

Right Thoracoabdominal Approach for Retrocardiac Paraganglioma Resection

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Paragangliomas are rare extra-adrenal tumors of sympathetic or parasympathetic paraganglia origin; of these, mediastinal paragangliomas are 2% of all cases. We present the case of a 21-year-old woman with uncontrolled arterial hypertension who had a functioning 6.5 × 6.2-cm retrocardiac paraganglioma firmly attached to the pericardium. The patient underwent tumor resection via a right thoracoabdominal incision; this surgical approach enabled adequate exposure for complete resection without institution of cardiopulmonary bypass or need for cardiac reconstruction or autotransplantation. Ten months postoperatively, the patient was doing well and was no longer hypertensive. (Tex Heart Inst J 2017;44(1):62-5)

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Paragangliomas are rare extra-adrenal tumors of sympathetic or parasympathetic paraganglia origin.¹ Mediastinal paragangliomas, slow-growing neoplasms that comprise nearly 2% of all paragangliomas, can be divided on the basis of their location: tumors in the anterior mediastinum (arising from parasympathetic paraganglia) or those in the posterior mediastinum (arising from sympathetic chain).²⁻⁴ Clinical manifestations are varied, and these depend on the tumor size, the compression of adjacent anatomic structures, and the secretion of excessive amounts of catecholamines. Although these tumors can be a cause of arterial hypertension, they account for only 0.01% of all such cases.⁵ We present the case of a young woman whose functioning retrocardiac paraganglioma we excised through a right thoracoabdominal incision.

Case Report

A 21-year-old woman was referred to our institution from a community medical facility. She had a 6-year history of progressive and uncontrolled arterial hypertension despite medical therapy with prazosin (2 mg/d) and propranolol (100 mg/d). A full investigation of possible causes included plasma-free metanephrines. The patient's normetanephrine value was highly elevated at 3,177 pg/mL (normal level, <145 pg/mL); other laboratory values were within normal ranges. An abdominal computed tomographic (CT) scan showed no abnormality. Then a complete body scan—obtained through hybrid single-photon emission computed tomography and CT (SPECT-CT) after injection of ¹³¹I-metaiodobenzylguanidine (¹³¹I-MIBG)—showed no sites of abnormal uptake. Next, the patient underwent positron emission tomographic-CT (PET-CT) scanning after injection of fluorine-18-L-dihydroxyphenylalanine (¹⁸F-DOPA); this study revealed a 6.5 × 6.2-cm retrocardiac paraganglioma (Fig. 1A). Finally, a chest CT scan confirmed a vascularized mass in the posterior mediastinum, adjacent to the left atrium (Fig. 1B).

Cardiothoracic and vascular surgery services were consulted, and after discussions with the patient regarding the various surgical options, she was taken to the operating room. In a left lateral decubitus position, the patient underwent an initial video-assisted thoracoscopy (VATS) that did not enable resection of the mass. A right thoracotomy was then performed at the 7th intercostal space. Because of the large mass and the small amount of space, we extended the incision to the upper abdomen, thereby completing a thoracoabdominal incision. This approach enabled us to expose the posterior mediastinum and to view the mass directly. We mobilized the deflated right lung laterally, and we found a plane between the tumor and the surrounding structures, thereby enabling circumferential dissection of the mass, which—firmly adhered to the pericardium, as it was—required a limited pericardiectomy (Fig. 2).

