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Perioperative Management of a Giant Pheochromocytoma Resection: A Case Report with Literature Review

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Abstract

Background: Pheochromocytoma, a rare endocrine tumor, produces substantial quantities of catecholamines, leading to severe cardiovascular complications. Perioperative management for its surgical resection entails significant risks. This case report, detailing the anesthetic and perioperative management of a giant pheochromocytoma excision, underscores the complexity inherent in managing these tumors and emphasizes the critical importance of employing multiple monitoring modalities for dynamic assessment to guide timely therapeutic interventions.

Case Report: We report a case of a 31-year-old female patient who presented with abdominal pain and was found to have a large mass in the left retroperitoneal adrenal region. The diagnosis of pheochromocytoma was confirmed by plasma catecholamine assays and high-resolution computed tomography (HRCT). Following preoperative pharmacologic preparation with alpha-adrenergic blockade, surgical resection of the tumor was performed. Intraoperatively, the patient developed significant hemodynamic instability, notably persistent hypotension after tumor removal that was refractory to empirical management. Combined transesophageal echocardiography (TEE) and pulse index continuous cardiac output (PiCCO) monitoring were employed for comprehensive assessment of cardiac function and volume status. Real-time guidance from this multimodal monitoring facilitated dynamic adjustments to the therapeutic strategy, enabling goal-directed hemodynamic support. Ultimately, the patient experienced an uncomplicated postoperative course.

Conclusion: This article integrates the features of the presented case with a systematic review of the literature on pheochromocytoma, comprehensively summarizing key diagnostic considerations, therapeutic strategies, and perioperative management protocols to serve as a clinical reference.

Keywords

Giant Pheochromocytoma, Perioperative Management, Hemodynamics, Anesthesiology, Multimodal Monitoring; Vasoplegia, Case report

Case Report

Background

Pheochromocytoma is a rare neuroendocrine tumor that can continuously or intermittently secrete catecholamines, leading to severe circulatory fluctuations, and in severe cases, multi-organ dysfunction [1]. Currently, surgical resection of the tumor is the first-line treatment for pheochromocytoma. However, the surgical and anesthetic procedures themselves can trigger a massive release of catecholamines, resulting in hemodynamic instability and even cardiovascular or cerebrovascular complications (such as arrhythmias, stroke, myocardial infarction, heart failure, etc.), which can be life-threatening in severe cases [2,3].

In patients with pheochromocytoma, larger tumor size is associated with a higher likelihood of severe perioperative hemodynamic fluctuations and postoperative complications. Particularly for tumors larger than 4.25 cm in diameter, strict preoperative preparation is essential [4]. Although α-adrenergic receptor blockers are one of the standard preoperative medications for pheochromocytoma, studies have found that even with adequate preoperative preparation using these drugs, maintaining stable perioperative hemodynamics remains challenging [5].

In this case, despite satisfactory preoperative preparation for a patient with a giant pheochromocytoma, severe hemodynamic instability still occurred and showed poor response to empirical treatment. We dynamically assessed the patient's cardiac function and volume status using a combination of TEE and PiCCO monitoring, allowing timely adjustment of treatment strategies, which ultimately helped the patient successfully overcome the critical condition.

Additionally, we reviewed previous literature and current research on pheochromocytomas and paragangliomas, summarizing and outlining their diagnostic approaches, treatment options, and perioperative management strategies to provide a reference for clinical practitioners.

Case Presentation

A 31-year-old Chinese female patient was admitted to the hospital after an outpatient examination for vague abdominal pain revealed a left retroperitoneal mass. Her blood pressure was 140/95 mmHg, and electrocardiography showed sinus tachycardia with a heart rate of 112 beats per minute. Abdominal CT indicated a large heterogeneous cystic-solid mass (approximately 14.7×15.9×25.3 cm, **Fig-1**) in the left adrenal region of the retroperitoneum. Plasma catecholamine levels were elevated: norepinephrine (NE) 49.59 nmol/L (reference: <5.17 nmol/L), metanephrine 17.46 nmol/L (reference: <0.42 nmol/L), and epinephrine (E) 15.06 nmol/L (reference: <0.34 nmol/L). Cardiac ultrasound showed mild aortic dilation, mild-to-moderate aortic regurgitation, and a patent foramen ovale. Thyroid function tests, blood glucose, and other examinations were normal. Based on preoperative laboratory and imaging findings, the patient was diagnosed with a left adrenal pheochromocytoma and was discharged with oral terazosin hydrochloride (an α-blocker) at 2 mg once daily for two months.

