

# Asymptomatic left posterior mediastinal functional paraganglioma

## A case report

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### Abstract

**Rationale:** Paraganglioma refers to a set of neuroendocrine tumors derived from the chromaffin cells of the adrenal sympathetic ganglion. Paragangliomas can be classified as functional or nonfunctional based on the ability to synthesize and release catecholamines.

**Patient concerns:** We report a 47-year-old man with a functional paraganglioma in the left posterior mediastinum and highlight the key elements of management of mediastinal paragangliomas.

**Diagnoses:** A left posterior mediastinal mass was found by computed tomography (CT) scan and Chest-enhanced CT. Preoperative ultrasound-guided biopsy suggested the possibility of a paraganglioma. A diagnosis of paraganglioma was established by immunohistochemistry.

**Interventions:** The patient underwent single-stage resection of the lesion *via* left thoracotomy after preoperative oral  $\alpha$ -adrenoceptor (phenoxybenzamine) therapy and intravenous fluid resuscitation for two weeks.

**Outcomes:** The postoperative period was uneventful. The patient exhibited no abnormal blood pressure or recurrence during the 12-month follow-up period.

**Lessons subsections as per style:** Pathological examination alone cannot determine whether it was a benign or malignant paraganglioma, which can be determined by pathological examination combined with distant metastasis. Long-term follow-up is required to assess the treatment effect.

**Abbreviations:** CT = computed tomography, MRI = magnetic resonance imaging, PET-CT = positron emission tomography-computed tomography.

**Keywords:** hypertension, mediastinal tumor, paraganglioma, pheochromocytoma

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Written informed consent was obtained from the patient for publication of the case.

The authors declare that they have no conflict of interest.

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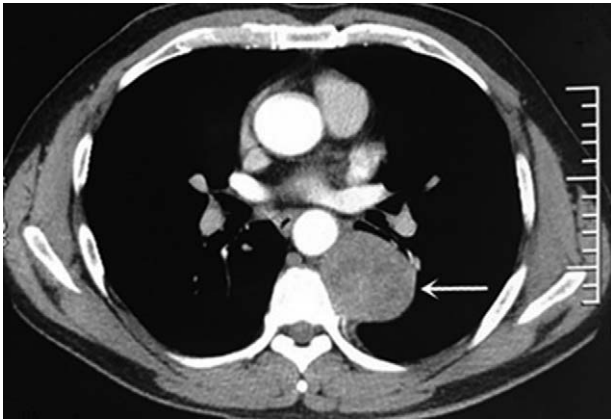
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## 1. Introduction

Paraganglioma refers to a set of neuroendocrine tumors derived from the chromaffin cells of the adrenal lateral ganglion. An estimated 90% of these tumors occur in the adrenal medulla, and are commonly known as pheochromocytoma. The remaining 10% derived from the adrenal gland are called paragangliomas.<sup>[1]</sup> Mediastinal paragangliomas are extremely rare entities that account for only 1%–2% of all paragangliomas and <0.3% of all mediastinal tumors.<sup>[2]</sup> Till date, approximately 150 cases have been reported in literature.<sup>[3]</sup> Paragangliomas can be classified as functional or non-functional based on the ability to synthesize and release catecholamines.<sup>[4]</sup> Mediastinal paragangliomas mainly occur in the anterior mediastinum and most are nonfunctional.<sup>[2]</sup> We report a 47-year-old man with a functional paraganglioma in the left posterior mediastinum and highlight the key elements of management of mediastinal paragangliomas.

## 2. Case report

This study was approved by the Ethics Committee of Chinese-Japan Union Hospital of Jilin University. All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki



