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Case Conference

Perioperative Management of Adrenalectomy and Inferior Vena Cava Reconstruction in a Patient With a Large, Malignant Pheochromocytoma With Vena Caval Extension

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PHEOCHROMOCYTOMA IS AN uncommon tumor that can present a significant perioperative challenge, potentially resulting in hemodynamic instability necessitating the use of multiple vasoactive agents. On rare occasions, malignant pheochromocytomas are encountered; these can invade adjacent structures, including the inferior vena cava (IVC), necessitating IVC cross-clamping to achieve surgical resection. This case conference describes the case of a 28-year-old man who presented with hypertension, nausea, and vomiting and was found to have a large, malignant pheochromocytoma with IVC invasion. His clinical course throughout the perioperative period is detailed, with discussion focusing on preoperative medical management, intraoperative hemodynamic and surgical considerations, and postoperative

complications and management in the intensive care unit (ICU) setting.

Pheochromocytoma is a rare neuroendocrine tumor that arises from chromaffin cells of the adrenal medulla or extra-adrenal paraganglia that produces and releases catecholamines and their metabolites.¹ The only cure for pheochromocytoma is surgical resection, with most recent studies demonstrating minimal or zero perioperative mortality, which commonly is attributed to improved preoperative preparation.^{2,3}

There are case reports demonstrating that large pheochromocytomas have the potential to invade into the IVC and result in significant IVC thrombus formation.^{4,5} To date, there exists minimal literature documenting the anesthetic planning and management of these complicated patients.^{6,7} In this case conference, the anesthetic management of a patient with a right adrenal pheochromocytoma with IVC extension who underwent an open radical adrenalectomy with IVC thrombectomy and reconstruction with a postoperative course complicated by rhabdomyolysis, acute kidney injury (AKI), and respiratory

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Fig 1. Coronal magnetic resonance imaging of the abdomen demonstrating right adrenal pheochromocytoma (A) with direct invasion of the infrahepatic inferior vena cava (B). White arrows in (B) delineate the superior and inferior margins of inferior vena cava invasion.

failure requiring prolonged mechanical ventilation is detailed. The discussion focuses on preoperative optimization, intraoperative management, and postoperative management in the ICU setting. The patient provided written informed consent for the publication of this case report.

Case Presentation*

A 28-year-old man with a 2-year history of hypertension presented with new-onset nausea and vomiting. Workup revealed an adrenal mass with IVC invasion. The patient's urine metanephrine and normetanephrine levels exceeded 40,000 µg/24 hours, consistent with pheochromocytoma. A metaiodobenzylguanidine (MIBG) scan was positive for radiographic uptake in the area of the mass, but no scintigraphic evidence of metastatic disease was demonstrated. Magnetic resonance imaging (MRI) confirmed the presence of a 10.3-cm heterogeneous tumor focused in the right adrenal gland with direct invasion into the infrahepatic IVC with no evidence of suprahepatic or intracardiac extension (Fig 1). Given his young age at presentation, the patient underwent genetic screening but was not found to have any known mutations associated with hereditary pheochromocytoma-paraganglioma syndromes. He was scheduled for an open adrenalectomy with IVC thrombectomy and possible nephrectomy to be performed by a team of endocrine and transplant surgeons. No further preoperative cardiac workup was undertaken because of the relatively short duration of symptoms and young age. Due to his significantly elevated preoperative catecholamine levels, metyrosine was added 2 weeks before his scheduled surgery to the terazosin and metoprolol prescribed for preoperative adrenergic blockade, which he tolerated without significant side effects. He underwent salt loading with oral sodium

chloride supplement for 5 days before surgery to promote intravascular volume retention and decrease the potential for postoperative hypotension.

