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CASE REPORT

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Successful thoracoscopic-assisted resection of a functional paraganglioma in the mediastinum with extracorporeal circulation: a case report

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Abstract

Background Paragangliomas are rare neoplasms arising from extra-adrenal chromaffin cells, with mediastinal paragangliomas representing an exceptionally rare subset. This report details the surgical management of a complex mediastinal paraganglioma case, presenting with refractory hypertension and invasion of critical surrounding structures. A comprehensive review of the current literature is included to underscore existing cases, enhance clinical awareness, and share our insights and experience in the diagnosis and treatment of this challenging condition.

Case presentation A 16-year-old female presented with recurrent headaches and persistent hypertension lasting over one year. Based on clinical findings and imaging studies, she was preliminarily diagnosed with a mediastinal paraganglioma. The patient underwent comprehensive preoperative management, including oral α- and β-adrenergic blockade, preoperative arterial embolization, and intravenous fluid volume expansion, to optimize endocrine control. Thoracoscopic resection of the mediastinal mass was initially attempted; however, the procedure became complex due to the high risk of uncontrolled hemorrhage and invasion of adjacent vital structures. Following the preoperative surgical plan, the incision was converted to a lateral thoracotomy, and cardiopulmonary bypass was initiated. Meticulous dissection enabled the complete removal of the tumor along with the affected posterior wall of the left atrium, followed by reconstruction of the left atrium and the right pulmonary vein. The surgery was successfully completed, and follow-up assessments showed no signs of tumor recurrence or metastasis.

Conclusions Functional mediastinal paraganglioma is a rare neuroendocrine tumor, with complete surgical resection being the gold standard treatment. Stringent perioperative management is crucial to mitigate the risk of cardiovascular complications associated with functional tumors. Lifelong surveillance is recommended to detect potential recurrence or metastasis. Effective collaboration within a multidisciplinary team is essential for ensuring accurate diagnosis and delivering optimal, individualized care.

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Keywords Mediastinal paraganglioma, Perioperative management, Surgery, Extracorporeal circulation, Multidisciplinary treatment

Introduction

Paragangliomas are rare tumors derived from extra-adrenal chromaffin cells, typically located in the head, neck, and abdomen [1, 2]. However, only about 2% arise in the mediastinum [3, 4], making mediastinal paragangliomas exceptionally uncommon. While most paragangliomas in the head and neck are non-functional, nearly half of those below the neck are associated with excessive catecholamine secretion [3]. Hypertension is the most frequent symptom, accompanied by headaches, palpitations, sweating, and cardiovascular manifestations [3]. Complete surgical resection remains the primary treatment for paragangliomas. However, functional paragangliomas demand strict perioperative management to mitigate the risks of intraoperative hypertensive crises, massive hemorrhage, and postoperative hypotension [1, 3]. In this report, we present a case of mediastinal paraganglioma with refractory hypertension and invasion of the left atrium and pulmonary vein. With the coordinated efforts of our multidisciplinary team (MDT), the patient received comprehensive perioperative management and underwent successful surgical resection, resulting in a favorable prognosis.

Case presentation

A 16-year-old female presented with a history of persistent hypertension and recurrent headaches for over one year, with occasional blood pressure spikes reaching 210/100 mmHg. She reported symptoms of headaches, excessive sweating, palpitations, nausea, and vomiting. Despite multiple visits to local hospitals, no definitive diagnosis was made, and she had not been on consistent medication. Her symptoms progressively worsened over time, leading to her referral to our hospital for further evaluation and management. Upon admission, a comprehensive diagnostic workup was initiated.

Laboratory tests ruled out Cushing's syndrome and primary aldosteronism, as the patient displayed a normal circadian cortisol rhythm (CCR) and the aldosterone-to-renin ratios (ARR) below 30 (Fig. 1). Biochemical analysis of urine samples showed elevated vanillylmandelic acid (VMA), while plasma norepinephrine and normetanephrine levels exceeded the upper reference limit by four-fold (Table 1), strongly indicating pheochromocytoma or paraganglioma.

