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Surgical Management of Recurrent Retroperitoneal Paraganglioma: Anatomical Challenges in Surgical Dissection

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Paragangliomas (PGL) are rare neuroendocrine tumors that arise from sympathetic or parasympathetic ganglia of the autonomic nervous system (1). PGLs of sympathetic origin are predominantly located in the retroperitoneum, particularly at the aortic bifurcation and the organ of Zuckerkandl, and are associated with high catecholamine secretion (1,2). Approximately one-third of PGL are hereditary; succinate dehydrogenase complex iron sulfur subunit B (SDHB) mutations and other germline mutations are associated with recurrence and metastasis (3). Since a histopathological distinction between benign and malignant lesions cannot be made, metastasis is the main indicator of malignancy (3). The main approach for curative treatment is complete surgical resection. However, in cases of recurrence with retroperitoneal involvement, surgical dissection can be challenging because of dense fibrosis, anatomical distortion, and proximity to major vessels (1).

This letter discusses the surgical strategy and anatomical challenges encountered in managing a recurrent retroperitoneal PGL, with reference to current literature.

A 21-year-old woman presented three years ago with persistent vomiting, headache, and hypertension. Abdominal imaging revealed a 6×5-cm mass in the left preaortic area adjacent to the celiac trunk, pushing the pancreas anteriorly. The mass, which was evaluated for pheochromocytoma and PGL spectrum, was completely excised; pathology revealed PGL, a Ki-67 proliferation index of 5%, vascular-lymphatic-capsular invasion, and a heterozygous SDHB mutation. The patient, who was hormonally inactive in the postoperative period, was followed annually with radiological and biochemical assessments.

On follow-up magnetic resonance imaging (MRI) and Ga-68 DOTATATE positron emission tomography-computed tomography (CT), a recurrent lesion measuring 2-2.5 cm and exhibiting intense receptor uptake was observed posterior to the pancreas and anterior to the aorta. Preoperative imaging demonstrated that the mass was in close proximity to the superior mesenteric artery, splenic artery, and renal artery and vein (Figure 1). Accordingly, patients were routinely



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informed about the potential risk of vascular injury and the need for additional surgical procedures, and written informed consent was obtained prior to surgery. For 1 week prior to surgery, the patient underwent preoperative fluid replacement for volume expansion at a rate of 1,000 mL/day with a crystalloid-to-colloid ratio of 1:1.

A subcostal incision was made, the gastrocolic ligament was released, and a partial Mattox maneuver was performed. The bilobed recurrent mass extending beneath the pancreas into the retroperitoneal space caused severe difficulty during dissection because of dense perivascular fibrosis resulting from prior surgery and significant anatomical distortion surrounding the superior mesenteric artery and the renal hilum. The patient's blood pressure remained stable throughout the intraoperative tumor excision, and the mass was excised en bloc without vascular injury. No postoperative complications developed, and the patient was discharged on the seventh postoperative day.