The anesthetic plan included a preoperative thoracic epidural for postoperative pain control and general anesthesia with placement of an endotracheal tube. An arterial catheter and the epidural were placed in the preoperative holding area, and the patient was transferred to the operating room. After induction of general anesthesia and endotracheal intubation, a right internal jugular 9-Fr double-lumen central venous catheter was placed for volume resuscitation and vasoactive drug administration, and a second right internal jugular 9-Fr single-lumen introducer and left femoral venous cannula were placed for possible veno-venous bypass (VVB) if the patient were to be unable to maintain adequate hemodynamics during cross-clamping of the IVC. A transesophageal echocardiography (TEE) probe was inserted atraumatically, and the preprocedure TEE demonstrated normal biventricular function, normal left ventricular diastolic function, no valvular abnormalities, euvoolemia, and evidence of tumor extension into the retrohepatic IVC with unobstructed blood flow around the thrombus (Fig 2). TEE also confirmed that there was no evidence of hepatic vein, suprahepatic, or intracardiac IVC extension.

The patient was positioned supine on the operating room table and a chevron incision was made 2 cm below the right subcostal edge, extending from the anterior axillary line on the right to the midclavicular line on the left subcostal edge, along the anterior abdomen. It was noted at the time of the dissection that the tumor was adherent to the retroperitoneum, and there was extension of the tumor into the IVC as was known previously based on preoperative cross-sectional imaging and intraoperative TEE. The tumor was noted to be adherent to the posterior and lateral wall of the IVC for approximately 5 cm, extending to the junction of the right renal vein. These findings made the case more difficult because it was necessary to gain suprahepatic, subhepatic, and infrarenal control of the IVC and

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Fig 2. Transesophageal echocardiographic imaging of the inferior vena cava (IVC) demonstrating the presence of a large mass in the lumen of the IVC with invasion into the lateral wall of the vessel.

both the right and left renal veins. The porta hepatis (vascular inflow to the liver) was dissected and encircled with a vessel loop in case control of liver inflow and outflow was required in the event of bleeding (Pringle maneuver).

During dissection and mobilization, hypertensive episodes during tumor manipulation occurred and were treated with a combination of nitroglycerin, esmolol, and clevidipine, in addition to a magnesium sulfate infusion. In anticipation of decreased ventricular preload with IVC cross-clamping, volume loading and a vasopressin infusion were initiated. IVC test-clamping demonstrated adequate hemodynamic stability, which showed that VVB was unnecessary. IVC clamps were placed superiorly in the subhepatic area and inferiorly just below the renal veins. Both right and left renal veins were controlled with vessel loops and clamps. The tumor thrombus was adherent to the IVC wall and the junction of the right renal vein and IVC. Approximately 50% of the IVC wall and the junction of the right renal vein were excised to achieve complete curative surgical resection with negative margins (R0 resection) and to extract the thrombus. The IVC then was reconstructed with a Dacron patch, and reimplantation of the right renal vein into the IVC was performed below the patch graft (Fig 3). The IVC and left renal vein clamp time was 50 minutes, with an additional 21 minutes of ischemic time to the right kidney due to the reimplantation of the right renal vein. The right adrenal tumor with the adjacent retroperitoneal tissue and tumor invading the IVC were removed en bloc.

After IVC unclamping, the patient briefly required hemodynamic support with norepinephrine and vasopressin infusions but was weaned from these quickly without difficulty while TEE-directed volume resuscitation was ongoing. The patient received 7 U of packed red blood cells, 5 U of fresh frozen plasma, and 1 U of cryoprecipitate. Estimated blood loss was 3,100 mL. Postprocedure TEE demonstrated normal biventricular function with absence of the IVC tumor thrombus. The right atrium, right ventricle, and pulmonary arteries were inspected thoroughly with TEE to ensure absence of tumor emboli. Volume assessment using TEE appeared consistent with euolemia, suggesting adequate fluid resuscitation.

The epidural catheter was dosed with 0.6 mg of hydromorphone at the beginning of surgery, and an infusion of 0.125% bupivacaine with 10 μ mL of hydromorphone at 6 mL/h for postoperative analgesia was initiated once all vasoactive infusions were stopped at the end of the procedure. Local anesthetic was not used until hemodynamic stability was confirmed because of concerns about worsening hypotension after catecholamine withdrawal.

