

## Retroperitoneoscopic Resection of a Primary Retroperitoneal Paraganglioma Occurring between the Aorta and Vena Cava

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Received: 26 May 2022

Accepted: 07 Jun 2022

Published: 13 Jun 2022

J Short Name: AJSCCR

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### Citation:

Nishikimi T. Retroperitoneoscopic Resection of a Primary Retroperitoneal Paraganglioma Occurring between the Aorta and Vena Cava. *Ame J Surg Clin Case Rep.* 2022; 4(18): 1-5

### Keywords:

Paraganglioma; Retroperitoneal neoplasm; Retroperitoneal laparoscopic surgery

## 1. Abstract

**1.1. Introduction:** Paraganglioma is a rare extra-adrenal pheochromocytoma that arises from paraganglionic cells of the autonomic nervous system and can occur at various sites. Herein, we present a case of retroperitoneoscopic resection of a primary retroperitoneal paraganglioma between the aorta and vena cava.

**1.2. Case presentation:** A 67-year-old man presented with lower limb weakness, palpitations, and insomnia. His urinary total metanephrine and normetanephrine levels were high. Computed tomography showed a 48×34-mm oval mass on the dorsal side of the pancreas and portal vein between the aorta and vena cava. Because of the tumor location, we performed laparoscopic surgery using a retroperitoneal approach.

**1.3. Conclusion:** We described a case of a patient with primary retroperitoneal paraganglioma between the aorta and vena cava who underwent retroperitoneoscopic resection. Our case suggests that this procedure can be performed safely.

## 2. Introduction

Paraganglioma is a rare extra-adrenal pheochromocytoma that arises from paraganglionic cells of the autonomic nervous system and can occur at various sites. Although it most frequently appears in the large vessels, retroperitoneoscopic resection has rarely been reported. Herein, we present a case of retroperitoneoscopic resection of a primary retroperitoneal paraganglioma between the aorta and the vena cava.

## 3. Case Presentation

**3.1. Patient:** A 67-year-old man.

**3.2. Chief complaints:** Weakness of the lower limbs following alcohol consumption, insomnia, and palpitations.

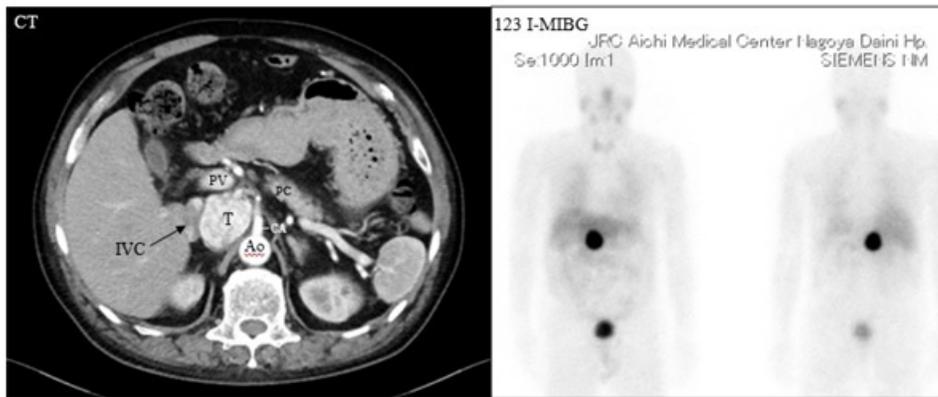
**3.3. Medical history:** Diabetes mellitus and hypertension.

**3.4. History of present illness:** The patient noted weakness of the lower limbs after drinking alcohol and was brought to our hospital via an ambulance. At the initial examination, his systolic blood pressure was low at 70–79 mmHg, but it promptly improved to 110–119 mmHg following fluid replacement. He was admitted for detailed examination.

Presentation status on admission: height, 165 cm; weight, 77 kg; body mass index, 28.28; body temperature, 36.0°C; blood pressure, 73/53 mmHg; pulse rate, 122/min.

