Laparoscopic Excision of an Extra-Adrenal Pheochromocytoma (Paraganglioma) of the Organ of Zuckerkandl

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Abstract

Background: Paraganglioma of the organ of Zuckerkandl (OZ) is a rare surgically challenging tumor because of its critical location and the nature of catecholamine secretion. We describe the technique of laparoscopic excision as well as provide a literature review to confirm its feasibility.

Case Presentation: In a 23-year-old male patient, laparoscopic excision of a 5 × 4 cm tumor located at the aortic bifurcation and indenting the vertebral column was performed. Preoperatively, the patient received α - and β -adrenergic blockers as well as underwent sperm banking. The patient was put in the lateral position, five ports were used: four in the midline and one in the left iliac fossa. The tumor was approached by the reflection of the colon. Ureter, gonadal vein, and sympathetic chain were preserved. Dissection of the tumor from the inferior mesenteric artery was done followed by control of three feeding arteries and two draining veins posteriorly and inferiorly. The procedure was completed laparoscopically with minimal blood loss. Intraoperatively, three episodes of hypertension developed and required stoppage and the administration of vasodilators. The patient recovered on the second day postoperatively and all antihypertensive medications were stopped. At 1 year of follow-up, the patient is tumor-free but developed anejaculation for which he is under current treatment.

Conclusion: Laparoscopic excision of paraganglioma located at the OZ is safe and feasible.

Keywords: pheochromocytoma, organ of Zuckerkandl, paraganglioma, laparoscopic, adrenalectomy

Introduction and Background

PHEOCHROMOCYTOMAS AND PARAGANGLIOMAS are extremely rare tumors arising from the neural crest chromaffin tissue located at the adrenal medulla and the extra-adrenal sympathetic nerves, respectively. The origin of abdominal paragangliomas varies but preferentially arises from the organ of Zuckerkandl (OZ), which refers to a collection of chromaffin tissue found in a para-aortic location near the inferior mesenteric artery.¹

Paragangliomas located at the OZ pose a surgical challenge when compared with classic pheochromocytoma surgery. The origin of the draining vein/veins is not clear, the close relation with the great vessels and the risk of bleeding are high, and the gentle manipulation of the tumor that is required to minimize intraoperative spells, all make open

surgery more appealing for many surgeons. Herein, we present the technique we used for the excision of paraganglioma of the OZ.

Presentation of a Case

The patient is a 23-year-old male who presented first to the endocrine unit for secondary hypertension workup. The patient reported episodic attacks of palpitation, sweating, and headaches, and was maintained on two antihypertensive medications. The 24-hours of urinary metanephrine/normetanephrine was high, and postcontrast CT scan of the chest, abdomen, and pelvis showed enhancing soft tissue mass (5 × 4 cm) located at the level of the aortic bifurcation on the left side partially encasing the aorta and indenting the vertebral column (Fig. 1). ⁶⁸Ga DOTA PET/CT scan

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FIG. 1. Postcontrast CT scan findings. (A) Axial arterial phase demonstrating the relation of the tumor with the inferior mesenteric artery (red arrow). (B) Axial arterial phase demonstrating posterior feeding vessels (red arrow). (C) Coronal arterial phase showing two feeding vessels (red arrows). (D) Sagittal arterial phase showing the tumor is indenting the vertebral column.

confirmed the presence of the tumor with no extra or metastatic sites.

Preoperatively, the patient received α -adrenergic blockers for 2 weeks for rapid control of hypertension followed by a β -adrenergic blocker for 1 week to control tachyarrhythmias and angina symptoms. The patient consented for laparoscopic excision, and because of the location of the tumor, the process of ejaculation was thoroughly explained to the patient and he underwent sperm banking before surgery.

The surgical technique

Initially, the patient was placed in the left lateral decubitus position. The first port was placed using the open technique just above the umbilicus, then right-hand port below the level of the umbilicus and the left-hand port midway between umbilicus and xiphisternum were placed. Additional two ports were required during surgery and were fixed just below the xiphisternum and close to the anterior superior iliac spine (Fig. 4A).

The procedure was performed through the transperitoneal approach with gaining access to the retroperitoneum after the reflection of the colon cranially from the Gerota's fascia and caudally as low as the pelvic rim (Fig. 2A). Initially, the ureter and gonadal vein were identified lying lateral to the mass and were dissected and left up to avoid injury. Then the gonadal vein was dissected up to its origin from the renal

vein (Fig. 2B). The inferior mesenteric artery was identified crossing the mass, then was dissected, preserved, and reflected medially (Fig. 2C). A feeding artery was controlled at the medial aspect. The mass was dissected caudally and a draining vein was dissected and divided (Fig. 2D).

The cranial end of the aorta was dissected and the left sympathetic chain was identified behind the aorta laterally. Then the main feeding artery entering the mass superiorly was identified, dissected, and ligated (Fig. 3A). Posterior dissection of the mass continues and the sympathetic chain was preserved on the left side of the aorta posteriorly (Fig. 3B). Then dissection of the mass from the surface of the aorta was performed using sharp and blunt dissection until the main vein (from a lumbar vein) was identified and controlled followed by control of two main feeding arteries inferiorly and directly from the surface of the aorta (Fig. 3C, D). The mass was extracted using an endobag through and extended port site (Fig. 4B).

