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A Case of Giant Pheochromocytoma with Type B Aortic Dissection

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Abstract: Pheochromocytoma is a chromaffin tissue originating from the adrenal medulla, sympathetic ganglia, or other sites, synthesizing and secreting catecholamines to cause persistent or paroxysmal hypertension alongside multifaceted organ dysfunction and metabolic disorders. However, no cases of giant pheochromocytoma complicated by type B aortic dissection have been previously reported. This retrospective analysis details the diagnosis and management of one such case, alongside a review of relevant literature, to inform treatment strategies for giant pheochromocytoma. The case is reported as follows.

Keywords: Giant pheochromocytoma; Type B aortic dissection; Perioperative management

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1. Introduction

Pheochromocytomas originate from the adrenal medulla or sympathetic ganglia. Due to paroxysmal or persistent excessive secretion of catecholamines, they can cause secondary hypertension and multi-organ dysfunction. Approximately 10% present as giant tumors (diameter > 6 cm). These tumors are large, highly vascularized, and anatomically complex, making them prone to inducing catecholamine crises during surgery and significantly increasing the risk of perioperative cardiovascular and cerebrovascular events. Stanford Type B aortic dissection (TBAD) is a life-threatening condition involving a false lumen formed after intimal tear, with hypertension being a core risk factor for its development and progression. Theoretically, hypertension caused by pheochromocytoma would further exacerbate aortic wall stress and elevate dissection risk. However, the coexistence of giant pheochromocytoma and type B aortic dissection is extremely rare, with no reported cases domestically or internationally. This dual condition creates multiple conflicting treatment strategies: preoperative alpha-blocker therapy for 2–4 weeks is required to lower blood pressure and expand blood volume in pheochromocytoma patients to reduce intraoperative fluctuations, while TBAD necessitates strict perioperative blood pressure control

to prevent dissection progression. Additionally, the need for anticoagulation therapy following endovascular aortic repair conflicts with the requirement for strict blood pressure control during pheochromocytoma surgery to prevent severe blood pressure fluctuations. This creates significant tension and demands higher standards for individualized management through multidisciplinary collaboration. This paper reviews the clinical data of a case of giant pheochromocytoma (maximum diameter 183 mm) with TBAD. By integrating literature findings, it explores the diagnostic and therapeutic challenges and proposes corresponding strategies, aiming to provide reference for managing such complex comorbidities.

2. Clinical data

Patient, male, aged 61, admitted to our hospital's Emergency Department on 11 April 2024 presenting with 'sudden onset of severe chest and back pain for 8 days', accompanied by dizziness, palpitations, nausea and vomiting. Systolic blood pressure peaked at 156/85 mmHg. The patient reported no history of hypertension and normal blood glucose levels. CTA findings indicated: A large tear measuring approximately 12 mm in width was observed in the distal aortic arch. Within the thoracic aorta and upper abdominal aorta, a linear shadow of torn intimal fragments was visible, dividing the aortic lumen into true and false lumens. A massive mass in the right upper abdomen, with the largest cross-section measuring approximately 183 mm × 139 mm. The right renal artery, right renal vein, inferior vena cava, and right branch of the portal vein were markedly compressed and displaced. The right adrenal artery originated from the false lumen to supply the right adrenal mass. Additionally, a right hepatic artery was observed arising from the false lumen of the abdominal aorta and entering the right upper abdominal mass. Urinary free adrenaline: 176.61 μg/24 h (normal range 0–20 μg/24 h); urinary free noradrenaline: 199.85 μg/24 h (normal range 0–90 μg/24 h); urinary vanillylmandelic acid 218 mg/24 h (normal range 0–12 mg/24 h). Other biochemical and imaging studies revealed no significant abnormalities.

2.1. Preliminary diagnosis

(1) Stanford type B aortic dissection (TBAD)

(2) Massive right adrenal mass: pheochromocytoma

2.2. Surgery performed

Given the risk of rupture and hemorrhage during perioperative management, the patient underwent endovascular repair with a thoracic aortic stent graft in the Department of Interventional Vascular Surgery on 17 April 2024. Postoperatively, the patient's condition stabilized and they were transferred to the Department of Urology for continued management. Preoperatively, the patient received 1 mg/day of prazosin for blood pressure control, 47.5 mg/day of metoprolol succinate for heart rate management, and 10 mg/day of rivaroxaban for anticoagulation. After four weeks of volume expansion, surgery was scheduled.