**3.5. Hematologic test findings:** The patient's blood catecholamine levels were as follows: adrenaline, 0.02 ng/mL (upper limit: 0.03 ng/mL); noradrenaline, 1.63 ng/mL↑ (reference value: 0.10–0.50 ng/mL).

**3.6. Urinalysis findings:** The patient's urinary catecholamine levels were as follows: adrenaline, 8.8 µg/day (reference value: 3.0–41.0 µg/day); noradrenaline, 796.1 µg/day↑↑ (reference value: 31.0–160.0 µg/day); dopamine, 677.1 µg/day (reference value: 365–961.5 µg/day); urinary total metanephrine, 3.39 mg/day↑↑ (reference value: 0.14–0.46 mg/day); and urinary normetanephrine, 3.26 mg/day↑↑ (reference value: 0.10–0.28 mg/day). Imaging findings (Figure 1) suggested paraganglioma.

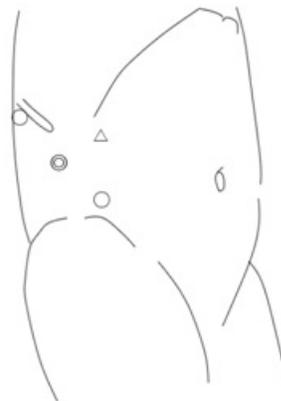


**Figure 1:** Imaging findings

Abdominal contrast-enhanced computed tomography (CT) A 48×34 mm oval-shaped mass lesion(T) was detected on the dorsal side of the pancreas(PC) and portal vein(PV) between the inferior vena cava (IVC) and celiac artery(CA). (Ao:aorta) 123-iodine metaiodobenzylguanidine (123 I-MIBG) scintigraphy, Abnormal radioisotope uptake was observed on the dorsal side of the pancreas between the IVC and celiac artery in the early phase that persisted in the late phase.

**3.7. Treatment course:** A catecholamine-producing primary retroperitoneal tumor between the aorta and vena cava was suspected based on the aforementioned findings. We planned preoperative treatment with oral administration of an  $\alpha$ -blocker (doxazosin) and  $\beta$ -blocker (bisoprolol fumarate), followed by surgical resection. Due to the tumor's location, we selected laparoscopic surgery using a retroperitoneal approach (Figure 2). Histopathological

findings appeared to be consistent with those of paragangliomas (Figure 3). Postoperative course: The patient started ambulation and oral intake the day after surgery and was discharged from the hospital on postoperative day 8. The palpitations detected before surgery disappeared. The catecholamine levels measured 1 week after surgery decreased as follows: blood catecholamines: nor-adrenaline, 0.31 ng/mL; urinary catecholamines: total urinary metanephrine, 0.11 mg/day; urinary normetanephrine, 0.04 mg/day.



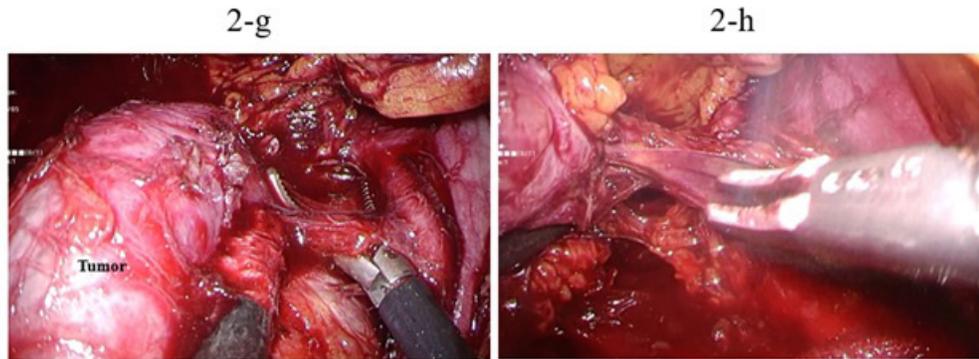
**Figure 2:** Site and size of trocars

⊙: Camera port (12 mm).