Computed tomography (CT) imaging identified a 5.3×3.3×3.5 cm mediastinal soft tissue mass above the right pulmonary artery, below the pulmonary vein, anterior to the left atrium, and posterior to the spine (Fig. 2A). The unenhanced CT showed a density of 30 Hounsfield units (HU), increasing to 90 HU with contrast, suggesting heterogeneous enhancement. The mass compressed the left atrium and displaced the pulmonary vein inferiorly. Magnetic resonance imaging (MRI) revealed isointense to slightly hypointense T1 signals and slightly to highly hyperintense T2 signals (Fig. 2C). A cardiac ultrasound confirmed a mildly hyperechoic mass near the posterior left atrial wall and the right pulmonary vein entrance, with well-defined margins.

The MDT diagnosed the patient with a functional mediastinal paraganglioma and recommended surgical resection under controlled hypotension. To optimize preoperative blood pressure and heart rate control, the patient was administered α-adrenergic blockade (phenoxybenzamine), β-adrenergic blockade (propranolol), and calcium channel blockade (nifedipine sustained-release). Intravenous fluid expansion was also implemented to prevent severe hypotension following tumor removal [1]. To reduce intraoperative bleeding, preoperative embolization of the tumor's feeding vessels was performed [5]. Given the tumor's proximity to the heart, the surgical plan included preparations for potential cardiopulmonary bypass (CPB) and atrial reconstruction, with a preoperative consultation and coordination with the cardiac surgery team.

Following preoperative optimization, the patient underwent thoracoscopic resection of the mediastinal mass under general anesthesia. The patient was positioned in the right lateral decubitus position, with thoracoscopic ports placed at the fourth and seventh intercostal spaces. Intraoperatively, a firm, irregular mass measuring approximately 5.0×4.0×4.0 cm was identified. The tumor exhibited an incomplete capsule with poorly defined margins. Even minimal manipulation triggered marked increases in heart rate and blood pressure. Further exploration revealed tumor invasion of the pericardium, left atrium, and partial involvement of the pulmonary vein. In consultation with the cardiac surgery team, it was determined that resection would require CPB support, along with reconstruction of the left atrium and pulmonary vein. The incision was converted to a lateral thoracotomy. The bilateral femoral artery and vein cannulation was performed to establish femoral-femoral bypass, enabling precise resection of the tumor, including the affected posterior wall of the left atrium. After complete excision, the left atrium and right pulmonary vein were reconstructed using bovine pericardial patches. The procedure was completed successfully, and postoperative pathology confirmed the diagnosis of mediastinal paraganglioma (Fig. 2D-F).

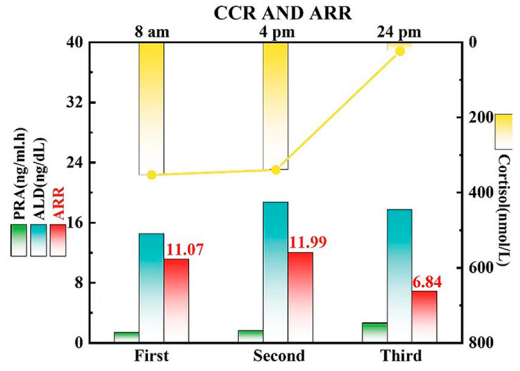

Fig. 1 Laboratory test results before the surgery. The patient had a normal CCR and all ARR were less than 30. (CCR: circadian cortisol rhythm, ARR: aldosterone-to-renin ratios)

Table 1 Laboratory test results before the surgery

Subject	First	Second
Urine VMA(≤ 60.24umol/day)	145.5	157.95
Dopamine(≤ 195.7pmol/L)	162.9	354.4
Adrenaline(≤ 605.4pmol/L)	65.5	91.4
Norepinephrine(414-4435.5pmol/L)	19005.7	35,732
3-Methoxytyramine(<0.18nmol/L)	0.10	0.34
Metanephrine(≤ 0.50nmol/L)	0.09	0.22
Methoxy-norepinephrine(≤ 0.90nmol/L)	> 20.56	18.86

(VMA: vanillylmandelic acid)

On the second postoperative day, the patient developed transient cardiac dysfunction, with increased cardiac volume load triggering pulmonary edema and impaired circulation. This complication was likely attributed to a sharp decline in catecholamine levels following tumor resection, leading to diminished myocardial contractility and reduced stroke volume. With intensive cardiac and respiratory support, alongside diuretics, vasodilators, and appropriate anti-infective therapy, the patient's condition steadily improved. Follow-up laboratory tests and imaging at two weeks post-surgery confirmed favorable recovery. The patient was discharged in stable condition on postoperative day 23. No tumor recurrence or metastasis was found during more than two years of outpatient follow-up (Fig. 2B). The patient's blood pressure normalized without needing medication, and she successfully returned to her normal daily activities.