2.3. Surgical procedure

Under general anesthesia, a supine position was adopted with a mid-abdominal inverted T-shaped incision. A retroperitoneal tumor measuring approximately 22 × 17 × 5 cm was identified. During dissection, marked adhesion between the tumor and liver was noted, with the inferior vena cava adhered to and displaced anteriorly under pressure. The portal vein was compressed, and the right renal artery and vein were partially encased by the tumor. Consequently, the right hepatic lobe, gallbladder, and right kidney were resected. Intraoperatively,

compression of the tumor released substantial catecholamines, causing blood pressure fluctuations with a peak systolic reading of 290/150 mmHg and heart rate rising to 130 beats per minute. During final tumor dissection, the right adrenal artery ruptured at its adhesion to the abdominal aorta false lumen, causing massive hemorrhage from the tear. Approximately 1500 mL of blood was lost instantaneously, with blood pressure plummeting to 20/5 mmHg. Immediate pressure was applied to the tear site whilst repairing the ruptured vessel. Concurrently, blood and fluid transfusion rates were accelerated, and the norepinephrine infusion rate was increased. The patient was transferred to the ICU for further monitoring postoperatively. Following hemodynamic stabilization after 2 days, they were moved to a general ward and discharged in good health after 14 days. The postoperative pathology report indicated a mass measuring 22 × 17 × 8 cm, solid in consistency with soft texture and regional cystic changes. Immunohistochemical findings were consistent with pheochromocytoma. See **Figure 1**–**3**.

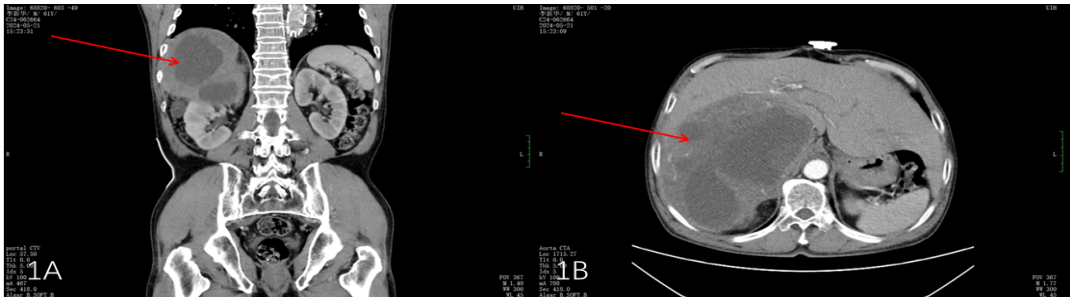

Figure 1. Preoperative contrast-enhanced CT revealed a right adrenal tumour and a thoracic aortic stent graft.

