# The Giant Paraganglioma of The Sigmoid Mesentery：A Case Report

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**Case report**

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# Abstract

**Background:** PGL is originated from pheochromaffin cells in the adrenal paravertebral ganglion and often presented as a slow-growing, painless mass in the clinical, so many PGL are detected by imaging during physical examination. At present, surgery is still the primary treatment for radical treatment of PGL. However, If the disease and medical history cannot be asked in detail before surgery or imaging examination and related tests cannot be finished for diagnosis, the preoperative preparation will be insufficient and increasing the risk of surgery. Here, we reports the treatment of a case with undiagnosed giant sigmoid mesangial PGL before operation.

**Case presentation:** The 56-year-old male patient was admitted to the hospital with "intermittent headache and dizziness for 3 years and aggravation for 1 week". Abdominal and pelvic CT showed that round soft tissue density shadow could be seen beside the left iliac artery in the pelvic cavity. The laparoscopic surgery was performed. The patient's blood pressure increased rapidly when the mass was removed intraoperatively, and decreased sharply when it was removed. Postoperative pathological examination showed that is a PGL.

**Conclusion:** For patients diagnosed as space-occupying lesions accompanied by long-term persistent or intermittent hypertension, careful differentiation and diagnosis should be made in combination with the history and relevant examinations to reduce the misdiagnosis rate of the PGL. Meanwhile, the adequate preoperative preparation, intraoperative anesthesia monitoring and postoperative symptomatic supportive treatment should be done to reduce the risk of surgical treatment of this disease in the perioperative period.


# Background

Pheochromocytomas (PCC)[1] is a rare neuroendocrine tumor originating from pheochromocytes in the embryonic neural crest tissue. Approximately 80% - 85% of PCCs are tumors caused by adrenal medulla pheochromocytes, and 15%-20% of PCCs are tumors originating from extradrenal pheochromocytes in sympathetic paravertebral ganglia of thoracic, abdominal or pelvic system[1]. According to the classification of endocrine organ tumors by the World Health Organization (WHO) in 2017, tumors generated by pheochromocyte in the adrenal medulla are called PCC and generated by pheochromocyte outside the adrenal gland are called paraganglioma (PGL) through anatomical positioning[2]. The PGL is usually performed a painless mass, so hypertension is often the first symptom of the patients. If the disease and medical history cannot be asked in detail before surgery or imaging examination and related tests cannot be finished for diagnosis, the preoperative preparation will be insufficient and increasing the risk of surgery. Here, the author reports the treatment of a case with undiagnosed giant sigmoid mesangial PGL before operation as follows

# Methods

The 56-year-old male patient was admitted to the hospital with "intermittent headache and dizziness for 3 years and aggravation for 1 week". Before 3 years, he had a sudden intermittent headache, dizziness and discomfort, and had measured his blood pressure is higher than 140/90 mmHg for many times, up to 180/116 mmHg. He began taking oral antihypertensive drugs for antihypertensive treatment after the diagnosis has been hypertension (amlodipine 1.5mg 1 time/day, irbesartabn 150mg 1 time/day), and regular monitored of blood pressure. The blood pressure control around 140/90 mmHg. His headache and dizziness had been worsened accompanied by nausea, visual rotation, chest shortness of breath and other symptoms.the blood pressure was 160–170/100-110mmHg at 1 week ago. He was admitted to the department of cardiology for hypertension after blood pressure was measured at 180/114mmHg. Physical examination on admission: the vital signs were stable, blood pressure was measured at 134/105mmHg. The abdomen was flat and soft, with palpable abdominal masses of about 4cm has been felt in the left lower abdomen, with regular boundary, hard texture, inactivity and low skin temperature. Other physical examination showed no abnormality. Abdominal and pelvic CT showed that round soft tissue density shadow could be seen beside the left iliac artery in the pelvic cavity, and small round low-density shadow could be seen inside, at the time, the lesion size was about 4.9*6.8cm (Fig. 1A). After antihypertensive treatment to make the condition stable, it was classified as "1. Sigmoid mesangial mass 2. Essential hypertension grade 3 (extremely high risk); 3.Type 2 diabetes" was transferred to general surgery. The laparoscopic surgery was performed after the preoperative examinations were completed and the contraindications for surgery were eliminated. Microscopically, there was a 4*6cm pliable mass (Fig. 1B) at the mesangial root of the sigmoid colon, with a fair degree of motion, clear boundary with the intestine, no effusion in the abdomen and pelvis, and no metastatic lesion on the abdominal wall. The tumor was carefully separated along the mesentery and the tumor capsule with ultrasound scalpel, the blood pressure of the patients was significantly increased after intraoperative instrument touching the tumor, with the highest blood pressure reaching 200/108mmHg, Paraganglioma was considered and surgical procedures were suspended at that time. After the communication with the anesthesiologist given phentolamine symptomatic treatment to reduce and stabilize blood pressure, the operator separated the mass carefully. The electrocautery unit blood carefully after complete resection of the mass. At this point, the patient's blood pressure was significantly reduced to 64/40mmHg. The norepinephrine and dopamine were administered as vasopressors to treatment after the anesthesiologist had evaluate. After the operation, the patient needed vasoactive drugs for maintenance due to low blood pressure, so he was transferred to ICU for further transitional treatment after the operator communicated with the anesthesiologist. On the third day after the operation, the patient had an average blood pressure of 98/65mmHg and was transferred to the department of general surgery for further treatment. On the seventh days after the operation, the patient recovered and was discharged.

