Oncology

Symptomatic paraganglioma of the urinary bladder: A rare case treated with a combined surgical approach

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ABSTRACT

Pheochromocytomas are tumors of the embryonic chromaffin cells, originating from the embryonic neural crest. The pheochromocytomas developing at extra-adrenal locations are termed paragangliomas, which are extremely rare and account for almost 0.06% of all bladder tumors. In this report, we present a 23-year-old woman who presented with a one-year history of repeated episodes of dizziness, hypertension, intermittent hematuria, and nausea/vomiting that occurred during urination and was operatively treated due to a diagnosis of paraganglioma of the urinary bladder.

Introduction

Pheochromocytomas are tumors of the embryonic chromaffin cells, originating from the embryonic neural crest. Almost 18% of these tumors are reported at extra-adrenal locations and are termed paragangliomas. In the genitourinary system, bladder is the most common location for paragangliomas (79.2%), followed by urethra (12.7%), renal pelvis (4.9%), and ureter (3.2%). In this report, we present a PB case who presented with typical symptoms (hypertension, hematuria, dizziness during urination) preoperatively and treated with a combined surgical approach.

Case presentation

A 23-year-old woman was previously admitted to another center with a one-year history of repeated episodes of dizziness, hypertension, intermittent hematuria, and nausea/vomiting that occurred during urination. In that center, an ultrasound (USG) examination of the lower urinary tract revealed a mass in the bladder and then the patient underwent transurethral resection for bladder tumor (TUR-BT). After being diagnosed with PB based on histological findings, the patient was referred to our clinic. A 68Ga-DOTATATE positron emission tomography-computed tomography (PET-CT) scan revealed a 34 × 33 mm irregular lobular mass in the bladder (primary malignant process?), showing an intensive Ga-68 pattern and an exophytic component extending to the bladder lumen at the right upper-side wall and also revealed a 15 × 16 mm nodular lesion at the left wall, medial-lateral region of the bladder, showing an intensive Ga-68 pattern and an exophytic component (synchronous malignant process?). No regional or distant metastasis was observed [Fig. 1].

Partial cystectomy was planned for the two lesions detected on PET-CT. Before cystectomy procedure, cystoscopy was performed in the same session and revealed normal ureteral orifices and an impression of a solid tumor in a 3-cm area at the junction between the left lateral wall and the dome. However, the second lesion detected on PET-CT was not visible on cystoscopy. The mucosa and the detrusor tissue surrounding the tumor were incised using a resectoscope and a monopolar system. To define safe surgical margins, methylene blue was injected around the tumor [Fig. 2 A-B-C]. Subsequently, surgical exploration was performed through a suprapubic transverse incision. The second lesion that could not be visualized by cystoscopy was viewed at the right bladder dome, extending into the detrusor muscle and protruding into the bladder lumen. Using the safe surgical margins defined by methylene blue, the lesion was removed via partial cystectomy. The second mass was removed after being dissected from the detrusor muscle [Fig. 2D]. The integrity of the bladder mucosa in the surgical site remained intact.
During the surgery, typical complications including hypertension and tachycardia were observed and were controlled by medical therapies. The defect in the bladder wall was closed in two layers during partial cystectomy. In the excision site on the right dome, the detrusor tissue was closed in a single layer. The procedure was completed after inserting a 16 Fr Foley catheter and a perivesical sump drain in the bladder. Histological examination was reported as PB with muscular tissue invasion [Fig. 3A]. No tumor was detected at the surgical margins of both masses. All the symptoms resolved postoperatively and no marked pathological peptide pattern was observed in the control 68Ga-DOTA TATE PET-CT scan performed at postoperative month 6 [Fig. 3B]. At 18-month follow-up, the patient had no urinary complaints and the USG revealed no urinary pathology.

Discussion

Functional paragangliomas may manifest with various symptoms including hypertension, palpitation, and micturition syncope during catecholamine secretion. Similarly, our patient had a one-year history of typical PB symptoms and hematuria and thus was considered to have active and symptomatic PB.

Distinguishing between benign and malignant PBs in histological examination of these tumors is highly difficult. Grignon et al. reported that although DNA ploidy has been shown to be a predictor of malignant behavior in adrenal pheochromocytomas, it cannot be used as a diagnostic criterion for malignancy in PB. On the other hand, it is commonly known that patients with malignant tumors may require life-long follow-up due to the risk of long-term recurrence. In our patient, no metastasis was observed in the 68Ga-DOTATATE PET-CT scans performed preoperatively and at postoperative month 6, which confirmed that the tumor in our patient had a benign nature. Additionally, no local recurrence was observed at 18-month follow-up as well.

Although CT and magnetic resonance imaging (MRI) are commonly used in the detection of primary tumors and metastases, administration of these two techniques with I-131 labeled metaiodobenzylguanidine (MIBG) has been shown to provide high sensitivity and specificity in the diagnosis of pheochromocytomas. In contrast, 68Ga-DOTATATE PET-CT has also been shown to have a high diagnostic performance in detecting PB, albeit not as effective as MIBG. In our patient, 68Ga-DOTATATE PET-CT was used for pre- and post-operative assessment of metastasis and recurrence due to the nonavailability of MIBG in our hospital.

In patients with PB, total resection of the tumor is often curative due to the low risk of malignancy. Moreover, partial or radical cystectomy is the most effective treatment approach in cases accompanied by muscular invasion. On the other hand, there are some studies suggesting that cystoscopic resection could be sufficient if the surgery was performed by an experienced surgeon and if the surgeon could ascertain...
that the tumor base has been removed completely. We performed combined cystoscopy and open surgery and after assessing tumor localization, size, and safe margins on cystoscopy, the tumor was removed completely during open surgery.

Conclusion

Our patient was referred to our clinic with a histological diagnosis established after TUR-BT and thus our surgical and anesthetic teams were prepared for probable intraoperative symptoms associated with paragangliomas. Accordingly, we suggest that surgical and anesthetic teams should be prepared for probably intraoperative symptoms in patients presenting with typical symptoms of PB and should specifically keep in mind the high risk of perioperative hypertensive crisis during cystoscopy and tumor resection.

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Declaration of competing interest

There are no conflicts of interest in connection with this paper.

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