult women.⁽¹⁾ The most prevalent involved sites are discussed controversially in the literature. In a recently published work, lateral and posterior walls of the bladder are described as the most prevalent sites, while in the latest edition of the *Campbell-Walsh Urology*, the trigones and the posterior wall have been stated as the most common tumor sites for paraganglioma.^(2,3) Common symptoms and signs are dysuria, hematuria, and hypertension, caused by local irritation of the tumor, and increased catecholamine levels. As many as 50% of the paragangliomas are hereditary and may be associated with familial paraganglioma, neurofibromatosis type 1, von Hippel-Lindau disease, and the Carney triad.⁽⁴⁾ We report a single case of paraganglioma in the urinary bladder primarily diagnosed as myoma.

CASE REPORT

A 37-year-old woman presented with frequency and dysuria. Physical examination showed no abnormalities. No constitutional symptoms suggesting an endocrine active paraganglioma, such as hypertension, were detected. Transabdominal ultrasonography showed a low-echoic mass at the bottom of the bladder. To prove the ultrasonographic diagnosis, conventional cystoscopy was performed and led to the diagnosis of a 2-cm myoma. Since infiltration of the muscle layer was ruled out by computed tomography, we decided to perform transurethral resection of the tumor (Figure 1). However, during the first transurethral resection, infiltration of the muscle layer was seen and confirmed by



Figure 1. Computed tomography of the pelvis showed the tumor mass on the right side of the urinary bladder.

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pathologic examination. The preliminary histological diagnosis of a granular cell tumor led to the decision for a second transurethral resection.⁽⁵⁾ Despite the high risk of bladder perforation, it was possible to resect the tumor in 2 surgical operations without any residual tumor tissue.

Endocrinologic examination showed physiologic levels of serum catecholamines. The histopathologic examination showed positive staining for CD56 (Figure 2A), synaptophysin (Figure 2B), and weakly positive but specific for chromogranin (Figure 2C). Sustentacular cells stained positive for S100 (Figure 2D). The Ki67 staining revealed a proliferation index lower than 2%. The patient was discharged without any complaints. Computed tomography of the pelvis and the abdomen in the 3-month follow-

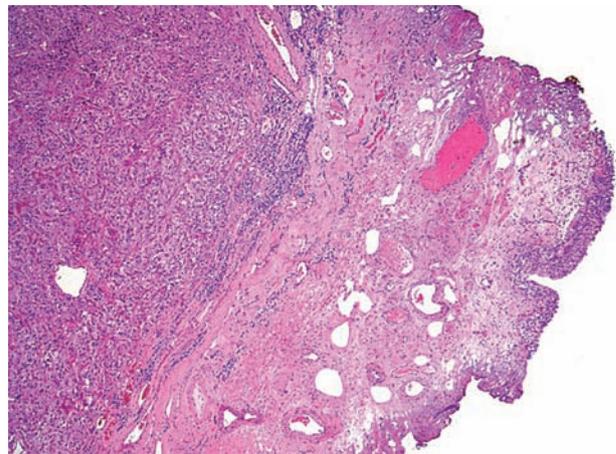


Figure 3. The picture shows the superficial urothelium (right) and the tumor (left; hematoxylin-eosin, $\times 100$).

up visit did not show any suspected lymph nodes or local recurrence.