Literature review

We identified 19 cases of surgically treated mediastinal paragangliomas published in PubMed (Table 2). Among these, 11 patients were female and 8 were male, ranging from 24 to 76 years (median age: 52 years). The tumors were distributed across various mediastinum regions: 6 in the middle, 11 in the posterior, and 2 in the anterior mediastinum. Clinical presentations were diverse, with

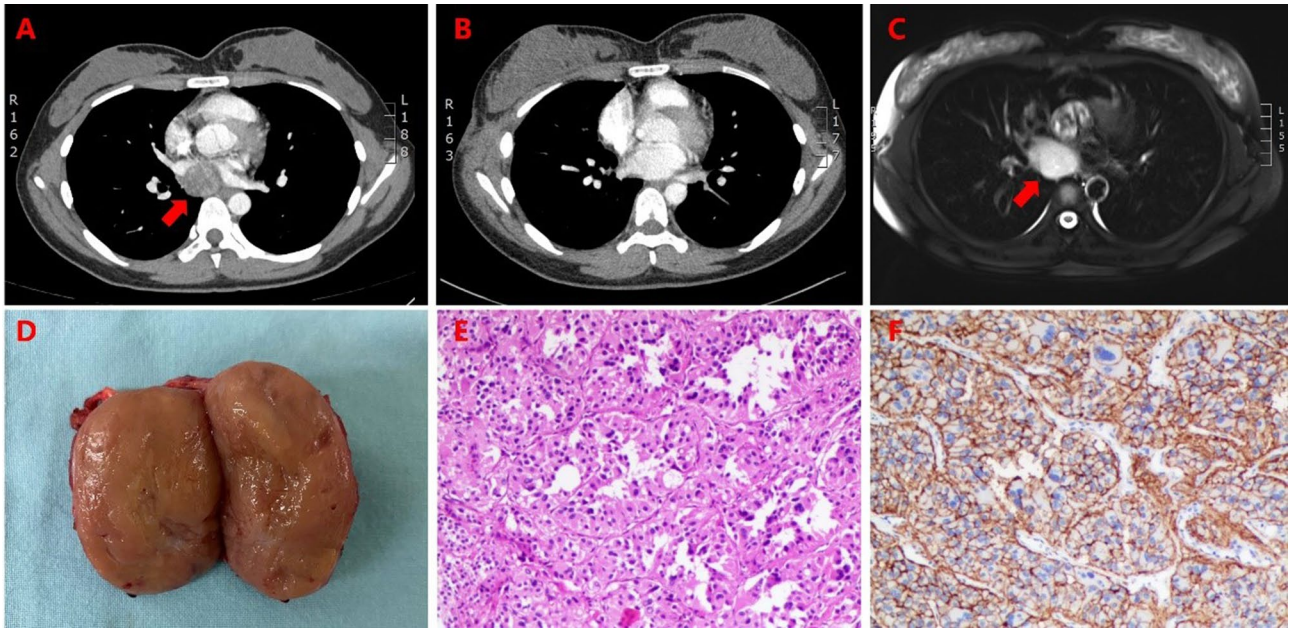

Fig. 2 Imaging and pathological results of the patient. **A**) The chest CT with contrast shows a heterogeneous mass measuring 5.3 × 3.3 × 3.5 cm located just below the right pulmonary artery and above the pulmonary vein in the mediastinum. There is significant compression of the left atrium and downward displacement of the pulmonary vein. **B**) A follow-up contrast-enhanced CT at 6 months post-surgery reveals no signs of significant recurrence and no notable pleural or pericardial effusion. **C**) The MRI showed isointense to slightly hypointense T1 signals and slightly to highly hyperintense T2 signals in the immediate vicinity of the right upper pulmonary vein, right upper pulmonary artery, ascending aorta, esophagus, and left atrium. **D**) The gross specimen of the tumor measures 4.5 × 4.5 × 3 cm, with a grayish-yellow cross-section and a soft texture. **E**-**F**) Tumor cells predominantly exhibit a nested (Zellballen) and organoid arrangement, with focal necrosis and some involvement of myocardial tissue. Immunohistochemistry results: CD56 (+), Syn (+), CgA (+), S-100 (+), CK (-), Ki-67 (5% +). (CT: computed tomography, MRI: magnetic resonance imaging)

