Laparoscopic Resection of Retrocaval Non-functioning Paraganglioma

Moon Soo Lee, M.D.¹, Yoon Jung Kang, M.D.¹, Hyun-Young Han, M.D.², Hyun-Jin Son, M.D.³, Jae Min Lee, M.D.⁴

Departments of ¹Surgery, ²Radiology, ³Pathology, ⁴Internal Medicine, Eulji University Hospital, Daejeon, Korea

INTRODUCTION

Paragangliomas are extra-adrenal chromaffin tumors that develop at the expense of the neuroectodermal cells of the autonomous nervous system. Retroperitoneal and non-functioning forms are very rare.¹ Laparoscopic adrenalectomy has been considered surgery for benign and partially malignant tumors.² However, laparoscopic retrocaval paraganglioma resection is technically challenging due to its close relation to the vena cava. Accordingly, the laparotomy has been preferred. However, in our case, laparoscopic resection of the retrocaval paraganglioma was performed successfully. To our knowledge, this is the first report of the laparoscopic resection of a retrocaval non-functioning paraganglioma.

Key words: Paraganglioma, Non-functioning, Retrocaval, Laparoscopic

CASE REPORT

A 49-year-old male was referred for an asymptomatic retroperitoneal tumor, detected by FDG-PET. He underwent right thyroidectomy for papillary carcinoma one year ago. Abdominal computed tomography (CT) scan showed a mass measuring 2 cm in size located behind the inferior vena cava (IVC), which deviate IVC anteriorly. A ¹²³I-metaidoiodobenzylguanidine (MIBG) scan also demonstrated abnormal focal activity. Result of preoperative adrenal function test was normal. A laparoscopic retrocaval tumor excision was performed successfully. The operative time was 160 minutes. The patient's postoperative recovery was uneventful and he was discharged on the fourth postoperative day. Pathologic examination was consistent with the diagnosis of paraganglioma. To the best of our knowledge, this is the first report on laparoscopic resection of a retrocaval non-functioning paraganglioma. In conclusion, laparoscopic retrocaval paraganglioma resection is safe and feasible.

Corresponding author: Moon Soo Lee
Departments of Surgery, Eulji University Hospital, 1306, Dunsan-dong, Seo-gu, Daejeon 302-799, Korea
Tel : +82-42-259-1330, Fax : +82-42-259-1125
E-mail : mslee01@eulji.ac.kr

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Fig. 1. (A) FDG-PET showing a mildly hypermetabolic, low density mass at retroperitoneum (retrocaval area). (B) Abdominal CT showing a 2 cm round highly enhancing hypervascular mass with central necrosis in the retrocaval space, which deviated from IVC anteriorly without definitive evidence of vessel-wall invasion. (C, D) I-\textsuperscript{123}MIBG scan showing abnormal focal activity in right upper quadrant area.

Fig. 2. Trocar site placements for the laparoscopic retrocaval tumor resection.

and medial to lower pole of the adrenal gland. The IVC was displaced and compressed anterio-medially by the tumor. After the localization of the tumor, the border between the anterior surface of the tumor and the posterior surface of the IVC was carefully dissected using a blunt dissector and a harmonic scalpel (Ethicon, Cincinnati, Ohio) (Fig. 3A). The tumor was progressively extracted from behind, on the right side of the vena cava. No direct invasion of the IVC was observed. Thereafter, we proceeded with dissection of the posterior attachment of the tumor. And the feeding vessel for the tumor was ligated and divided. Finally, the paraganglioma was divided from the paravertebral ganglia (Fig. 3B). The operative time was 160 minutes. During mobilization of the tumor, no elevations of the patient’s blood pressure were observed. These procedures were successfully conducted without vascular injury and without conversion to open surgery. The patient’s postoperative recov-
Fig. 3. (A) Laparoscopic view at the localization of retrocaval paraganglioma. (B) View after laparoscopic paraganglioma resection. L = liver; A = adrenal gland; T = tumor; G = paravertebral ganglia; IVC = inferior vena cava.

