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Case report

Paraganglioma of the urinary bladder with pelvic metastasis

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ABSTRACT

A 52-year-old male, diagnosed with paraganglioma of the urinary bladder, underwent transurethral resection of the bladder tumor 10 years ago. He was lost to follow-up after the operation but was recently admitted to our hospital for the treatment of nasopharyngeal cancer. However, refractory hypertension with palpitation was noted and a computed tomography scan revealed a round, well-defined mass at the right pelvic region. Retroperitoneal tumor excision surgery was performed and a subsequent pathological analysis revealed paraganglioma. The diagnosis of paraganglioma of the urinary bladder with pelvic metastasis was confirmed and his blood pressure returned to normal level without medication after the operation.

1. Introduction

Catecholamine-secreting tumors that arise from chromaffin cells of the adrenal medulla and the sympathetic ganglia are referred to as pheochromocytomas and catecholamine-secreting paragangliomas (extra-adrenal pheochromocytomas), respectively. Approximately 10% of pheochromocytomas originate from extra-adrenal organs, primarily from para-aortic sympathetics. Bladder pheochromocytoma is a rare neoplasm accounting for less than 1% of all pheochromocytomas[1] and less than 0.06% of all bladder tumors.[2] The bladder pheochromocytomas usually arise from submucosal or muscularis propria and not from perivesical tissue and they are most common in the trigone region.[3] Malignant pheochromocytomas are histologically and biochemically the same as benign ones. The only reliable clue to the presence of a malignant pheochromocytoma is local invasion or distant metastases, which may occur as long as 20 years after a resection. We herein present a case of bladder paraganglioma with pelvic metastasis and discuss the clinical, image, and surgical findings.

2. Case report

A 52-year-old male had painless gross hematuria for days and he also mentioned urination with palpitation and headache for 2 years. He denied any history of hypertension or arrhythmia. An abdominal echo showed a bladder tumor over the right side of the urinary bladder. Cystoscopy revealed a submucosal protruding mass (Fig. 1A) and transurethral resection (TUR) of the bladder tumor was done on February 7, 2002. A pathological analysis confirmed paraganglioma. 123-I-metaiodobenzylguanidine (MIBG) scintigraphy was done and the results showed low probability of distant metastases (Fig. 1B). He was lost to follow-up in our outpatient department after the operation.

On February 3, 2012, he was admitted to our hospital for the treatment of nasopharyngeal cancer. However, refractory hypertension as high as 170–190 mmHg (systolic blood pressure) with palpitation was noted. Abdominal computed tomography (CT) was arranged and a 4.2 cm × 3.9 cm × 4.0 cm round, well-defined mass at the right pelvic region beside the urinary bladder (Fig. 2) was found. The 24-hour urine vanillylmandelic acid (VMA; 15.67 mg/day) was administered (normal range: 1–7.5 mg/day).

He was transferred to our urologic ward and an oral form of alpha blocker, terazosin hydrochloride (Hytrin), was administrated for 1 week prior to the operation. Then, retroperitoneal tumor excision surgery was performed on February 13, 2012. Fluctuating blood pressure (systolic blood pressure as high as 230 mmHg) was noted perioperatively, and sodium nitroprusside was administrated. However, it had poor response and perdipine (nicardipine) pump, a short-acting calcium-channel blocker, was used to control the blood pressure. We ceased surgery several times until the systolic blood pressure returned to normal level and the tumor was

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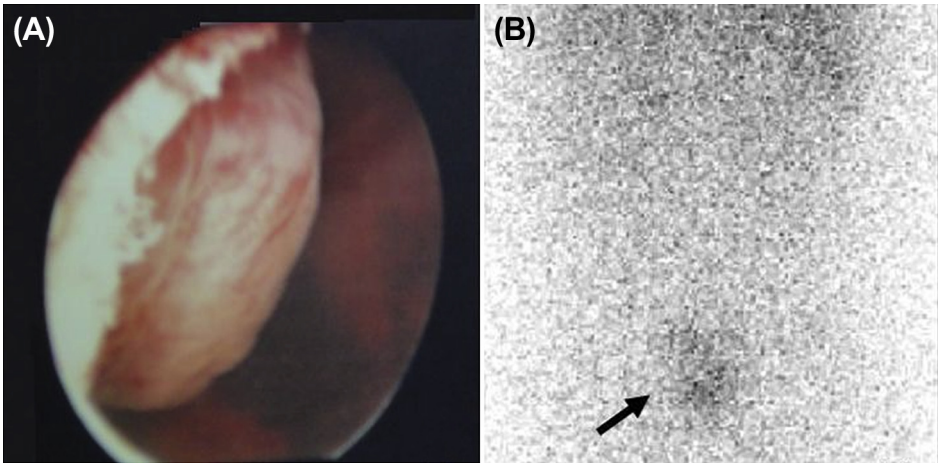