The procedure went uneventful, the operating time was 4.5 hours with minimal blood loss. During the procedure, three spells of hypertension (200/120) were recorded and necessitated the stoppage of the procedure until blood pressure was managed by the administration of direct vasodilators. Immediately after the control of the two draining veins, hypotension ensued and the patient received vascular replenishment by intravenous fluids to maintain blood pressure. The patient stayed in the intensive care unit for 24 hours for close

FIG. 2. Surgical procedure is demonstrated. (A) Reflection of the colon was required to expose the tumor. Dissection reached the renal hilum with the adrenal vein drains into the renal vein. (B) Dissection of the ureter and gonadal vein from the lateral surface of the mass. (C) The inferior mesenteric artery (*red*) is seen attached to the medial aspect of the tumor. (D) A draining vein (*blue*) was dissected and controlled inferior to the tumor.

FIG. 3. (A) Dissection cranial to the tumor and identification of a feeding artery. (B) Posterior dissection of the mass with identification of the sympathetic chain (medial) and the ureter (lateral). (C) Posterior dissection and control of the main draining vein (*blue*) behind the aorta. (D) A main feeding artery from the aorta is controlled, an inferior artery also is controlled with clips (seen inferiorly on the aorta).

monitoring of the vital parameters. Afterward, the patient was referred to the ward while under medical supervision, anti-hypertensive medications were withdrawn, and the patient fully recovered on the second day after surgery. The length of stay was 3 days and the postoperative course was uneventful.

Histopathologic examination confirmed the presence of paraganglioma with no malignant features. At 1 year follow-up, oncology wise, the patient was cured with no evidence of disease recurrence. Nevertheless, the patient developed anejaculation and is under current management in the andrology unit.

FIG. 4. (A) Port distribution with the proposed site of the tumor is determined by a circle. (B) The tumor after extraction. (C, D) The tumor bed after excision.

Discussion

Extra-adrenal paragangliomas arising from the OZ are not only rare but are also seldom encountered in the general urologic practice. Its overall incidence is 1–2:100,000 and the usual treatment of choice is open surgical removal.² Laparoscopic excision can be challenging and requires expertise that is anchored on the lengthy experience of various extirpative and reconstructive minimally invasive surgery.

Apart from the early convalescence and return to daily activities provided by laparoscopy, we believe that the approach has several merits with regard to the surgical technique. Initially, the laparoscope can be used for evaluating the extent and respectability of the tumor before making a decision of open surgery. In addition, the magnification provided enables adequate observation and identification and control of smaller blood vessels. Moreover, it is feasible in the pediatric population.³

In our case, the colon is reflected medially to get exposure of the aortic bifurcation area. This approach could minimize the risk of inferior mesenteric vessels injury as well as provide a wider exposure. Ragavan et al. have published the first case done robotically and utilized the same approach.¹ Arsanjani and associates gained access to an interaortocaval tumor through the colon reflection and duodenum Kocherization.³ However, the transmesocolic approach is feasible and could be utilized to gain direct access to the aortic bifurcation area without mobilizing the colon. It is to be noted that the inferior mesenteric artery or even a bowel segment might be resected to allow a complete tumor excision.² Cozzupoli and colleagues reported the resection of the inferior mesenteric artery to be able to dissect a 10 cm tumor.⁴

One of the surgical challenges to consider is that early control of the draining vein is not as easy as in cases of pheochromocytoma. The constant anatomy of the adrenal vein makes it a target in the early beginning of the procedure. Nonetheless, in the case of paragangliomas, meticulous dissection is required as usually the vein is not apparent during preoperative imaging or is located dorsally close to the vertebral column, which requires extensive dissection before control. Furthermore, the surgeon should expect multiple variable feeding arteries.¹

Intraoperatively, the anesthetic team should be aware of two major hemodynamic changes that might occur: namely hypertension episodes during tumor manipulation and hypotension after controlling the draining vein as α -blocking action is then unopposed. Postoperatively, the patient developed anejaculation that is a well-known consequence of retroperitoneal surgery. This was anticipated preoperatively and he underwent sperm banking. The rare location of the tumor at the proposed site of the hypogastric plexus might make the oncologic outcomes questionable if nerve preservation was attempted. Nevertheless, careful attention to any nerve fibers to be preserved might have avoided this complication.

Conclusion

Laparoscopic excision of paraganglioma of the OZ is feasible and safe as documented in this report. Colonic reflection or transmesocolic approaches are utilized to gain access to the mass, control of multiple feeding vessels might be required, and sacrifice of inferior mesenteric artery might be needed. Surgeon experience and adequate perioperative management of hemodynamic changes are of utmost

importance to optimize the outcome. Ejaculatory dysfunction should be highlighted before surgery.

Disclosure Statement

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References

1. Ragavan N, Ramesh M, Kandasamy N, Reddy YV. Robot-assisted laparoscopic excision of organ of Zuckerkandl. *J Robot Surg* 2016;10:373–374.
2. Thapar PM, Dalvi AN, Kamble RS, Vijaykumar V, Shah NS, Menon PS. Laparoscopic transmesocolic excision of paraganglioma in the organ of Zuckerkandl. *J Laparoendosc Adv Surg Tech A* 2006;16:620–622.
3. Arsanjani AH, Chang MF, Franco I. Laparoscopic transperitoneal resection of extra-adrenal pheochromocytoma of the organ of Zuckerkandl in a pediatric patient. *J Urol* 2008; 179(4S):303.
4. Cozzupoli P, Veneziano D, Sicuro O, Carbone L, Sgrò E. VID-04.08 laparoscopic resection of a large functional

paraganglioma in the organ of Zuckerkandl (PZ). *Urology* 2011;78:S168.

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Abbreviations Used

CT = computed tomography
OZ = organ of Zuckerkandl

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