Fig. 4. Surgical specimen showed the grayish soft tissue, measuring 2.3×2.0 cm-sized mass.

ery was uneventful and he was discharged on the 4th postoperative day. The surgical specimen showed 2.3×2.0 cm-sized, well-demarcated, soft mass (Fig. 4). Histological examination coupled with an immunohistochemical study was consistent with the diagnosis of paraganglioma with Ki 67 < 1% (Fig. 5). SDHB and SDHD exon mutation analyses were performed for exclusion of hereditary paraganglioma syndrome, because the patient suffered from papillary carcinoma and paraganglioma. DNA blood analysis revealed no SDHB and SDHD exon mutation.

DISCUSSION

According to the World Health Organization in 2004, paraganglioma is described as intra-adrenal paraganglioma, and is classified as pheochromocytoma and extra-adrenal paraganglioma. Paragangliomas is usually a functional tumor which secretes catecholamine or their metabolites. Functioning paraganglioma can be diagnosed from typical clinical symptoms such as palpitation and flushing face. However, a non-functional paraganglioma is very rare and the diagnosis may be a significant challenge because it is characterized by its asymptomatic profile and normal levels of catecholamines in the urine and blood. Therefore, a non-functional paraganglioma is usually discovered after the tumor has become sufficiently enlarged to produce symptoms related to the compression on adjacent organs. Also, non-functioning paraganglioma tumors tend to be difficult to resect due to their size and location. Computerized tomography has been established as the investigation method of choice for demonstrating the location, extent, and relation to surrounding tissues of a retroperitoneal tumor. Occasionally, as in our case, I-123MIGB scan or PET-CT may be alternative diagnostic image modalities.

It may not be possible to differentiate benign from malignant paraganglioma tumors, even at the time of diagnosis. Therefore, complete surgical excision is considered as the treatment of choice. Laparoscopic adrenalectomy has become the standard approach for resection of benign adrenal diseases, including the pheochromocytoma. Also, open surgical procedures have been used to remove most extra-adrenal paragangliomas because they are mainly located at the para-aortic or para-caval area and are thus difficult to approach. However, recent improvements in minimally invasive surgery have made laparoscopic resection of paragangliomas possible. Tetsuo Nozaki et al. reported the first case of the laparoscopic resection of a retrocaval paraganglioma, demonstrating its feasibility, despite the high level of compression of the IVC. Also, Saud Alrasheedi...
reported that a robotic approach as well as laparoscopic for retrocaval paraganglioma resection is feasible and safe. Most resection of laparoscopic or robotic retrocaval paraganglioma is confined to the functioning tumor. To our knowledge, this is the first case where a non-functioning paraganglioma located at the retrocaval area has been resected successfully laparoscopically.

The main risk of the laparoscopic approach for retrocaval tumor excision is injury and hemorrhage of the IVC during the tumor resection. The atraumatic grasper and 5 mm Endo peanut™ (auto-suture) were useful for blunt dissection between the retrocaval tumor and the IVC. Most connective tissue and the feeding vessels were controlled using a harmonic scalpel and clips. In our case, no vessel invasion was observed due to the relatively small-size of the (2 cm) tumor. On the other hand, a more precise localization was needed for the small size tumor to avoid unnecessary dissection. This could be achievable using an anatomical landmark through the multimodality images at the preoperative preperation. If vessel-wall invasion is observed or the tumor is too large, laparoscopic resection of the paraganglioma should be considered inappropriate and the procedure should be converted to open surgery.

In conclusion, retroperitoneal retrocaval non-functioning paraganglioma is rare and may be difficult to diagnose. In addition, open surgical procedures were used to remove most extra-adrenal paraganglioma due to the difficulty in approaching the location. However, a laparoscopic resection of a retrocaval paraganglioma can be performed safely through proper patient selection and preoperative planning.
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