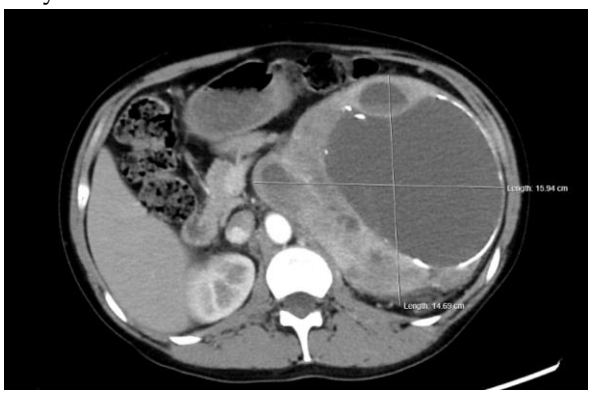

Fig-1: Maximum Diameter and Adjacent Conditions of Left Adrenal Tumor

Upon readmission, the patient's plasma catecholamine levels were within normal ranges, with blood pressure at 115/87 mmHg and heart rate at 90 bpm. Her body weight had increased by 5 kg, and hematocrit (Hct) had decreased by 6%. For preoperative volume expansion, she received daily infusions of 1000 mL crystalloids and 500 mL colloids for three days before surgery. One day prior to surgery, a multidisciplinary team (including endocrinology, intensive care, urology, and anesthesiology specialists) conducted a comprehensive evaluation and concluded that preoperative preparation was adequate and the tumor margins were well-defined. Consequently, a laparoscopic left

retroperitoneal tumor resection was planned.

Upon entering the operating room, the patient's vital signs showed a heart rate of 120 bpm, blood pressure of 140/100 mmHg, respiratory rate of 20 breaths/min, and SpO_2 of 100%. Considering preoperative anxiety, 2 mg midazolam was administered, resulting in mild reductions in heart rate and blood pressure. After adequate sedation and local anesthesia, a left radial arterial catheter was placed for continuous blood pressure monitoring. Anesthesia induction was achieved with 100 mg propofol, 25 μg sufentanil, 50 mg rocuronium, and 0.5 mg penehyclidine hydrochloride. The oropharynx and trachea were then topically anesthetized with 5 mL of 2% lidocaine spray under videolaryngoscope guidance. After ensuring adequate drug effect, a 6.5F endotracheal tube was inserted. Anesthesia maintenance consisted of intravenous remifentanil at 0.2 μg/kg/min and propofol at 5 mg/kg/h, supplemented with 1% sevoflurane inhalation, with dosages adjusted according to anesthesia depth. Following induction, a right internal jugular venous catheter was placed for intraoperative fluid administration and CVP monitoring, along with BIS and core temperature monitoring.

Prior to tumor resection, the patient developed severe hypertension (peak BP 230/100 mmHg) and tachycardia (peak HR 150 bpm). To control the hypertensive crisis, intravenous nitroglycerin infusion was initiated, with intermittent boluses of nicardipine, phentolamine, and esmolol administered. Following tumor removal, the patient's blood pressure precipitously dropped to 65/34 mmHg. Despite aggressive fluid resuscitation combined with intravenous methylprednisolone (80 mg), continuous infusions of epinephrine and norepinephrine, and intermittent norepinephrine boluses (50–100 μg), the patient exhibited refractory hypotension with minimal blood pressure improvement.