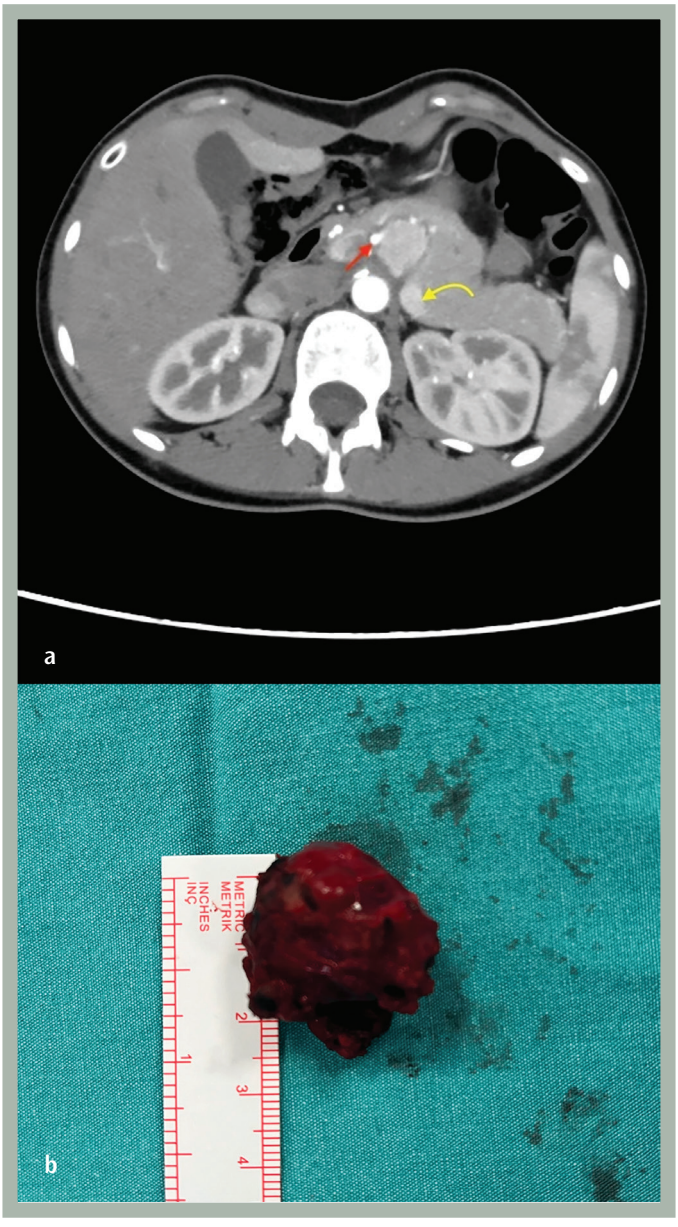


Figure 1. (a) Axial slice from a triphasic computed tomography scan. Red arrow: splenic artery; yellow arrow: splenic vein. (b) Macroscopic specimen

Histopathological examination revealed two foci of PGL measuring 3 cm and 1.5 cm, respectively. Mitotic activity was low (1 mitosis/2 mm^2); no atypical mitoses, vascular invasion, or lymphovascular invasion were observed, and surgical margins were reported as negative. Since the surgical margins were negative and the patient remained asymptomatic, no adjuvant therapy was administered and follow-up was planned with clinical and biochemical evaluations every 6 months and cross-sectional imaging (CT or MRI) annually during the first five years. At the 6-month postoperative follow-up, the patient remains disease-free.

Surgery for recurrent retroperitoneal PGL is a high-risk procedure due to anatomic proximity and fibrotic scarring. In addition, since recurrence rates are higher in patients with SDHB mutations, long-term, close follow-up is recommended (2). Visceral rotation techniques (e.g., the Mattox maneuver) increase the likelihood of successful resection by facilitating safe exposure of major vascular structures (4). In addition, ensuring intraoperative hemodynamic stability during PGL surgery requires close coordination between the surgical and anesthesia teams, and is critical for operative safety (1).

In conclusion, retroperitoneal PGLs are rare tumors whose surgical management requires advanced expertise because of their anatomical location. Successful surgical treatment in recurrent cases is possible with detailed preoperative planning, appropriate hemodynamic preparation, precise dissection to preserve vascular structures, and multidisciplinary teamwork. Long-term radiological and biochemical follow-up is of great importance, especially in patients with an SDHB mutation.

Footnotes

Informed Consent: The written informed consent was obtained prior to surgery.

Authorship Contributions

Surgical and Medical Practices: F.G., Y.Y.K., N.A., E.K., Concept: F.G., Y.Y.K., N.A., E.K., Design: F.G., Y.Y.K., N.A., E.K., Data Collection or Processing: F.G., Y.Y.K., N.A., E.K., Analysis or Interpretation: F.G., Y.Y.K., N.A., E.K., Literature Search: F.G., Y.Y.K., N.A., E.K., Writing: F.G., Y.Y.K., N.A., E.K.

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