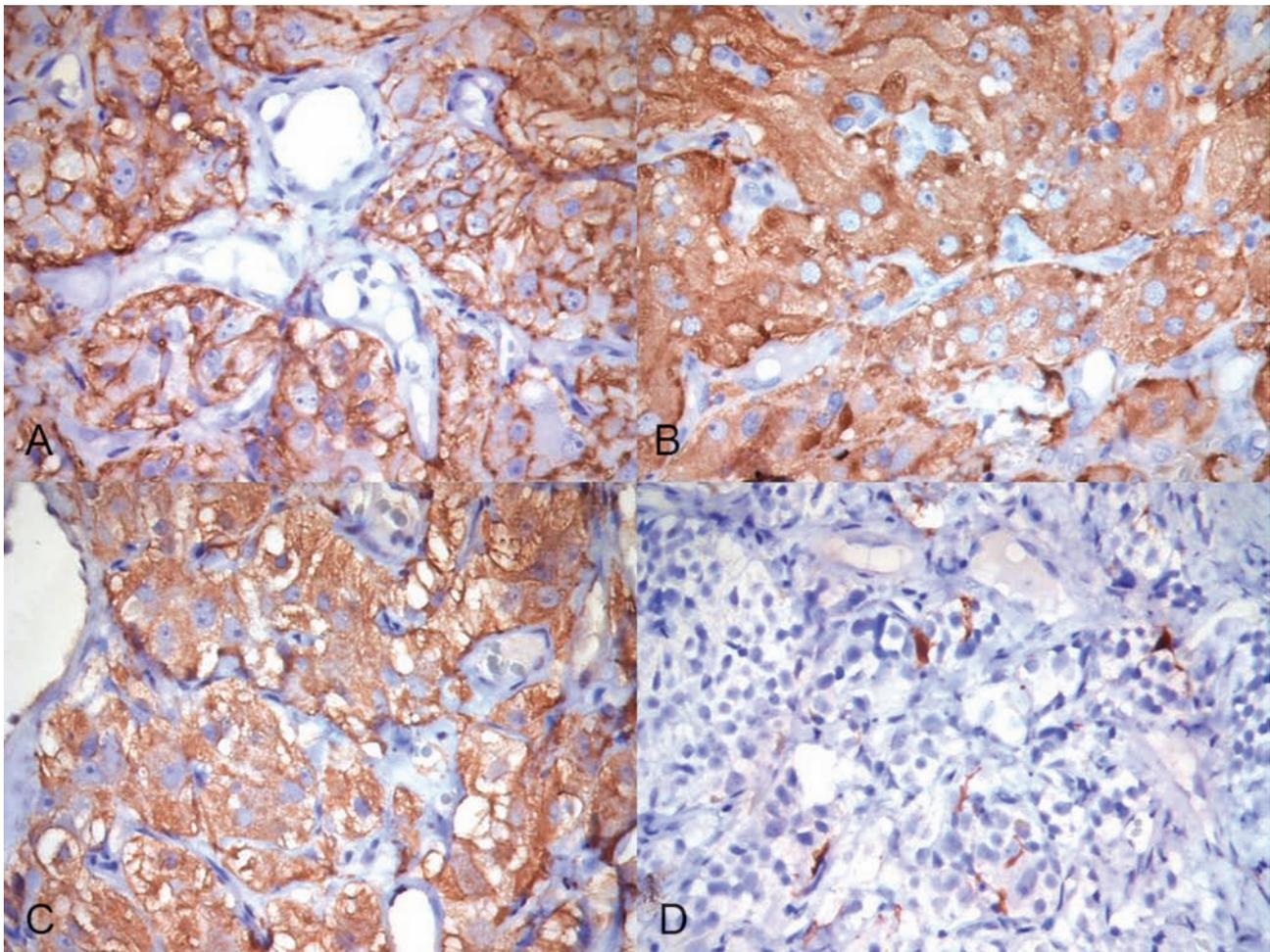


Figure 2. Immunohistochemical study of the tumor specimen for different stains shows the tumor cells ($\times 400$). Brown staining is indicative of a positive result for tumor. **A**, Positive staining for CD56. **B**, Positive staining for synaptophysin. **C**, Positive staining for chromogranin. **D**, Typically, between the tumor cells are located the sustentacular cells which are stained positive for S100 protein.

DISCUSSION

Paragangliomas are extra-adrenal neoplasms of the neural crest derivation, and if hormonally active, they are termed *pheochromocytoma*. There are at least 20 reported cases of the malignant paraganglioma in the English literature.⁽⁶⁾ Histologically, they are characterized by cells arranged in discrete nests separated by a prominent sinusoidal network. No defined histological features have been identified to safely distinguish benign from malignant paragangliomas. If the paraganglioma of the bladder is suspected, cystoscopy should only be performed following adrenergic blockade in a controlled environment such as an operating room. Biopsy should be avoided.⁽⁶⁾ Options for treatment include transurethral resection and laparoscopic or open cystectomy (partial or radical) with the aim of complete local excision of the tumor. Surgery is planned in a similar fashion as in adrenal pheochromocytomas and includes volume expansion and adrenergic blockade.⁽⁷⁾ Regarding the disposition of malignant paragangliomas to recur locally and develop

metachronous metastasis, lifelong follow-up is indicated.

CONFLICT OF INTEREST

None declared.

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