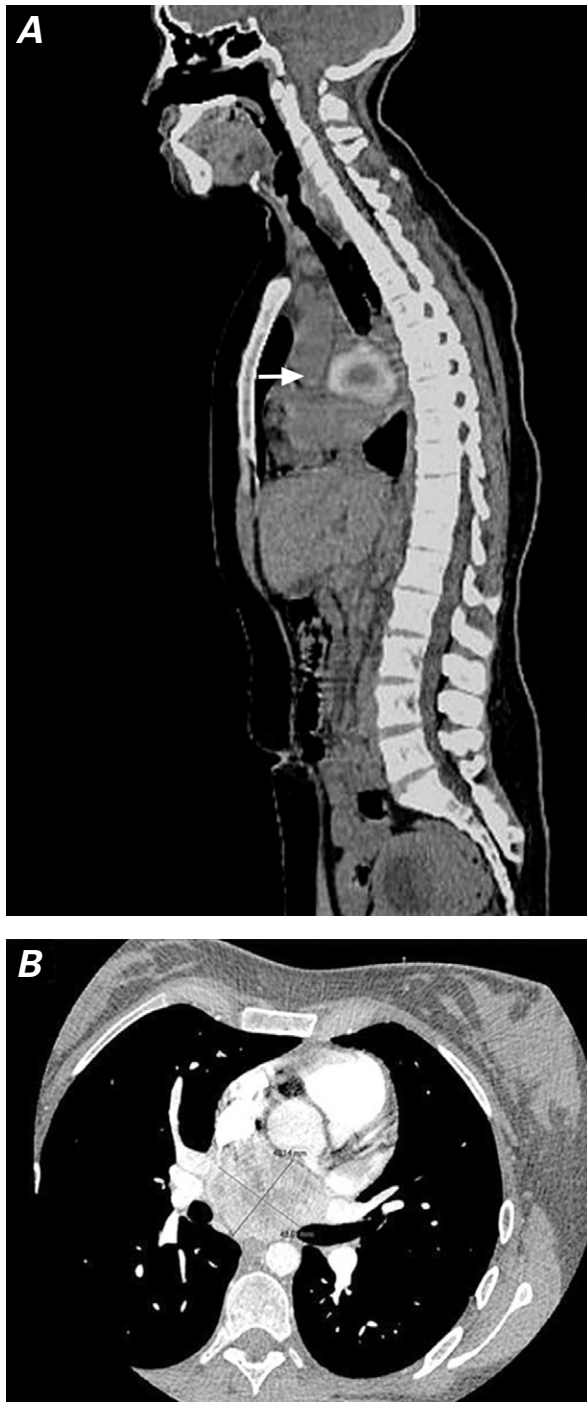


Fig. 1 **A)** Fluorine-18-L-dihydroxyphenylalanine positron emission tomogram–computed tomogram shows a retrocardiac paraganglioma (arrow). **B)** Chest computed tomogram confirms a 6.5 × 6.2-cm vascularized mass in the posterior mediastinum.

The mass was removed successfully, and the margins were tumor-free (Fig. 3A). The patient tolerated the procedure well, her recovery was uncomplicated, and she was discharged from the hospital on postoperative day 10. The histologic features of the mass were compatible with paraganglioma (Fig. 3B). Ten months postopera-