At the end of the procedure, the patient was hemodynamically stable on no vasoactive agents and breathing spontaneously with a pulse oximetry reading $> 95\%$. His trachea was extubated successfully, and he was transferred to the ICU in stable condition. En route to the ICU, he was noted to be experiencing increased work of breathing with diffuse rhonchi noted on examination. Oxygen saturation declined, and the decision was made to reintubate him shortly after arrival in the ICU. A chest x-ray after intubation demonstrated left basilar atelectasis and bilateral pulmonary edema. The initial arterial blood gas reading after reintubation showed a pH of 7.26, partial pressure of carbon dioxide of 51, and partial pressure of oxygen of 89 on 100% oxygen.

The patient's postoperative course was complicated by rhabdomyolysis, with a peak creatine kinase level of 13,691 u/L and peak creatinine of 2.8 mg/dL (baseline 1.2 mg/dL) on postoperative day (POD) 2. Acute kidney injury from rhabdomyolysis was confounded by the warm ischemia to the kidneys during the IVC cross-clamping and IVC

Fig 3. Intraoperative photograph taken after resection of the adrenal tumor and inferior vena cava and right renal vein reconstruction. IVC, inferior vena cava.

reconstruction, which likely worsened his tolerance of volume expansion. During treatment of the rhabdomyolysis, his respiratory insufficiency worsened due to aggressive fluid administration to promote diuresis. The patient's nadir partial pressure of oxygen dissolved in arterial blood/fraction of inspired oxygen ratio of 70 occurred on POD 3. On POD 3, he also experienced increasing purulent drainage from his sinuses. Bacterial culture of this drainage and a bronchoalveolar lavage on POD 4 both demonstrated 3+ beta-lactamase-negative *Haemophilus influenzae*, consistent with a diagnosis of ventilator-associated pneumonia (VAP). As his renal function improved, he underwent aggressive diuresis with mannitol and furosemide with rapid improvement in his respiratory status and extubation on POD 7. He was discharged home on POD 12. Final surgical pathology was consistent with malignant pheochromocytoma with negative margins.

Discussion

Overview of Preoperative Management

Preoperative pharmacologic preparation for patients with pheochromocytoma is critical for optimal intraoperative hemodynamic control. Several strategies available may be used in combination to tailor an optimal regimen for each patient, based on the patient's level of tumor biochemical activity and pre-existing comorbidities. All these strategies share the basic principle of adequate alpha (α)-blockade. All patients with pheochromocytoma should be started on an α -blocker at least 1-to-2 weeks before surgery to reduce intraoperative blood loss, blood pressure lability, and arrhythmias.^{8–11} Phenoxybenzamine, a nonreversible, nonselective α -blocker used since the 1950s, previously was the preoperative treatment of choice, but concerns about prolonged postoperative hypotension have resulted in the favoring of selective α -1 blocking drugs.¹² Phenoxybenzamine mostly has been replaced by these shorter-acting, selective α -1 blockers, including doxazosin, prazosin, and terazosin. Compared with phenoxybenzamine, the use of selective α -adrenergic blockade is associated with less reflex tachycardia and less postoperative hypotension.¹² Doxazosin has the longest half-life of the selective α -adrenergic blockers and requires only once-a-day dosing; its effect subsides rapidly after surgery, making it an excellent option for a first-line α -adrenergic antagonist medication for patients with pheochromocytoma. Once α -blockade dosing is maximized to the goal of orthostatic hypotension, some patients may need additional β -blockade if tachycardia becomes an issue.¹³ A goal resting heart rate of 60 beats/min is optimal for intraoperative hemodynamic control.¹⁴ Most often, β 1-selective drugs are used. Dihydropyridine calcium channel blockers also can be used as part of the preoperative preparation for patients with pheochromocytoma, particularly in patients who are intolerant of α -blockers, or as an adjunct to α -adrenergic blockade. Calcium channel blockers reduce arterial pressure by blocking norepinephrine-mediated calcium influx into the smooth muscle cells of the vessel wall.^{9,13}