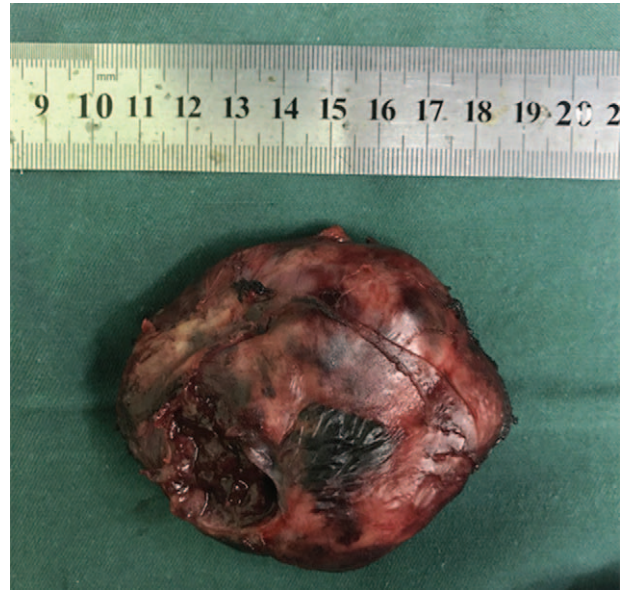
**Figure 1.** Chest enhanced CT showing a round, solid mass in the left posterior mediastinum with low-density cystic lesions in the central area (arrow).

declaration and its later amendments or comparable ethical standards. Written informed consent was obtained from the individual participant included in the study.

A 47-year-old man was admitted to our hospital due to intermittent tearing pain in the left side of the chest and back since >10 months. A left posterior mediastinal mass was found by computed tomography (CT) scan. At admission, his blood pressure was 120/80 mmHg, pulse rate was 82 beats/minute, respiratory rate was 18 beats/minute, and body temperature was 36.5°C. The patient had no history of hypertension, headache, palpitations, night sweats, weight loss, or facial flushing. Furthermore, there was no family history of hypertension. Chest-enhanced CT revealed a round, solid mass in the left posterior mediastinum, with low-density cystic lesions in the central area; there were no enlarged lymph nodes in the hilum or the mediastinum (Fig. 1). Preoperative ultrasound-guided biopsy suggested the possibility of a paraganglioma. No distant metastasis was observed by whole body positron emission tomography-computed tomography (PET-CT) scan. Plasma level of epinephrine was 83 pg/mL (0–100 Pg/mL), and that of norepinephrine and dopamine was 420 pg/mL (0–600 Pg/mL) and 82.6 pg/mL (0–100 Pg/mL), respectively. Urinary excretion of epinephrine was 3.22 μg/24 hours (0–20 μg/24 hours), and that of norepinephrine and dopamine was 67.8 μg/24 hours (0–90 μg/24 hours) and 130.5 μg/24 hours (0–600 μg/24 hours), respectively.

The patient underwent single-stage resection of the lesion *via* left thoracotomy after preoperative oral α-adrenoceptor (phenoxylbenzamine) therapy and intravenous fluid resuscitation for two weeks.

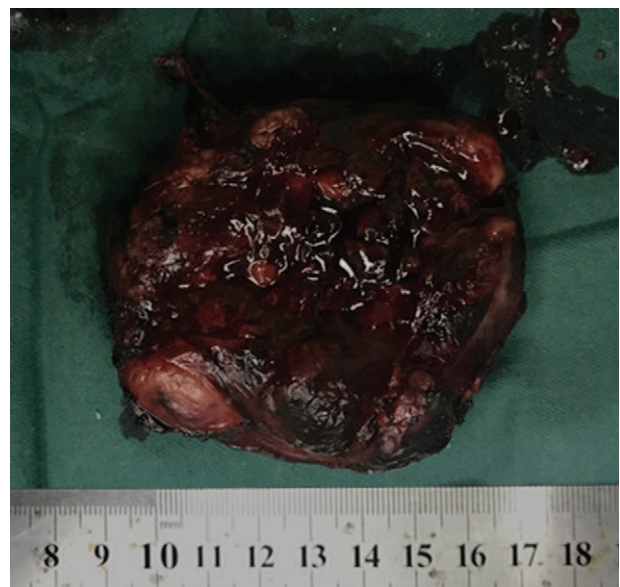
For the operation, after double-lumen endotracheal intubation anesthesia, the patient was placed in the right lateral position. The tumor was located in the paravertebral groove at the level of 7–8 intercostal space between the descending aorta and the sympathetic chain. The tumor was derived from the left sympathetic chain and measured approximately 7 × 6 × 4 cm in size. It was dark red in color with a clear border, abundant blood supply, and incomplete capsule. The posterior chest wall and descending aorta adventitia were compressed. Intraoperative manipulation with the tumor led to a sudden rise in blood pressure to 220/120 mmHg. Therefore, the patient was treated with rapid infusion of antihypertensive agent. After control of