Fig. 1. (A) Cystoscopy showed a hypervascular, protruding mass over the right lateral wall of the urinary bladder. (B) I^{131}-MIBG revealed a high uptake at the urinary bladder and low probability of distant metastases. MIBG = metaiodobenzylguanidine.

finally removed smoothly (Fig. 3A and B). A pathological analysis of the isolated tumor confirmed paraganglioma (Fig. 3C and D). After the operation, the blood pressure became normal without medication and the patient was discharged uneventfully.

3. Discussion

Paraganglioma of the urinary bladder is a rare disease and accounts for 10% of extra-adrenal pheochromocytoma. The clinical presentation of paraganglinoma of the bladder is painless hematuria (50–60%), hypertension (65–80%), headache, and palpitation during micturition.[3] The average age at diagnosis is the 4^{th}–5^{th} decade and has an incidence of approximately 1:3 in men and women. Malignant pheochromocytomas are defined as tumors with local invasion or distant metastases. These tumors account for 10% of all pheochromocytomas. However, extra-adrenal pheochromocytomas are malignant in 29–40% of cases.[4,5] Thus, extra-adrenal pheochromocytomas might have a higher malignant potential than adrenal pheochromocytomas. For this reason, long-term follow-up should be recommended in patients with extra-adrenal pheochromocytoma.[6,7]

The diagnostic tests for paraganglioma of the urinary bladder include 24-hour urine or plasma fractionated catecholamines and metanephrines. Because bladder paragangliomas sometimes lack converting enzymes called phenylethanolamine *N*-methyltransferase, elevated norepinephrine levels can be found.[8] Imaging

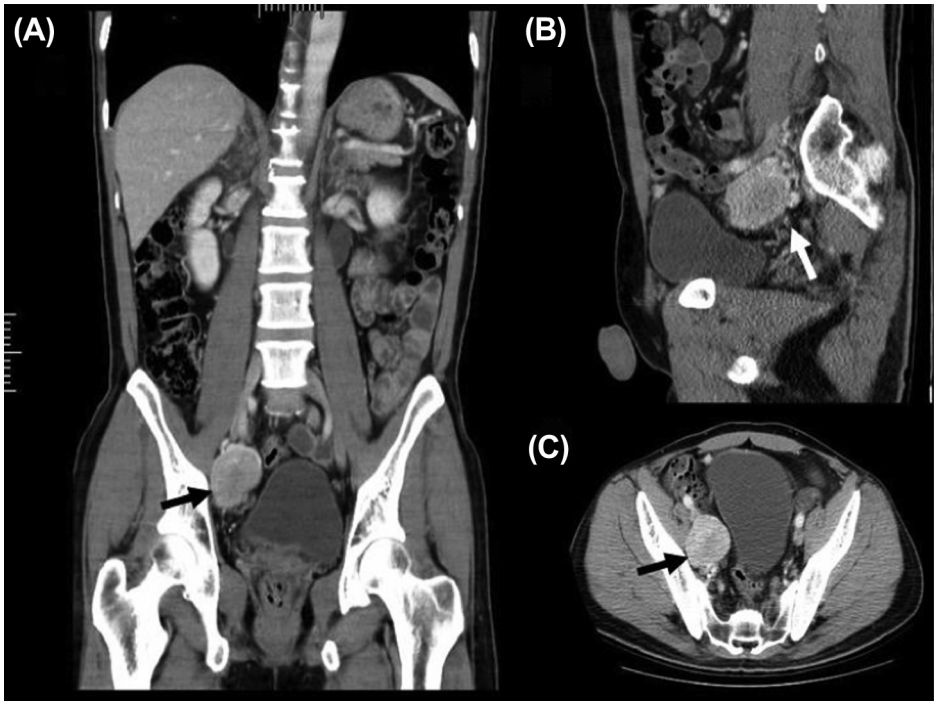

Fig. 2. Biphasic computed tomography revealed a 4.2 cm × 3.9 cm × 4.0 cm round, well-defined and enhanced mass at the right pelvic region beside the urinary bladder (arrow): (A) coronal view; (B) sagittal view; (C) transverse view. There was no connection between them.