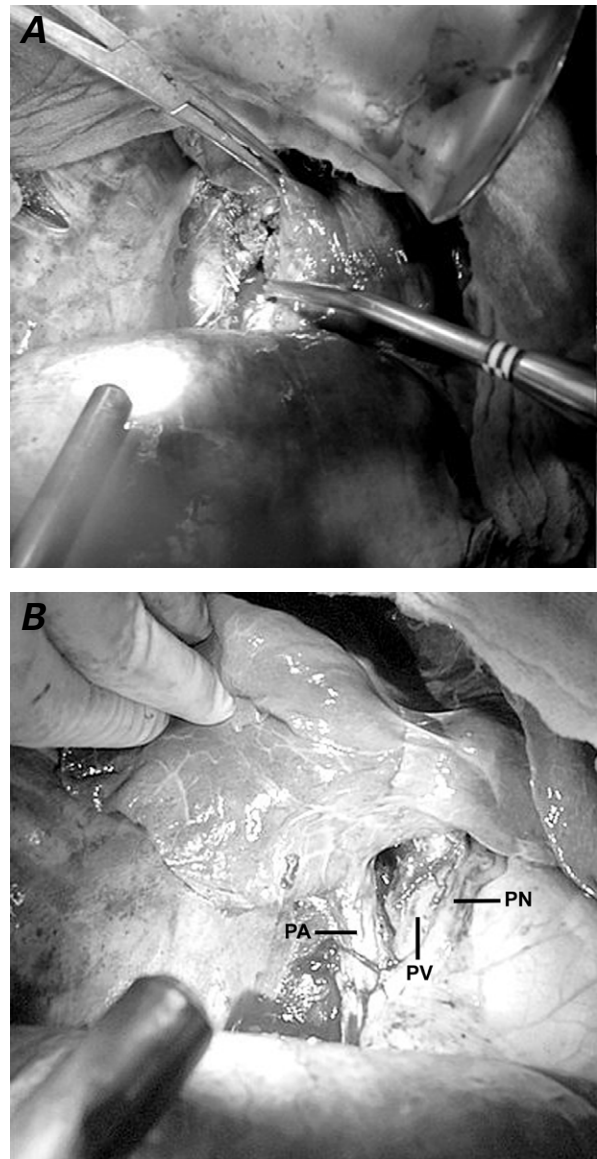


Fig. 2 Intraoperative photographs. **A)** The deflated right lung was mobilized laterally, and a plane was found between the tumor and the surrounding structures, enabling circumferential dissection of the mass via a limited pericardiectomy. **B)** The mass was successfully removed without major intraoperative bleeding.

PA = pulmonary artery; PN = phrenic nerve; PV = pulmonary vein

tively, the patient was doing well; her serum normetanephrine level was 61 pg/mL, and she no longer needed antihypertensive therapy.

Discussion

Paragangliomas are slow-growing tumors arising from the autonomic nervous system; mediastinal paragangliomas comprise only 1% of all mediastinal tumors and less than 2% of all pheochromocytomas.⁶ As in

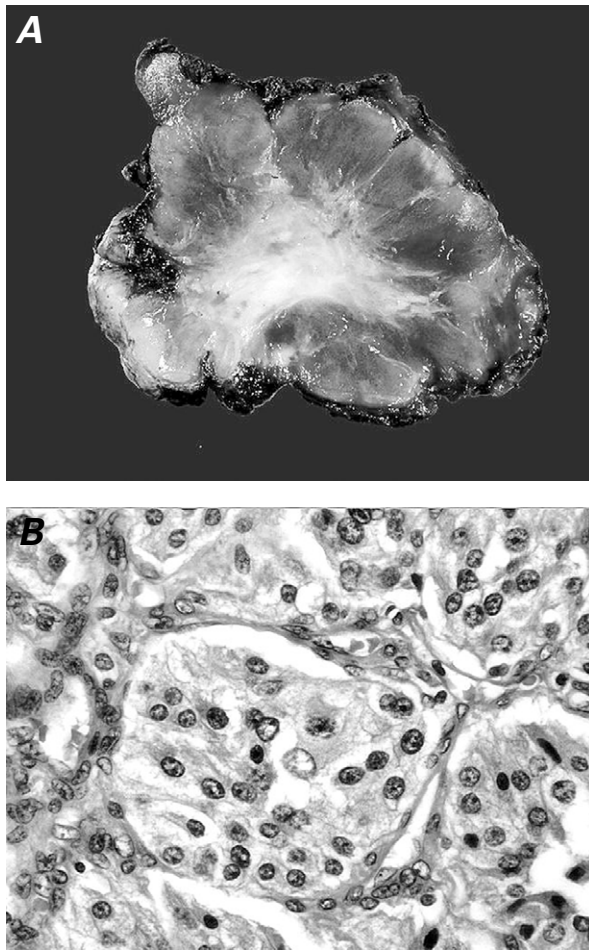


Fig. 3 **A)** Macroscopically, the resected tumor was well-demarcated, highly vascular, and removed with free margins. **B)** Cytologic examination revealed the Zellballen pattern of paraganglioma (H & E, orig. $\times 40$).