Table 2 Previous studies documented 19 cases of surgical treatment for mediastinal paragangliomas

No.	PMID	Patient	Tumor location	Surgery	CPB	Pathological diagnosis
1	24,570,537	52/F	Middle mediastinum (Invasion to the intrapericardial pulmonary artery)	Median sternotomy	Yes	Non-functional paraganglioma
2	31,363,483	47/M	Left posterior mediastinum (Adjoining the T7-T8 vertebral body)	Left thoracotomy	No	Functional paraganglioma
3	36,107,546	31/M	Middle mediastinum (Enclosed the proximal LAD)	Standard midline sternotomy	Yes	Non-functional paraganglioma
4	35,636,209	47/F	Posterior mediastinum (In the paravertebral gutter)	Posterolateral thoracotomy	No	Non-functional paraganglioma
5	33,708,339	58/F	Middle mediastinum (Epicardial mass)	Median sternotomy	Yes	Paraganglioma
6	32,384,867	65/M	Posterior mediastinum	VATS	No	Functional paraganglioma
7	31,043,977	24/M	Posterior mediastinum (Left para-aortic area)	VATS	No	Functional paraganglioma
8	32,138,719	31/F	Posterior mediastinum (Invading the left 2nd rib)	RATS + Chest wall resection	No	Non-functional paraganglioma
9	32,997,281	76/F	Middle mediastinum (Between the aorta and the left pulmonary artery)	VATS	No	Functional paraganglioma
10	31,305,440	47/M	Left posterior mediastinum (In the paravertebral groove at 7–8 intercostal space)	Left thoracotomy	No	Functional paraganglioma
11	35,256,545	53/F	Middle mediastinum (Contiguous with the aortic arch and LPA)	Left-sided posterolateral thoracotomy	No	Paraganglioma
12	28,427,002	44/F	Posterior mediastinum (abutting the aorta)	Left-sided posterolateral thoracotomy	No	Functional paraganglioma
13	22,707,541	54/F	Posterior mediastinum (Abutting the esophagus, aorta and diaphragm)	Left thoracolaparotomy	No	Functional paraganglioma
14	24,782,980	55/F	Anterior mediastinum (Under the left innominate vein, above the aortic arch)	VATS	No	Paraganglioma
15	35,668,526	73/M	Left posterior mediastinum (Adjacent to the descending aorta)	VATS	No	Functional paraganglioma
16	34,434,294	56/F	Anterior mediastinum	Sternotomy	Yes	Functional paraganglioma
17	32,953,448	60/F	Middle mediastinum	Sternotomy	No	Paraganglioma
18	25,589,992	48/M	Posterior mediastinum (Adjoining the T11-T12 vertebral body)	VATS	No	Functional paraganglioma
19	33,225,892	36/M	Posterior mediastinum	Lateral thoracotomy	No	Functional paraganglioma

(CPB: cardiopulmonary bypass, VATS: Video-assisted thoracoscopic surgery, RATS: Robot-assisted thoracoscopic surgery, LPA: Left pulmonary artery, LAD: Left anterior descending artery)

common symptoms including chest or back pain, chronic cough, hypertension, palpitations, headaches, dyspnea, nausea, vomiting, dysphagia, spinal pain, episodic lower limb paralysis, and, in some cases, diabetes.

Of the 19 cases, nine patients received preoperative management for endocrine symptoms. Strategies included oral α-blockers (Cases No. 7, 15), α-blockers with intravenous fluid resuscitation (No. 2, 10, 12), α- and β-blockers (No. 13, 16, 19), and combined α-/β-blockers with intravenous fluid resuscitation (No. 18). All patients underwent surgical resection. In 4 cases, intraoperative blood pressure surges occurred during tumor manipulation, managed with rapid-acting antihypertensives to ensure safe resection (No. 2, 6, 10, 13). CPB was used in 4 cases to minimize bleeding and resect tumors invading critical structures, such as coronary arteries or pulmonary arteries (No. 1, 3, 5, 16). Two patients with left coronary artery invasion required coronary artery bypass grafting (CABG) (No. 3, 5). Postoperative complications included left recurrent laryngeal nerve palsy and persistent hypotension (No. 9, 13). With intensive care, all patients recovered and were successfully discharged.