Postoperative pathological examination showed that: the size of the tumor is about 6.6*4.8*3.2cm and is a PGL (Fig. 1C, D).

# Discussion And Conclusion

PGL is originated from pheochromaffin cells in the adrenal paravertebral ganglion. It was more common near the abdominal aorta that is accounting for about 10%-15%, and followed by the hilum of the kidney and the inferior vena cava[3]. It's incidence is between 0.01% and 0.03%[4]. The PGL that have function can secrete large amounts of catecholamines which leading to persistent or paroxysmal hypertension and related signs in more than 95% of patients[5]. Headache, sweating, and palpitations are the typical triad of PGL. Some patients also experience orthostatic hypotension, pallor, tremor and even psychological symptoms including acute anxiety and panic attacks. In addition, large amounts of catecholamines can lead to constipation, intestinal dilatation, cholestasis, or gallstones and so on.

PGL is often presented as a slow-growing, painless mass in the clinical, so many PGL are detected by imaging during physical examination. CT and MRI are common imaging methods. On CT, benign PGL usually presents as round or quasi-round soft tissue masses with well-defined boundaries and uneven density[6]. In MRI, PGL usually presents hypointensity on T1WI, while T2WI and DWI show hyperintensity [7]. In addition, the content of catecholamines, normetanephrine, metaneph-rine in plasma in blood, and $^{131}$I-MIBG, PET-CT and genetic tests can be used as auxiliary examination methods.

At present, surgery is still the primary treatment for radical treatment of PGL, but sudden increase in intraoperative blood pressure and sharp drop in postoperative blood pressure may increase the risk of death. In order to reduce the incidence of surgical complications and mortality, preoperative medication should be adequately prepared to control blood pressure. At the same time, restoring blood volume is indispensable. Adrenergic block is usually administered at least 7 days before surgery with nonselective or selective alpha-receptor antagonists, along with a high-sodium diet and high fluid intake[6, 8]. Sudden reduction of catecholamines after surgery can lead to sudden drop in blood pressure, decreased circulation resistance and relatively insufficient circulating blood volume which can lead to severe hypotension. At this point, α-receptor agonists and fluid supplementation should be given to perform the symptomatic treatment.

In this case, the possibility of PGL was not considered duo to the patient's good blood pressure control with previous antihypertensive drugs. So preoperative preparation for the treatment of α-receptor antagonist was not performed on the patient during the perioperative period. The rapid release of catecholamines during the separation mass during surgery causes a sudden increase in blood pressure, which increases the risk of adverse outcomes such as a hypertensive crisis, acute left heart failure, pulmonary edema, cerebrovascular accident, or severe arrhythmias. After considering the possibility that the tumor was PGL, the surgeon suspended the operation. And the tumor was carefully and slowly removed after the anesthesiologist gave phentolamine to reduce and stabilize the blood pressure. The patient showed hypotension after tumor resection, and the anesthesiologist and the ICU physician gave norepinephrine dopamine, fluid rehydration and volume dilation symptomatic treatment. On the second day after the operation, the patient's blood pressure returned to the normal range. After the condition was stable, the patient was transferred to the general ward for observation and treatment. After the condition was stable, the patient was transferred to the general ward for observation and treatment.

Early detection, complete tumor resection and appropriate clinical follow-up are key management strategies for patients with PGL. The case report of the sigmoid mesangial PGL is helpful to improve the preoperative understanding, diagnosis and treatment of parageneuroma by clinicians. For patients diagnosed as space-occupying lesions accompanied by long-term persistent or intermittent hypertension, careful differentiation and diagnosis should be made in combination with the history and relevant examinations to reduce the misdiagnosis rate of the disease. The adequate preoperative preparation, intraoperative anesthesia monitoring and postoperative symptomatic supportive treatment should be done to reduce the risk of surgical treatment of this disease in the perioperative period.

# Abbreviations

| Full name | Abbreviations |
|---|---|
| Pheochromocytomas | PCC |
| paraganglioma | PGL |

# Declarations

### Ethics approval and consent to participate

This manuscript is not reported studies involving human participants, human data or human tissue. So the “ethics approval and consent to participate” isn’t appropriate for our manuscript.