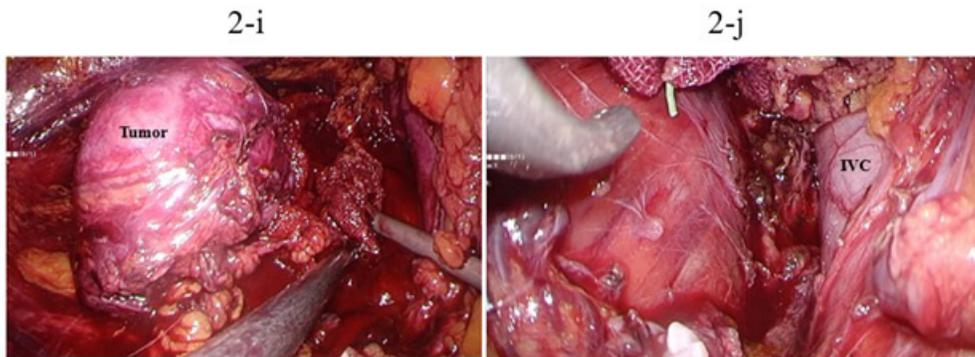
○: Ports for the operator's hands (12 mm).

△: Port for the assistant (5 mm).

The 12-mm port for the operator's left hand was placed at the intersection between the back muscles (lumbar quadratus muscle) and 12th rib. The camera port (12 mm) was placed between the 12th rib and iliac crest (almost on the axillary midline) at a site 3–4 fingers' width away from this port. Furthermore, the port for the operator's right hand was placed at a site approximately 3–4 fingers' width away from the camera port, and the 5-mm port for the assistant was placed at the cranial side of the port for the operator's right hand.

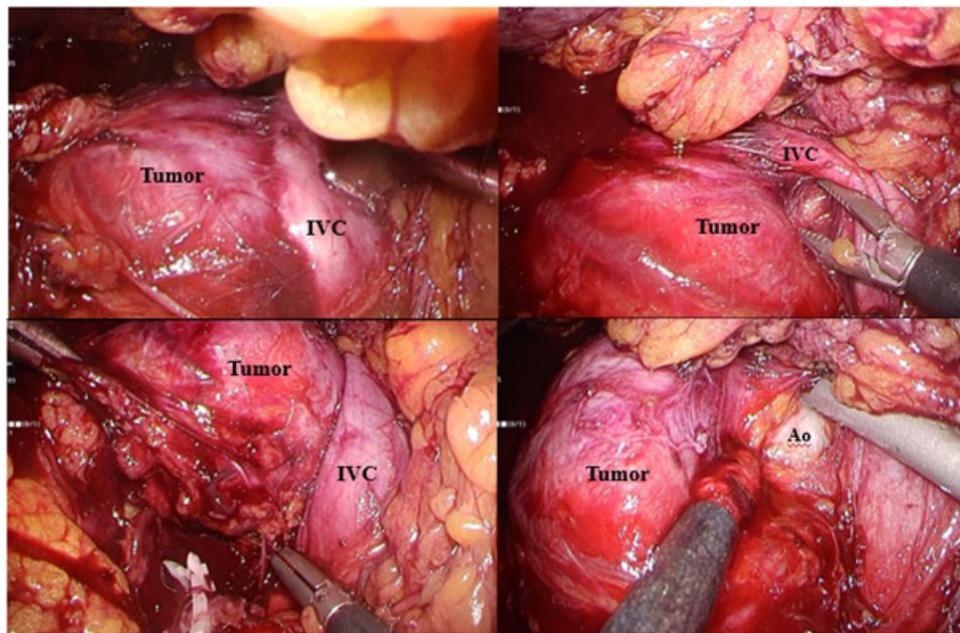


**Figure 2-g, h:** When the medial side of the tumor was dissected to some extent, forceps were inserted on the medial side. The tumor was pulled laterally and ventrally to create a column-like surgical field. A bundle of tissues containing feeding vessels was gradually dissected using a sealing device.