our patient's case, the anterior mediastinal location is more common than is the posterior. Of paragangliomas in the posterior mediastinum, 50% are functional, and appropriate antihypertensive therapy may be instituted (an α -adrenergic blocker is recommended 7 to 10 days preoperatively, followed by β -blockers or calcium channel blockade as adjuncts when blood pressure or tachycardia are not controlled⁷). The ^{123}I -MIBG scintigraphy and fluorodeoxyglucose PET scans have proved useful in locating and staging this entity: the more functional the tumor, the higher the MIBG accumulation.⁴

Pericardial or cardiac paragangliomas are very rare. Standard therapy consists of complete surgical resection, because these tumors are resistant to chemotherapy and radiotherapy.^{4,8} The surgical approaches that are usually described include median sternotomy under cardiopulmonary bypass (CPB) or circulatory arrest, necessitating dissection and division of the major vessel, or reconstruction of the heart itself.^{4,8,9} The extreme vascularity and locations of these tumors mandate me-

ticulous surgical planning and might necessitate complex surgical procedures for success.⁸ In our patient, we observed that the tumor had not invaded surrounding structures, which precluded the necessity of CPB to ensure safe dissection. The latter might be indicated if invasion of the heart, coronary arteries, or another great vessel is seen.

The largest reported surgical experience, from the Mayo Clinic, included 14 patients with mediastinal paragangliomas, only 6 of whom needed CPB for resection.⁷ The authors reported positive-margin resections in 3 of 14 patients and, during follow-up, 2 recurrences in patients with initially negative margins. There was one intraoperative death because of massive hemorrhage. Among 11 patients at the University of Michigan, there were 2 postoperative deaths.⁹ In 2012, 4 of 7 patients underwent successful resection with the heart autotransplantation technique⁸; the authors consider the use of this alternative when the tumors involve the pulmonary veins or large portions of the atrium.⁸ A mortality rate of 14% was reported in that series.

In 2014, Suzawa and colleagues³ reported the complete surgical resection by VATS of a 2.2-cm asymptomatic paraganglioma, a minimally invasive approach that could be a feasible option for small tumors.³ Although our initial approach was by means of VATS, no safe pulmonary vein dissection plane was identified, and conversion to open surgery became necessary. First, we performed a right thoracotomy, but because of the tumor's size and the small operative space, we decided to extend the incision to the abdominal region for adequate exposure and complete resection of the paraganglioma.

While performing a posterolateral thoracotomy for an intrapericardial paraganglioma resection, Yamamoto and colleagues¹⁰ encountered substantial hemorrhage, leading to cardiac arrest as they dissected the tumor from the left atrial wall. Although they were able to repair the wall laceration, these authors suggested, on the basis of this experience, that CPB should be considered if sites for arterial and venous cannulation can be secured while a right posterolateral thoracotomy is performed.¹⁰

During our dissection, the tumor was found to be closely attached to the pericardium, necessitating limited pericardiectomy. Under this circumstance, the use of the HARMONIC[®] vessel-sealing system (Ethicon US, LLC, a Johnson & Johnson company; Cincinnati, Ohio) and careful finger dissection can enable optimal pulmonary vein vascular control. Preoperative embolization is in some cases attempted in order to reduce major intraoperative bleeding from these hypervascular tumors that adhere to adjacent structures and are close to great vessels.⁸ After careful evaluation of the imaging studies, we opted not to perform that procedure for fear of an uncontrollable hypertensive crisis through cat-

echolamine release (caused by post-embolization tumor necrosis). Complete surgical removal was achieved in our patient without sequelae.

The expected survival rates for retrocardiac paraganglioma range from 85% to 95% with appropriate surgical care, but incomplete removal considerably reduces the survival rate—to 50%, even when adjuvant therapies are instituted.⁴ Close monitoring is mandatory.

In our patient, a right thoracoabdominal incision enabled optimal exposure of the tumor for complete resection, without need of CPB, hypothermic circulatory arrest, heart reconstruction, or autotransplantation.

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