Discussion

The diagnosis and management of functional paragangliomas are particularly challenging due to their rarity and diverse clinical presentations. Optimal perioperative care and surgical planning rely on seamless collaboration within a multidisciplinary team involving endocrinologists, anesthesiologists, radiologists, and surgeons. This integrated approach ensures precise hemodynamic

control, tailored pharmacologic management, and successful surgical outcomes.

Precise monitoring and meticulous perioperative care are critical to mitigating complications and ensuring patient safety. Preoperative α-adrenergic blockade should commence 7 to 14 days before surgery, with phenoxybenzamine (Dibenzyline) being the most commonly used agent. The initial dose is typically 10 mg twice daily, titrated based on the patient's response. If tachyarrhythmias develop, β-adrenergic blockers can be introduced 2 to 3 days after initiating α-blockade. Standard options include atenolol (12.5–20 mg, 2–3 times daily), metoprolol (25–50 mg, 3–4 times daily), and propranolol (20–80 mg, 1–3 times daily) [6, 7]. A high-sodium diet and intravenous fluid expansion are recommended to counteract catecholamine-induced hypovolemia and prevent severe hypotension following tumor resection [1]. Additionally, as Camilla S. Carlsen et al. recommended, preoperative selective embolization of tumor vessels can effectively reduce intraoperative blood loss [5].

Resecting functional paragangliomas presents significant challenges due to the tumor's size, limited mediastinal space, hemodynamic instability, and invasion of vital structures. Key intraoperative priorities include controlling bleeding and minimizing damage to major organs. CPB offers critical support, enabling safe and complete tumor removal. Given the risk of severe cardiac compression and the potential need for structural reconstruction, the early involvement of an experienced cardiac surgery team is essential for comprehensive preoperative planning. Following complete resection, vigilant postoperative monitoring is crucial to ensure optimal recovery. Close surveillance facilitates the early detection of complications and allows for prompt intervention, promoting favorable patient outcomes.

Studies report that patients undergoing complete resection have higher survival rates and longer median survival than those receiving partial resection and adjuvant therapy [8]. Additionally, the 5-year recurrence rate for secretory paragangliomas can reach up to 20% [9]. Therefore, regular follow-up and lifelong monitoring are essential for postoperative management. A comprehensive follow-up plan should include periodic imaging, hormonal evaluations, and clinical assessments. This approach enables early detection and prompt management of recurrent or residual lesions while providing valuable insights into the patient's long-term prognosis.

Conclusions

Functional mediastinal paragangliomas are exceedingly rare in clinical practice. Their rarity underscores the necessity of thorough evaluation, meticulous perioperative management, and carefully planned surgical strategies to ensure patient outcomes.

Abbreviations

MDT	Multidisciplinary team
CCR	Circadian cortisol rhythm
ARR	Aldosterone-to-renin ratios
VMA	Vanillylmandelic acid
CT	Computed tomography
HU	Hounsfield units
MRI	Magnetic resonance imaging
CPB	Cardiopulmonary bypass
CABG	Coronary artery bypass grafting

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Author contributions
ZHY: Conceptualization, Data curation, Visualization and Writing – original draft. XJH: Conceptualization, Formal analysis and Writing – original draft. DTZ: Conceptualization, Data curation, and Writing – original draft. TT: Methodology and Resources. HXL: Methodology and Visualization. WLL: Project administration. FLY: Resources and Supervision. CC: Conceptualization, Project administration and Writing – review & editing.All the authors have read and approved the final version of the manuscript.

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Data availability
No datasets were generated or analysed during the current study.

Declarations

Ethics approval and consent to participate
The Institutional Review Board of the Second Xiangya Hospital approved the research protocol of this study. Written informed consent was obtained from all subjects.

Consent for publication
Informed consent was acquired from the patient, and the patient consented to the publishing of all images, clinical data, and other data included in the manuscript.

Competing interests
The authors declare no competing interests.

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