TEE revealed an underfilled heart with a cardiac output of ~6 L/min. PiCCO monitoring via the left radial artery showed markedly reduced systemic vascular resistance (SVRI: 650 $dyn{\cdot}s/cm^3{\cdot}m^3$; normal range: 1970–2390 $dyn{\cdot}s/cm^3{\cdot}m^3$), consistent with catecholamine withdrawal-induced vasodilation. The persistent hypotension was attributed to acute catecholamine withdrawal following tumor resection, resulting in peripheral vasodilation. Consequently, we escalated the dosage of vasoactive agents and initiated vasopressin infusion, while continuously optimizing the treatment strategy based on real-time TEE and PiCCO monitoring findings.

One hour before surgery completion, norepinephrine, epinephrine, and vasopressin were gradually tapered. Upon leaving the OR, only norepinephrine and vasopressin infusions were maintained, effectively sustaining BP within 100–120/50 mmHg. The surgical procedure lasted 5 hours in total, with an estimated blood loss of 2000 mL and urine output of 1100 mL. Intraoperative fluid resuscitation included: packed red blood cells 3.5 units, leukocyte-depleted red blood cells 1.5 units, fresh frozen plasma 300 mL, crystalloids 4000 mL, and colloids 1000 mL.

The patient was successfully extubated in the intensive care unit (ICU) on postoperative day 1. Vasopressor support with norepinephrine and vasopressin was continued until day 3, when hemodynamic stability was achieved and medications were discontinued, allowing transfer to the general ward. The patient was discharged with clinical improvement after one week. Follow-up at 3 and 6 months postoperatively demonstrated good recovery without evidence of pheochromocytoma recurrence or residual disease.

Discussion

Pheochromocytomas and paragangliomas (PCC/PGL) originate from the adrenal medulla and extra-adrenal sympathetic paraganglia, respectively. Among these, pheochromocytomas and some paragangliomas are classified as functional tumors due to their ability to secrete catecholamines [3]. The released catecholamines - including epinephrine, norepinephrine, metanephrines, and dopamine - can induce clinical manifestations such as hypertension, tachycardia, headaches, palpitations, and profuse sweating. In severe cases, patients may experience panic attacks, syncope, or sudden death due to catecholamine-induced cardiovascular collapse [4].

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Patients harboring undiagnosed and untreated functional tumors face a significantly elevated risk of cardiovascular events. Consequently, enhancing the sensitivity and accuracy of diagnostic methods is paramount. Current diagnosis of PCC/PGL primarily relies on plasma free metanephrines or 24-hour urinary fractionated metanephrines (normetanephrine [NMN] and metanephrine [MN]). Measurements exceeding twice the upper reference limit are generally considered diagnostically significant [1,3]. When measuring plasma catecholamines, patients are advised to avoid tyramine-rich foods for 2–3 days prior to testing to enhance accuracy [6]. HRCT is established as the diagnostic gold standard due to superior spatial resolution compared to magnetic resonance imaging (MRI). However, when HRCT yields negative results, radioisotope imaging may provide complementary diagnostic value [7,8]. Ultrasonography is also a valuable modality for pheochromocytoma detection, demonstrating a sensitivity of approximately 90% [3]. Given that a subset of pheochromocytomas and paragangliomas harbor genetic predispositions [9], which may alter surgical management, universal genetic screening is recommended for all patients with PCC/PGL.

Surgical resection represents the sole curative intervention for these tumors. However, factors including patient anxiety, anesthetic and surgical manipulation, and pharmacological agents may precipitate massive catecholamine release [10], resulting in severe hemodynamic instability. Consequently, meticulous preoperative preparation is critical. Current guidelines designate α-adrenergic receptor blockers as first-line pharmacotherapy for preoperative optimization in pheochromocytoma, recommending initiation 7 to 14 days prior to surgery. Patients with established catecholamine-induced end-organ damage may require an extended preparation period [11]. β-blockers may be added for persistent tachycardia despite adequate α-blockade. The objectives of comprehensive preoperative preparation are to achieve normalization of blood pressure and heart rate, ensure adequate volume expansion to restore intravascular volume, and mitigate perioperative catecholamine surges and associated adverse cardiovascular sequelae [8].