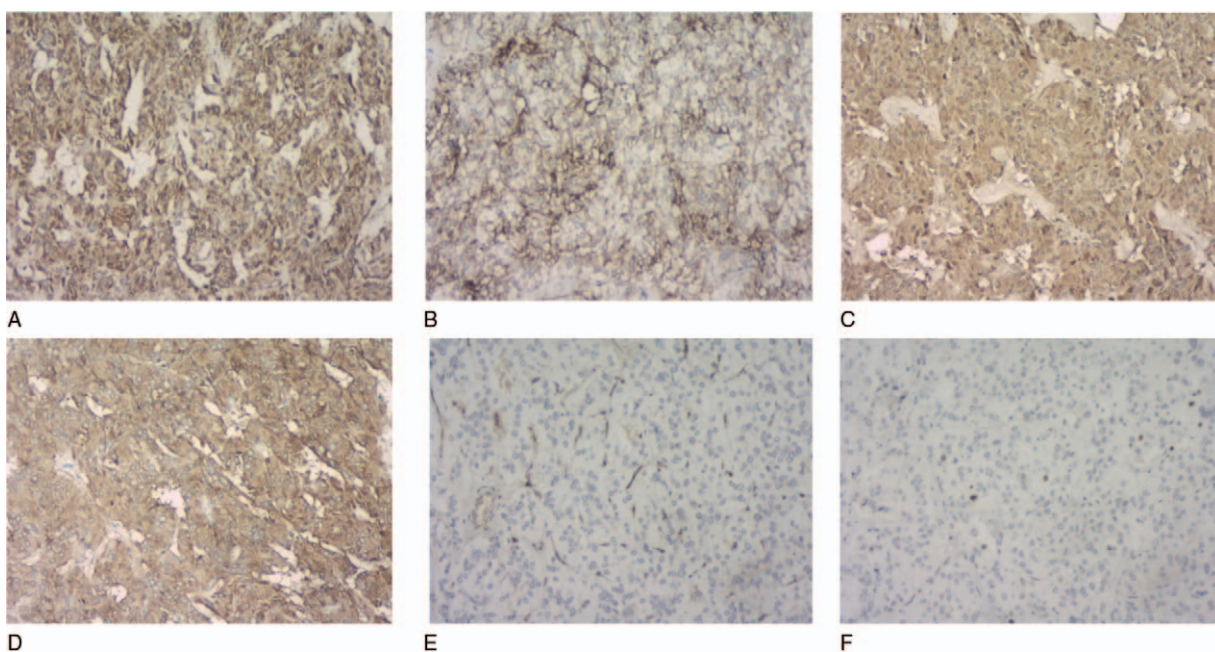
**Figure 2.** Intraoperative image showing a tumor with a clear border and incomplete membrane.

blood pressure, the tumor was quickly peeled off and removed (Fig. 2). Postoperatively, the blood pressure was maintained around <120/75 mmHg. Postoperatively, the plasma levels of epinephrine, norepinephrine, and dopamine were in the normal range. The patient exhibited no abnormal blood pressure or recurrence during the 12-month follow-up period.

Histopathological examination revealed a mass sized 6.5 × 6.0 × 4.0 cm with cystic changes in the central area (Fig. 3). Immunohistochemistry revealed CK(–), EMA(–), Vimentin(+), Inhibinα(–), CD34(–), S-100(–), CD56(+), CgA(+), SyN(+), and



**Figure 3.** The grayish part of the cut surface was soft with few capsule and cystic changes in the central area.



**Figure 4.** Immunohistochemistry revealed (A) Vimentin(+), (B) CD56(+), (C) CgA(+), (D) SyN(+), (E) CD34(vessel+), and (F) Ki-67 (<5%+) (100×).

Ki-67 (<5%+) (Fig. 4). Thus, a diagnosis of paraganglioma was established.