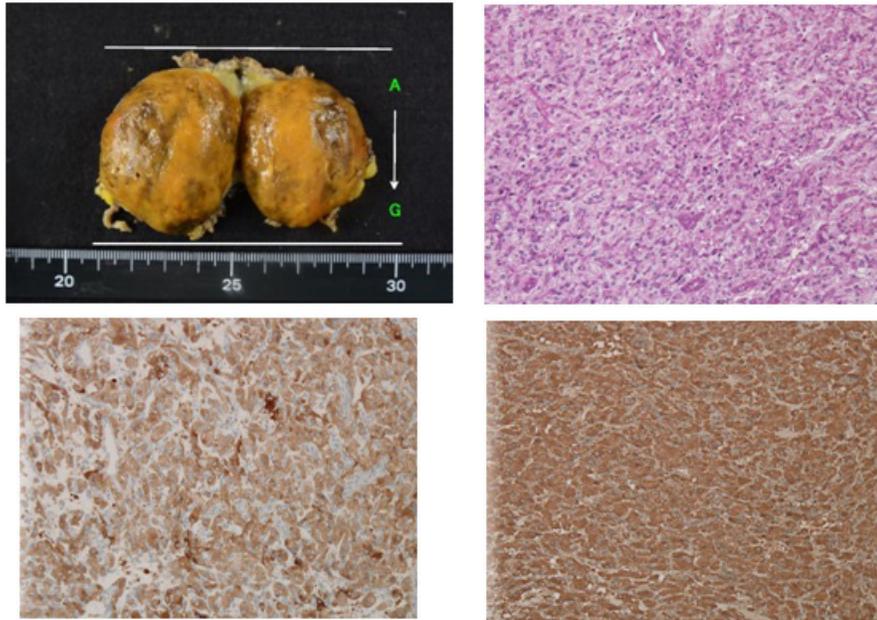
**Figure 2-i:** The tumor was removed.

Figure 2-j: After the absence of major bleeding at the site of resection and wound closure were confirmed. (Total operation time: 3hr 52min, Blood loss: 320ml.)



**Figure 3:** Surgical findings

Similar to nephrectomy or adrenalectomy performed via the retroperitoneal approach, the lateral conical fascia was incised to expose the surgical field and the right adrenal gland and right kidney were dissected. Consequently, a tumor was identified. As we dissected further, the IVC adjacent to the tumor was identified. The tumor was carefully dissected from the IVC using dissection forceps. When the IVC and the tumor were partially dissected, the tissue around the tumor was grasped. After the anesthesiologist confirmed the absence of marked changes in blood pressure, the tumor was pulled laterally to a non-damaging extent and dissected further. The aortic wall was also identified



**Figure 3:** Histopathological findings

Macroscopically, the tumor measured 5×4cm, was yellow-brown in color, and had a distinct border (Figure 3-a). Histologically, tumor cells with a large cytoplasm that were stained as slightly eosinophilic granules during hematoxylin-eosin staining (Figure 3-b) proliferated in an alveolar form with an abundant capillary network (Zellballen pattern). There were no signs of necrosis, increased cell density, or vascular invasion. Both synaptophysin staining (Figure 3-c) and chromogranin-A staining (Figure 3-d), which are highly specific to paraganglioma, were positive in the alveolar area. These findings appeared to be consistent with those of paragangliomas.