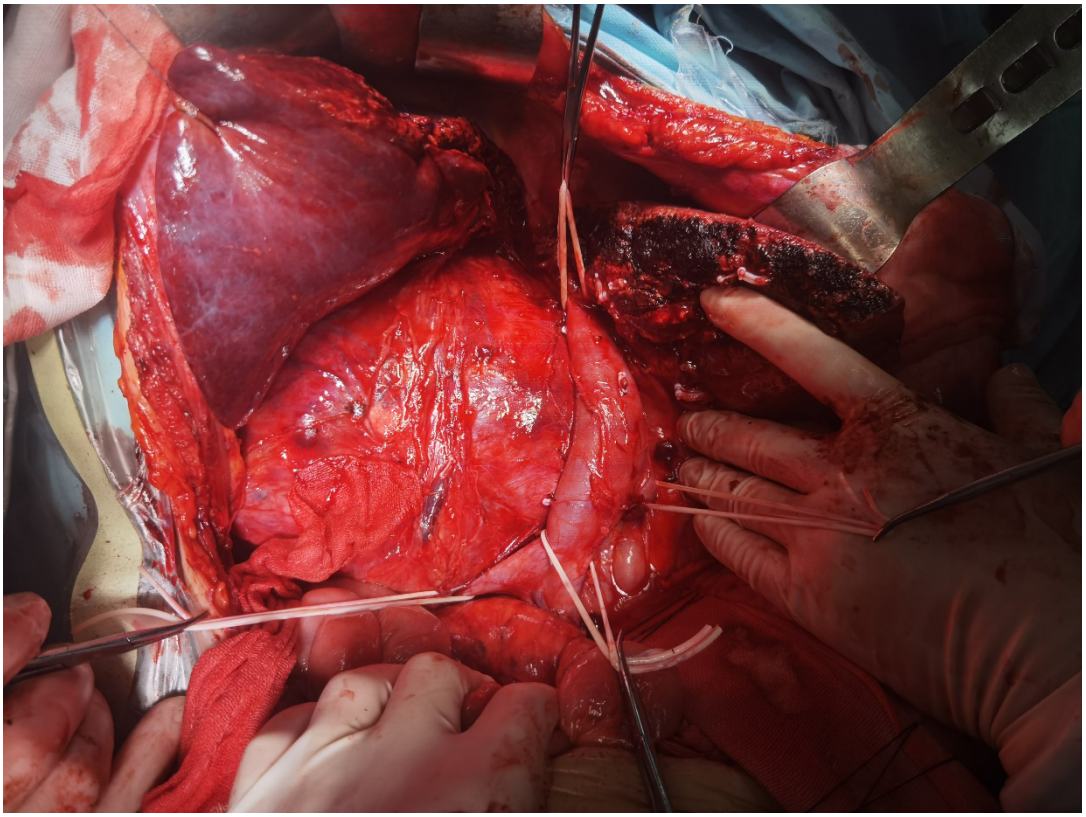

Figure 2. Following intraoperative division of the right hepatic lobe, the mass was fully exposed.

Figure 3. Tumor resection specimen. Left: Right kidney; Centre: Gallbladder; Right: Right hepatic lobe and adrenal

3. Discussion

Pheochromocytoma is a rare neuroendocrine tumor of chromaffin cell origin, primarily arising in the adrenal glands. Approximately 10% originate from the sympathetic nerves of the adrenal glands and are termed paragangliomas. These tumors continuously or intermittently release substantial amounts of catecholamines, such as noradrenaline, adrenaline, and dopamine, inducing a clinical syndrome characterized by elevated blood pressure or metabolic alterations, alongside severe complications affecting the cardiovascular and cerebrovascular systems [1]. Pheochromocytoma, also termed the "10% tumor", exhibits near-equal incidence between genders. Approximately 10% occur in children, 10% arise extra-adrenally, 10% are malignant, 10% represent multiple or bilateral adrenal tumors, and 10% are familial in origin [2].

Preoperative diagnosis of pheochromocytoma encompasses both qualitative and localizing assessment [3]. Qualitative diagnosis involves measuring plasma and urinary free catecholamine levels, including epinephrine, norepinephrine, dopamine, and their metabolites such as vanillylmandelic acid, alongside plasma free methoxymethamphetamine. Localization diagnosis involves adrenal contrast-enhanced CT and MRI, with multi-slice spiral contrast-enhanced CT demonstrating exceptionally high clinical value for pheochromocytoma diagnosis [4]. For giant pheochromocytomas, determining the tumor's relationship with surrounding vessels and organs necessitates supplementary CT angiography (CTA).