### 3. Discussion

Paraganglioma most frequently occurs in patients with an average age of 40–50 years old, with no predilection for a particular sex. However, only 3% of these tumors secrete catecholamines. In contrast, in almost half of all young patients with paravertebral paragangliomas, the tumors synthesize catecholamines.<sup>[5]</sup> Paragangliomas typically occur in areas with abundant parasympathetic nerves, such as the head, neck, mediastinum, adrenal glands, retroperitoneum, bladder, duodenum, and thyroid.<sup>[6]</sup> Mediastinal paragangliomas are mainly located in two areas, that is, in the aortic sinus sympathetic ganglion in the posterior mediastinum and the autonomic ganglion in the superior or middle mediastinum.<sup>[7]</sup> Paragangliomas are commonly found in the urinary system, but are rare in the thoracic cavity. Approximately 25%–70% of patients with extra-adrenal paragangliomas exhibit symptoms and signs of excessive catecholamine secretion;<sup>[8]</sup> these patients typically present with hypertension, facial flushing, palpitations, and night sweats. However, our patient exhibited no such symptoms, thereby belonging to normotensive pheochromocytoma.<sup>[9]</sup> Plasma biochemical marker (epinephrine, norepinephrine, and chromogranin A) tests are the initial laboratory investigations of choice in patients with suspected pheochromocytoma or paraganglioma.<sup>[10]</sup> CT and magnetic resonance imaging (MRI) are the preferred imaging modalities for the localization and qualitative diagnosis of paraganglioma. The lesions appear iso-dense or slightly hypodense on CT, and exhibit significant enhancement on contrast-enhanced CT. On MRI T1W1 sequence, the lesions exhibit an equal or low signal, while T2W1 sequence reveals a medium, high, or nonuniform mixed signal. Diffusion-weighted imaging shows a high signal. The lesions typically exhibit significant enhancement on enhanced MRI.<sup>[11]</sup> In addition to common neurogenic tumors,

such as schwannomas, rare ectopic tumors should also be considered in the differential diagnosis of masses located in the posterior mediastinum, in order to avoid misdiagnosis.

Surgical resection is the treatment of choice for paraganglioma. Thoracoscopic surgery affords adequate exposure of the operative field and reveals the fine structure of the lesions. Compared with traditional thoracic surgery, thoracoscopic surgery offers the advantage of lesser trauma and postoperative pain.<sup>[12]</sup> However, since the tumor had compressed the posterior chest wall and the adventitia of the descending aorta, we opted for open surgery owing to safety concerns; moreover, the safety and efficacy of thoracoscopic resection of left posterior mediastinal functional paraganglioma is not well characterized. Ma et al considered that although thoracoscopic surgery has been successfully applied in some cases, thoracotomy remains the best choice for tumors that have an abundant blood supply.<sup>[13]</sup> Perioperative management is also important, including adequate preoperative preparation, meticulous intraoperative monitoring, and strict postoperative care. Furthermore, since the preoperative pathological immunohistochemistry results were positive for some antibodies, the lesion was considered to be a paraganglioma with endocrine function. For this reason, the following measures were adopted: first, sufficient peripheral vasodilation before surgery by  $\alpha$ -adrenoceptor blocker therapy (oral phenoxybenzamine, 40 mg/day divided in three doses) for two weeks; second, blood volume expansion by low molecular dextran (500 mL/day for two weeks); third, preoperative blood preparation; fourth, real-time intraoperative monitoring of blood pressure; and fifth, close monitoring of blood pressure and heart rate after surgery, and maintenance of water and electrolyte balance.

An estimated 90% of all paragangliomas are benign. Furthermore, it is difficult to judge the characteristics based on the morphology alone.<sup>[14]</sup> The typical sign of malignant paraganglioma is metastasis. Hence, pathological examination

may not determine the feature. Long-term follow-up is needed to assess the treatment effect.

### Author contributions

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**Resources:** Yang Yue, Hua Xin, Fu-Qiang Li, Jun-Duo Wu, Le-Ning Zhang.

**Supervision:** Yang Yue, Le-Ning Zhang.

**Validation:** Yang Yue, Hua Xin, Fu-Qiang Li, Jun-Duo Wu, Jun-Zhi Liu, Le-Ning Zhang.

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**Writing – review & editing:** Yang Yue, Hua Xin, Fu-Qiang Li, Jun-Duo Wu, Jun-Zhi Liu, Le-Ning Zhang.

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