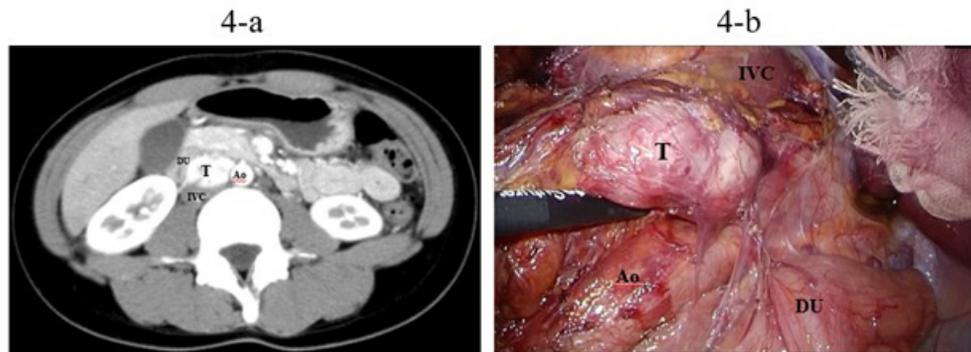
#### 4. Discussion

Catecholamine-producing tumors that occur outside the adrenal gland are referred to as paragangliomas [1]. Thus, primary retroperitoneal paragangliomas are relatively rare [2]. Although paragangliomas can occur anywhere in the sympathetic ganglia, the common sites of occurrence are the head and neck region, as well as the retroperitoneum around the aorta [3]. Measuring catecholamines and their metabolites is useful for diagnosis, and CT and magnetic resonance imaging are useful for localization [4]. MIBG scintigraphy, a radioisotope examination, has a high sensitivity and specificity (90% and almost 100%, respectively) [5]. Surgical resection is considered the first-line treatment strategy for both pheochromocytomas and paragangliomas. However, the indications for laparoscopic surgery have expanded to include select cases of retroperitoneal paraganglioma in recent years because of rapid advances in the procedure. Laparoscopic surgery may be a standard treatment for tumors with a diameter of  $\leq 7$  cm [6], but no clear indication has been established. There is a risk of damage to large vessels since paragangliomas often occur in contact with large vessels, as seen in our case. Thus, laparoscopic surgery for paraganglioma is considered more difficult to perform than that for other diseases. Given the lack of clear criteria for the indications for the procedure, we consider that the indications should be determined carefully. Since the procedure necessitates strict intraoperative control of blood pressure in addition to preoperative treatment with  $\alpha$ -blockers and other drugs, close cooperation with endocrinology and anesthesiology departments is also essential.

In the present case, because the tumor was adjacent to the pancreas and portal vein, as well as large vessels such as the IVC, aorta, and celiac artery, the magnification effect of laparoscopy was expected to be useful to secure the view of the retroperitoneum and identifying the dissection plane between the tumor and large vessels. Therefore, a laparoscopic surgery was performed. After consulting with the Department of Gastroenterological Surgery, we decided to first dissect the anterior and posterior surfaces of the tumor, which was located on the dorsal side of the IVC, as much as possible through the retroperitoneal approach rather than the transperitoneal approach from the beginning. If resection proves difficult, we planned to switch from the retroperitoneal approach to the transperitoneal approach to continue laparoscopic surgery, while keeping an option for conversion to laparotomy. In our case, the tumor was located on the dorsal side of the IVC, which is difficult to reach in open surgery using the transperitoneal or translumbar approach. However, the tumor was identified relatively easily when the right adrenal gland and right kidney were dissected to create a space using the retroperitoneal approach. Identifying the dissection plane between the tumor and large vessels under laparoscopic magnification is useful for safe tumor resection. Xu et al [7]. also reported the efficacy and safety of the retroperitoneal approach. Meanwhile, we have also experienced resection of paraganglioma by laparoscopic surgery using the transperitoneal approach at our department (Figure 4). Since CT revealed a tumor on the ventral side of the IVC (Figure 4-a), the transperitoneal approach was selected. As with nephrectomy and adrenalectomy

using the conventional transperitoneal approach, displacing the duodenum allowed us to directly observe the tumor between the IVC and aorta, and resect the tumor in a good visual field (Figure 4-b). Although no established approach for laparoscopic resection of paraganglioma currently exists, we believe that the approach should be comprehensively determined based on the tumor location, association with adjacent organs, and the surgeons' skill. Conversion to laparotomy due to difficult dissection from the large and renal vessels has also been reported [8,9]. For tumors adjacent to large vessels, such as the IVC and aorta, we believe that sur-