### Availability of Data and Materials

All the data are available in the patient’s medical record.

### Consent for publication

Written informed consent was obtained from the patient for the publication of this case report and the accompanying images.

### Competing interests:

We confirm that the manuscript has been read and approved by all named authors and that there are no other persons who satisfied the criteria for authorship but are not listed. We further confirm that the order of authors listed in the manuscript has been approved by all of us. We also declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

We understand that the Corresponding Author is the sole contact for the Editorial process. He is responsible for communicating with the other authors about progress, submissions of revisions and final approval of proofs.

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### Authors' contributions:

**Xin Xin Wang:** manage patients and collect clinical data and write the manuscript; **Yuan Li:** collect clinical data**Xiao Jun Yang:** manage patients and revision of the manuscript.

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# Figures

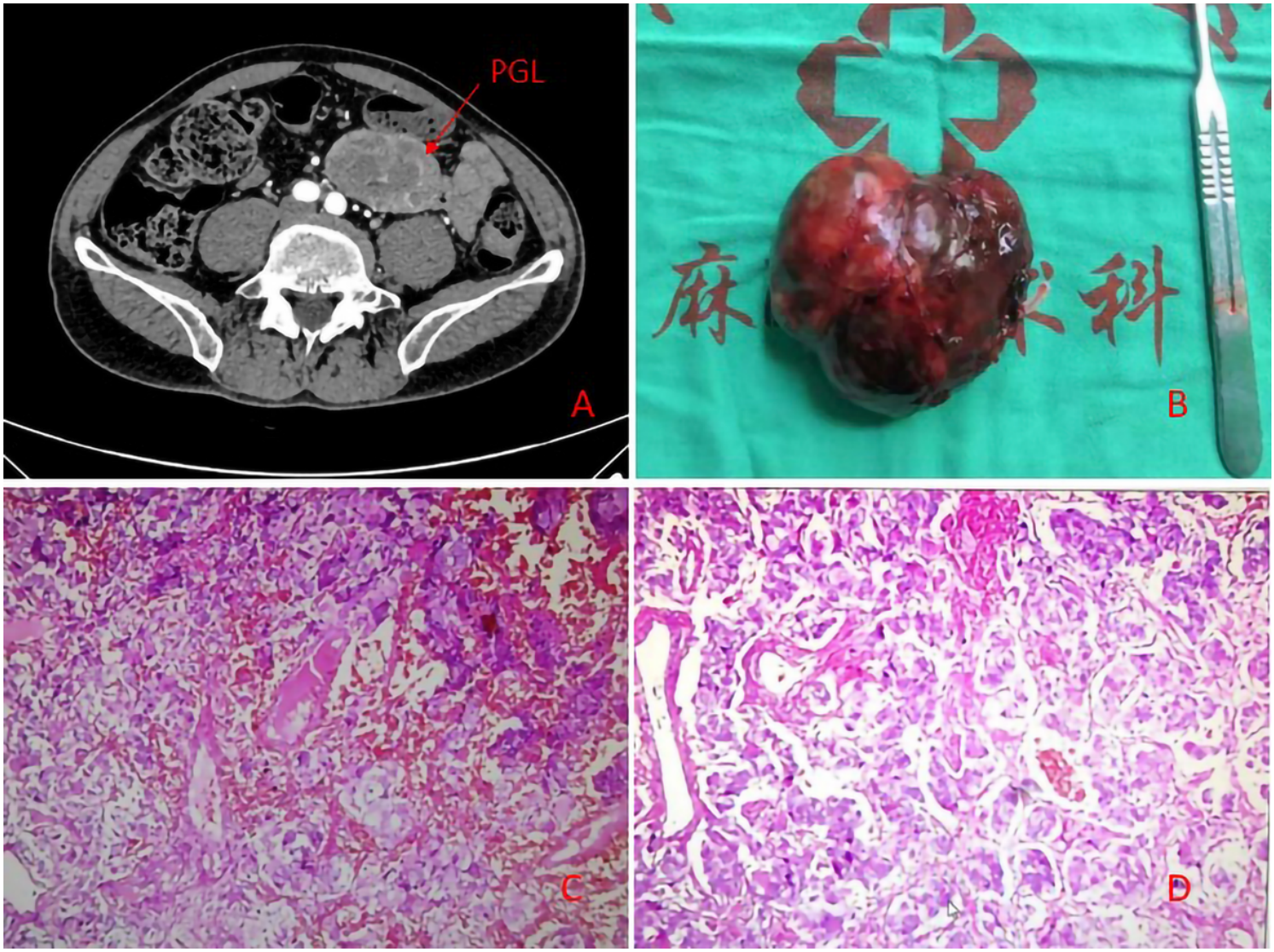


**Figure 1**

A: Arterial phase of enhanced CT；B: Postoperative image of whole tumor specimen；C、D: HE staining suggested paraganglioma (100×).