Patients whose tumors secrete significant excess amounts of vasoactive products may benefit from taking metyrosine for a short course (usually 1–2 weeks) directly before surgery. Metyrosine depletes adrenal catecholamine stores by inhibiting tyrosine hydroxylase, the rate-limiting enzyme in catecholamine synthesis. Pheochromocytomas have enhanced tyrosine hydroxylase activity significantly compared with normal adrenal tissue.¹⁵ The authors use metyrosine selectively for approximately 2 weeks before surgical resection, in patients with pheochromocytomas that secrete very large amounts of catecholamines, as was done with the patient described here. Even though the use of tyrosine analogs in the preoperative period has been shown to decrease cardiovascular complications of pheochromocytoma resection, they are not without side effects.¹⁶ Patients may develop diarrhea, somnolence, anxiety or agitation, tremors, or extrapyramidal symptoms.¹⁷ These can be minimized if the treatment is limited to the 2 weeks leading up to surgery. In the patient described here, the risk of significant perioperative lability due to his very high preoperative catecholamine levels suggested that a short course of metyrosine outweighed the risk of any systemic side effects. Fortunately, the patient tolerated metyrosine without issue.

Patients with pheochromocytoma typically experience significant volume contraction and thus require preoperative intravascular volume resuscitation after starting α -blockade to help minimize severe postoperative hypotension after resection of the tumor.¹⁸ This is achieved by increasing oral salt intake in the form of salt tablets for 3-to-5 days before surgery, which will allow for increased fluid intake and retention. Some patients with multiple comorbidities may benefit from a short preoperative hospital admission to optimize their intravascular volume status with isotonic intravenous fluids.¹⁴ Patients who have harbored a pheochromocytoma for a long time should undergo cardiac evaluation and echocardiography to rule out catecholamine-induced cardiomyopathy.

Intraoperative Planning and Management

Although no specific anesthetic protocol has been demonstrated to be superior in pheochromocytoma resection, it is clear that a comprehensive approach, starting preoperatively, has helped to smooth the overall perioperative course of these patients, resulting in very low overall morbidity and mortality.^{3,16} Even though the patient in this case had undergone extensive preoperative adrenergic blockade and metyrosine therapy to decrease tumor catecholamine synthesis, the large tumor size and very high preoperative catecholamine levels suggested that he was still at risk for significant intraoperative hypertension during tumor manipulation. Thus, a magnesium sulfate infusion was used to promote hemodynamic stability by preventing the reuptake of catecholamines and reducing peripheral α -adrenergic receptor sensitivity.¹⁹ For control of hypertension, nitroglycerin and clevidipine were selected because these drugs are titrated rapidly and have minimal residual activity once discontinued.^{20,21} The authors initially

planned to use only these vasodilators, but worsening intraoperative tachycardia with tumor manipulation led them to add esmolol to optimize hemodynamic control. The selection of short-acting antihypertensive medications is important because the transition from hypertension to hypotension occurs abruptly after ligation of the venous blood supply from the tumor and withdrawal of excess catecholamines.

In this patient, the typical hypotensive response to acute catecholamine withdrawal was compounded by venous outflow ligation from the tumor occurring during the period of IVC cross-clamp application. This created a challenging clinical scenario, with an acute decrease in ventricular preload followed by the withdrawal of intrinsic catecholamine secretion. With no evidence of IVC flow obstruction on TEE and the presence of a grossly normal lower extremity examination, it was presumed that extrahepatic venous collateral formation had not taken place.