gery should be performed with vascular surgeons standing by in case the large vessels are damaged. We described a patient with a primary retroperitoneal paraganglioma occurring between the aorta and vena cava who underwent retroperitoneoscopic resection. Although reports on laparoscopic resection of paragangliomas are limited, this procedure is considered difficult to perform as it is associated with the risk of damaging large vessels. However, our case suggests that this procedure can be performed safely. Laparoscopic surgery exerts a magnification effect if the indications were carefully determined.



**Figure 4:** Another case of paraganglioma on the ventral side of the IVC

We have also experienced resection of paraganglioma by laparoscopic surgery using the transperitoneal approach at our department. Since CT revealed a tumor on the ventral side of the IVC (Figure 4-a), the transperitoneal approach was selected. As with nephrectomy and adrenalectomy using the conventional transperitoneal approach, displacing the duodenum (DU) allowed us to directly observe the tumor (T) between the IVC and aorta (Ao), and resect the tumor in a good visual field (Figure 4-b).

## 5. Conflicts of Interest

The authors declare no conflicts of interest. In consideration of ethical aspects, the contents of this article comply with the provisions of the ethics committee of the Japanese Red Cross Aichi Medical Center Nagoya Daini Hospital.

## 6. Author Contributions

All authors read and approved the final manuscript.

## 7. Ethical Approval Statement

The protocol for this research project has been approved by a suitably constituted Ethics Committee of the institution. Informed consent was obtained from the subjects.

The registration No.001219.

## 8. Data Availability Statement

Data sharing is not applicable to this article as no new data were created or analyzed in this study.

## 9. Supporting Information

Additional supporting information may be found in the online version of the article at the publisher's website.

## References

1. The Japanese Urological Association, the Japanese Society of Pathology, Japan Radiological Society, et al., eds. (2015) General Rule for Clinical and Pathological Studies on Adrenal Tumors, 3rd ed. Kanahara and Co., Ltd. Tokyo.
2. Whalen RK, Althausen AF, Daniels GH. Extraadrenal pheochromocytoma. *J Urol.* 1992; 147: 1-10.
3. Erickson D, Kudva YC, Ebersold MJ, Thompson GB, Grant CS, van Heerden JA, et al. Benign paragangliomas: Clinical presentation and treatment outcomes in 236 patients. *J Clin Endocrinol Metab.* 200; 86: 5210-6.
4. Moulton JS, Moulton JS. CT of the adrenal glands. *Semin Roentgenol.* 1988; 23: 288-303.
5. Nakajo M, Fujiyoshi F, Tsuchimochi S, et al. Pheochromocytoma and its metastasis. *Jpn J Imag Diagn.* 2002; 22: 167-73.
6. Soulie M, Mouly P, Caron P, Seguin P, Vazzoler N, Escourrou G, et al. Retroperitoneal laparoscopic adrenalectomy: Clinical experience in 52 procedures. *Urology.* 2000; 56: 921-5.
7. Xu W, Li H, Ji Z, Yan W, Zhang Y, Zhang X, et al: Retroperitoneal laparoscopic management of paraganglioma: a single institute experience. *Plos One.* 2016; 11: e014943.
8. Walz MK, Alesina PF, Wenger FA, Koch JA, Neumann HP, Petersenn S, et al. Laparoscopic and retroperitoneoscopic treatment of pheochromocytomas and retroperitoneal paragangliomas : Results of 161 tumors in 126 patients. *World J Surg.* 2006; 30: 899-908.
9. Hwang J, Shoaf G, Uchio EM, Watson J, Pacak K, Linehan WM, et al. Laparoscopic management of extraadrenal pheochromocytoma. *J Urol.* 2004; 171: 72-76.