Recent studies have revealed that despite adequate preoperative preparation, patients exhibit cardiovascular complication rates and perioperative mortality similar to those without preoperative optimization, with comparable peak intraoperative systolic blood pressure [12–14]. Tumor size has been identified as significantly correlated with intraoperative hemodynamic instability, particularly for lesions exceeding 4.25 cm. Furthermore, larger tumors are associated with increased risk of adrenal crisis following resection [4,15]. This phenomenon may be attributed to greater hormonal activity, more pronounced volume depletion, and acute hormone withdrawal after excision in larger tumors. Increased surgical difficulty associated with larger tumor size may also contribute to hemodynamic fluctuations [16]. Additionally, catecholamine levels substantially impact circulatory stability: epinephrine induces compensatory downregulation of cardiac β-adrenergic receptors, reducing myocardial contractility, while both epinephrine and norepinephrine cause vasoconstriction via adrenergic receptor activation, exacerbating hypovolemia [15]. Frequent administration of antihypertensive agents for severe preoperative hypertension may lead to residual pharmacological effects, prolonging the duration and severity of post-resection hypotension.

The choice of surgical approach critically influences patient outcomes. Compared with open surgery, minimally invasive techniques (laparoscopic or robotic) significantly reduce hospital length of stay and postoperative complication rates [17]. Robotic-assisted procedures demonstrate superior clinical outcomes relative to conventional laparoscopy in selected cases [18]. However, the optimal approach requires comprehensive evaluation based on individual patient factors and the surgical team's expertise.

Despite the large tumor size in this case, laparoscopic resection was performed following multidisciplinary discussion, considering the patient's absence of comorbidities, adequate preoperative preparation, and well-demarcated tumor boundaries. Our surgeons are highly skilled in laparoscopic giant tumor resection, and given the minimally invasive approach's benefits—less

pain, quicker recovery, and reduced hospitalization—the multidisciplinary team unanimously recommended laparoscopic surgery. Preoperatively, three-day volume expansion therapy was administered to ameliorate vasoconstriction and replenish intravascular volume. Nevertheless, significant hemodynamic fluctuations persisted prior to tumor resection, necessitating continuous nitroglycerin infusion and intermittent intravenous antihypertensive boluses for blood pressure control.

Following tumor excision, refractory hypotension developed that proved unresponsive to empirical management. To assess volume status and cardiac function, TEE and PiCCO monitoring were emergently implemented. These modalities enabled dynamic assessment of myocardial contractility, blood volume, and systemic vascular resistance (SVR), guiding real-time therapeutic adjustments that effectively reversed hemodynamic instability and prevented adverse outcomes. TEE provided real-time visualization of systolic/diastolic function and valvular status, offering quantitative cardiac performance data. PiCCO dynamically tracked stroke volume (SV), cardiac output (CO), and SVR, generating evidence-based parameters for clinical decision-making. A notable limitation was the absence of archived monitoring images.

Postoperative management of PCC/PGL is critical. Evidence indicates that maintaining a moderately elevated mean arterial pressure (MAP) reduces the incidence of acute kidney injury following tumor resection [19]. Larger tumor size correlates significantly with persistent postoperative hypotension and higher rates of adrenal crisis [4,15]. Consequently, intensive hemodynamic monitoring and rigorous maintenance of homeostatic balance are essential to mitigate adverse postoperative events.

Conclusion

In this case, laparoscopic resection was selected for a large retroperitoneal tumor based on the patient's young age, absence of comorbidities, and well-demarcated tumor boundaries. Despite comprehensive preoperative optimization, intraoperative hypertensive crisis (SBP >220 mmHg) and refractory post-resection hypotension (MAP <55 mmHg for >30 minutes) occurred. Empirical management failed to provide therapeutic direction.

TEE and PiCCO monitoring were implemented to guide perioperative therapy, enabling dynamic titration of interventions through real-time assessment of cardiac function, volume status, and systemic vascular resistance. Hemodynamic management in giant pheochromocytoma resection remains challenging, with core objectives being the maintenance of hemodynamic stability and the optimization of vital organ perfusion. Concurrently, multidisciplinary team coordination and personalized precision protocols are critical determinants of outcomes.

Conflict of Interest

The authors have read and approved the final version of the manuscript. The authors declare no conflicts of interest.

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