Even though the typical hemodynamic response to the decrease in cardiac output associated with IVC cross-clamping is a compensatory increase in systemic vascular resistance (SVR), it was unclear whether this patient would be able to mount a satisfactory increase in SVR in light of his aggressive preoperative adrenergic blockade.^{22,23} Maintaining hemodynamic stability during this portion of the surgery required a multifaceted approach, starting during the preoperative phase. The initiation of preoperative salt loading with oral sodium chloride supplementation was undertaken to help promote fluid retention and avoid hypovolemia.²³ The patient then underwent early intraoperative fluid loading with colloid and crystalloid solutions to ensure adequate resuscitation before IVC cross-clamping. It was suspected that his preoperative adrenergic blockade combined with chronic exposure to high concentrations of circulating catecholamines would make him relatively insensate to vasoactive agents that act on these same receptors, such as epinephrine, norepinephrine, and phenylephrine.²¹ Thus, the authors chose to use vasopressin, which bypasses the adrenergic system to increase SVR through vasopressin-1 receptors, as the primary vasopressor after catecholamine withdrawal.^{21,24,25} Furthermore, continuous intraoperative TEE was used to allow for periodic assessment of volume status and to evaluate contractility after the loss of intrinsic catecholamine secretion. The risk of hemodynamic deterioration was believed to be substantial enough that preparations were made to emergently initiate VVB, if necessary. Finally, in the unlikely event of catastrophic embolization of the tumor to the pulmonary vasculature, the femoral venous cannula also would have allowed for emergency veno-arterial extracorporeal membrane oxygenation (VA-ECMO) support with the placement of an arterial cannula in the contralateral groin.

VVB is used most commonly in orthotopic liver transplantation in patients unable to tolerate IVC cross-clamping.²⁶ Even though its use has declined, it still remains a viable option for the maintenance of end-organ perfusion in patients with significant hemodynamic instability.²⁷ VVB historically has been established by pumping blood from a femoral venous cannula to an axillary cannula, but concerns about infection,

lymphatic injury, and nerve injury with axillary cannulation have led some authors to advocate for internal jugular cannulation.²⁸ Even though VVB may improve intraoperative hemodynamics, it is not without potential complication. Accidental removal of the cannula, thrombosis, and air embolism all are potentially significant complications of VVB. The risks of systemic anticoagulation also must be balanced with the risk of thromboembolism because heparinization also may increase surgical transfusion requirements.²⁷ Fortunately, this patient demonstrated a robust response to the intraoperative administration of vasopressin and was able to tolerate IVC cross-clamp application without requiring VVB.

VA-ECMO can be used to provide temporary hemodynamic support to patients with refractory hemodynamic instability after a broad array of physiologic insults.²⁹ VA-ECMO can be established either through direct central cannulation or peripheral cannulation of the femoral vessels. If this patient had experienced catastrophic embolization of the tumor to the pulmonary arteries, the placement of a femoral arterial cannula would have allowed for the rapid initiation of VA-ECMO to provide temporary support until definitive extraction of the embolic tumor was undertaken. This approach was described previously in patients with massive pulmonary embolism resulting in hemodynamic collapse with excellent results.^{30,31} Fortunately, early proximal control of the IVC before tumor extraction prevented embolization and made extracorporeal support unnecessary.

Postoperative Intensive Care Management

Perioperative rhabdomyolysis and myoglobinuric renal failure are uncommon, but described, complications of pheochromocytoma that significantly complicated this patient's postoperative course.^{32–36} In the perioperative period, massive catecholamine release during tumor manipulation leading to intense vasospasm and ischemia of the patient's muscle tissue has been implicated as the underlying etiology.³⁶ Interestingly, rhabdomyolysis can be the presenting symptom of pheochromocytoma, with case reports demonstrating profound rhabdomyolysis and myoglobinuric renal failure in patients undergoing medical or surgical treatment for unrelated disorders.^{33,36–38} In this patient, a prolonged surgical time of approximately 10 hours likely also contributed to the development of rhabdomyolysis. Even though IVC cross-clamping may cause rhabdomyolysis via a phlegmasia-type mechanism, it was deemed unlikely in this patient because he underwent a relatively brief period of IVC cross-clamp application and his lower extremities were grossly normal during the perioperative period.

