

Robotic assisted partial cystectomy with pelvic lymph node dissection for metastatic paraganglioma of the urinary bladder

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Paraganglioma of the urinary bladder is a rare tumor that often presents with hypertensive crisis during micturition. We herein present the unusual case of a 45-year-old

female with metastatic paraganglioma treated with robotic assisted partial cystectomy and pelvic lymphadenectomy. We review the literature regarding the evaluation and management of paraganglioma of the bladder..

Key Words: urinary bladder neoplasms, positron emission tomography, paraganglioma, robotic cystectomy, pelvic lymphadenectomy

Introduction

Paraganglioma (extra-adrenal pheochromocytoma) of the urinary bladder is a rare tumor, making up less than 0.06% of all diagnosed bladder cancers. It originates from embryonic rests of chromaffin cells in the sympathetic nervous system.¹ The diagnosis is usually made by measurement of urine metanephrines, catecholamines, vanillylmandelic acid, serum clonidine

or chromogranin A.² Only once in published data has metastatic urinary bladder paraganglioma been evaluated with PET-CT imaging.³ We present the first reported case of a patient undergoing robotic assisted partial cystectomy (RAPC) with pelvic lymph node dissection to treat the disease.

Case report

A 45-year-old Caucasian female with a past medical history of hypertension was evaluated for suprapubic discomfort and microscopic hematuria. An abdominal ultrasound suggested a bladder mass. The patient underwent transurethral resection of the bladder tumor which revealed paraganglioma in the muscularis

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propria. She denied history of severe headaches, palpitations, hot flashes or excessive sweating. A PET-CT utilizing 100 cc of intravenous Ultravist contrast and 14.2 mCi of Fluorine-18 FDG displayed a 15 mm hypermetabolic left external iliac lymph node with maximum SUV of 7.5 that was consistent with malignancy. Further, there was an abnormal soft tissue density at the base of the bladder measuring approximately 3.8 cm x 3 cm with maximum SUV of 13.5, Figure 1.

The patient was treated preoperatively with an alpha blocker and underwent a RAPC with bilateral pelvic lymph node dissection without complication. The boundaries of the lymph node dissection were as follows: iliac bifurcation proximally, node of Cloquet distally, genitofemoral nerve laterally, midline over the sacrum medially, and obturator fossa inferiorly. A total of 39 lymph nodes were removed. Final histopathological diagnosis was consistent with an extra-adrenal paraganglioma (3.8 cm x 1.9 cm x 1.6 cm) that stained positive for synaptophysin, chromogranin A, and S-100 with one lymph node positive for metastatic paraganglioma.

The patient was discharged on POD #1, urinary catheter was removed POD #8, and experienced no postoperative complications. At 6 months, the PET-CT revealed no evidence of active malignancy with normal levels of chromogranin A, neuron specific enolase, urine catecholamines and metanephrines.

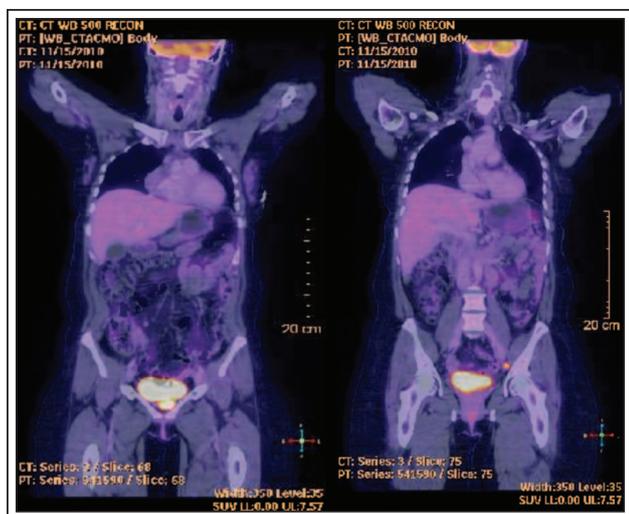


Figure 1. Coronal FDG PET-CT scan reveals the soft tissue mass at the base of the bladder with associated hypermetabolic activity, consistent with tumor (left) and the enlarged left iliac lymph node (right) is well visualized.

Discussion

Most publications of paraganglioma of the bladder have been in the form of case reports or small case series. However, Cheng et al, reported their experience of 16 cases over 53 years to determine indicators that could predict outcome. They determined that histological criteria (DNA ploidy, p53 alteration, MIB-1) had no definitive association with prognosis but tumor stage ($\geq T3$) and incomplete resection of T1 and T2 disease correlated with the risk of metastasis, death from disease, and recurrence.⁴ Our case was atypical in that the patient did not present with the usual hypertensive crisis, headache, diaphoresis, or palpitations which are typically seen in patients with urinary bladder paragangliomas.

To date, there is only one reported case of the use of PET-CT in urinary bladder paraganglioma. Thambugala and colleagues described metastatic urinary bladder paraganglioma with pelvic lymphadenopathy that was accurately staged with FDG PET-CT.³

Controversy exists regarding outcomes of robotic assisted radical cystectomy (RARC) compared to open radical cystectomy. A report of 100 consecutive cases of RARC revealed an average blood loss of 271 cc, OR time of 4.6 hours, 2.1 days to the passage of flatus, 2.8 days to bowel movement, 4.9 days until discharge, and a lymph node yield of 19. Overall there were 41 complications in 36 patients, with 8% being Clavien grade 3 or higher, with gastrointestinal (nausea, vomiting, ileus) and urinary tract infections being the most common. With no positive margins in any specimen, they concluded that RARC has comparable surgical and pathological outcomes to open cystectomy.⁵ The same authors also conducted a prospective randomized controlled trial of open versus RARC in 20 and 21 patients respectively. There were significant differences favoring RARC in mean estimated blood loss, time to flatus as well as bowel movement, and pain medication requirement. Of note, neither group had a positive surgical margin and mean nodal yield for RARC was 19 compared to 18 for open.⁶

Another debated topic is the effectiveness of robotic assisted pelvic lymphadenectomy at the time of radical cystectomy. To ensure staging accuracy, Capitano et al, reported that 25 lymph nodes could be the minimum for lymphadenectomy. Through their review of 731 patients over 20 years, there was a 75% probability of detecting lymph node metastasis if 25 lymph nodes were removed compared to only 50% for removal of 15 lymph nodes.⁷ Lavery and colleagues evaluated 15 consecutive cases of RARC. Their extended pelvic lymph node dissection borders were the genitofemoral

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nerve laterally, the node of Cloquet distally, 2 cm above the bifurcation of the aorta proximally, and the midline over the sacrum medially. Overall, their mean nodal yield was 41.8 (18-67). In comparison, a review of 120 patients in their open series revealed a mean nodal yield of 36.9 lymph nodes.⁸

Conclusion

In conclusion, even though current data shows promise with the use of robotics in bladder cancer and pelvic lymph node dissection, additional study is needed to determine the long term efficacy as well as oncological outcomes when compared to open surgery. □

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