Surgical resection remains the most effective treatment for pheochromocytoma. For tumours under 6 cm, most scholars recommend laparoscopic surgery, which offers advantages over open surgery including reduced intraoperative catecholamine release, smaller blood pressure fluctuations, less postoperative pain, and faster

recovery [5]. No definitive size threshold currently exists for giant pheochromocytomas. Some literature proposes tumors > 6 cm in diameter as giant pheochromocytomas, while others define those > 10 cm as such [6]. These larger tumors are rarer and exhibit non-specific clinical presentations. Maharaj's retrospective analysis of 36 cases exceeding 10 cm revealed only 6 presented with typical symptoms, whilst 12 remained asymptomatic [7]. The present patient admitted with thoracic and dorsal pain, lacking the classic triad of headache, sweating, and palpitations. For giant pheochromocytomas or invasive tumors, open surgery is preferable for achieving complete resection and preventing tumor rupture [8,9]. Given the characteristics of pheochromocytoma, preoperative alpha-adrenergic blockade with agents such as phenoxybenzamine, prazosin, trazolide, or doxazosin. These agents antagonize catecholamine-induced arterial vasoconstriction, relax peripheral vascular smooth muscle, and reduce vascular resistance, thereby effectively preventing adrenal crisis during anesthesia and surgery [10]. Preoperative preparation for pheochromocytoma surgery requires at least two weeks, with larger tumors necessitating extended preoperative management [11].

Thorough perioperative preparation by experienced surgeons and anesthetists is pivotal for successful surgery on giant pheochromocytomas. Studies indicate that large tumor size and elevated preoperative catecholamine levels increase perioperative complications, including severe arrhythmias, hypertensive crises, and cardiovascular or cerebrovascular events. A rare feature in this case was the presence of regional cystic components, whereas pheochromocytomas are typically solid. Goldberg A et al. reported a case where cystic fluid contained high concentrations of catecholamines, suggesting that compressing the mass during surgical resection could release substantial amounts of these substances [12]. Despite adequate preoperative alpha-adrenergic blockade initiated four weeks prior, an hypertensive crisis necessitated surgical interruption during the procedure.

4. Conclusion

In summary, this case represents an unreported association of giant pheochromocytoma with type B aortic dissection. Hypertension constitutes a primary cause of postoperative mortality in TBAD patients. with postoperative blood pressure recommended at 120/80 mmHg and heart rate 60–80 bpm [13]. Despite active anesthetic intervention during surgery, blood pressure and heart rate remained substantially above these recommended ranges. This may have contributed to residual dissection expansion and perioperative aortic complications, undoubtedly increasing the surgical complexity in this case. From this case, it summaries the following:

(1) Due to the substantial catecholamine secretion from giant pheochromocytomas, preoperative preparation requires over four weeks. Should alpha-blockers prove inadequate for blood pressure control, calcium channel blockers may be added. For patients with elevated heart rates (> 90 bpm), β-blockers should be administered to maintain blood pressure and heart rate within optimal parameters. Caution is warranted against sole reliance on β-blockers, as selective β2-receptor blockade may impair the vasodilatory effects of epinephrine, potentially precipitating hypertensive crises, myocardial infarction, pulmonary oedema, and other complications [14].

(2) Thorough preoperative discussion and comprehensive preoperative investigations are essential. Multidisciplinary consultation is required to determine tumor location, size, and relationships with surrounding organs and vessels. This is critical for establishing the surgical approach, minimizing intraoperative injury, and reducing postoperative complications. In this case, CT and CTA examinations

provided relatively accurate descriptions of the tumor and its relationship with surrounding organs and major vessels. Consequently, during preoperative communication with the patient, it was explained that the right hepatic lobe, gallbladder, and right kidney might be resected during surgery.

(3) An inverted T-shaped midline incision was employed in this case, extending from the xiphoid process to the umbilical level, and from the anterior axillary line on the left to the right. Intraoperative exposure was satisfactory. Apart from tearing the adhesion between the right adrenal artery and the pseudocavity during final tumor dissection, there was minimal vascular or organ injury. Even when injury occurred, the favorable exposure allowed for prompt hemostasis. Given the rich blood supply and extensive collateral circulation of giant pheochromocytomas, ligation or occlusion of feeding vessels during surgery obstructs tumour venous return, potentially increasing intraoperative oozing [15]. Consequently, ample plasma reserves must be prepared for volume replacement, alongside sufficient α-adrenergic blockers and noradrenaline to manage blood pressure fluctuations.

Disclosure statement

The authors declare no conflict of interest.

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