Initially, this patient's hypoxic respiratory failure was believed to be due to pulmonary edema from aggressive volume resuscitation in the setting of AKI. Even though this was likely a major contributing factor given that he eventually responded favorably to diuresis, his persistent respiratory dysfunction appeared to extend beyond volume overload. He also developed VAP secondary to *H influenzae* with associated

acute respiratory distress syndrome, and he may have experienced either transfusion-associated circulatory overload (TACO) or transfusion-related acute lung injury in the setting of multiple intraoperative transfusions. Despite aggressive efforts to prevent their development, VAP and acute respiratory distress syndrome remain common within the first 96 hours of the perioperative period in mechanically ventilated patients.³⁹ It is likely that this patient may have developed VAP due to microaspiration of purulent sinus drainage given that the causative pathogen was the same in both sinus cultures and his bronchoalveolar lavage. Even though he had normal cardiac function, which typically argues against TACO, his robust improvement in response to diuretic administration favored TACO over transfusion-related acute lung injury as the primary transfusion-related pathology that contributed to his respiratory failure.⁴⁰

Conventional treatment of rhabdomyolysis with high-volume fluid resuscitation was particularly problematic given the patient's poor postoperative pulmonary status.⁴¹ In this case, renal protection was prioritized over optimizing his pulmonary function. He was treated with aggressive fluid resuscitation with lactated Ringer's solution and underwent diuresis with mannitol, with furosemide added to aid diuresis.⁴² Because adequate urine output (> 100 mL/h) was achieved with this strategy, urine alkalization was deferred. Urine alkalization with sodium bicarbonate typically is recommended when sufficient urine output is unable to be achieved, as previously described.⁴² Unfortunately, the use of sodium bicarbonate has not been shown to decrease the incidence of AKI or mortality in rhabdomyolysis.⁴³ Even though questions also remain about its efficacy, mannitol was selected for this patient due to its ability to increase renal blood flow and maintain renal filtration fraction and oxygenation.^{43,44} Veno-venous extracorporeal membrane oxygenation was considered briefly on POD 3 due to profound hypoxemia and refractory hypercarbia but ultimately was not necessary, given the rapid improvement in his respiratory status after diuresis was initiated. Fortunately, his rhabdomyolysis and renal failure resolved, allowing for rapid ventilator weaning, successful extubation, and subsequent recovery.

Commentary 1[†]

The authors of this case conference reported preoperative, intraoperative, and postoperative care of a 28-year-old patient undergoing removal of a malignant, secreting pheochromocytoma extended to the IVC. Vena cava cross-clamping was used intraoperatively and tolerated well, and VVB was not necessary. Preoperative care included, "as still recommended,"⁴⁵ volume loading and administration of several hypotensive drugs together with an inhibitor of tyrosine hydroxylase enzyme. Intraoperatively, 4 hypotensive drugs were administered to control acute episodes of arterial hypertension resulting from tumor manipulation. Arterial hypotension resulting from intraoperative vascular exclusion after tumor removal

was controlled using arginine vasopressin. Postoperatively, (1) major early respiratory failure required mechanical ventilation, and (2) rhabdomyolysis associated with transient renal failure was controlled using volume loading, diuretics, and mannitol. Fortunately, the patient recovered from this challenging medical misadventure. An analysis of available data likely to support the authors' therapeutic decisions is provided.

This clinical report provided the opportunity of broadening the debate on pheochromocytoma care. The author of this commentary believes that the time is ripe for a reappraisal of the available data regarding perioperative care of patients with pheochromocytoma.⁴⁵ Although a mortality rate of close to zero has been reported in all recent series of adrenalectomy for pheochromocytoma, irrespective of preoperative care,⁴⁵ this improved prognosis currently is attributed (1) to the widespread preoperative administration of vasodilating drugs, even to patients with normal arterial pressure in some series, and (2) to the administration of large fluid volumes, even to patients with normal or borderline elevation of catecholamine levels in other series.¹ None of these practices was evidence based.⁴⁵

Are Patients With Pheochromocytoma Hypovolemic?

The evidence does not support the concept of clinically significant hypovolemia associated with pheochromocytoma.^{46,47} As early as 1966, plasma volume was determined in 15 patients with pheochromocytoma before any therapy and in 10 of these patients after hypotensive drug administration or adrenalectomy, using human albumin labeled with ¹²⁵I.⁴⁶ All but 2 patients had normal predicted plasma volume at first assessment.⁴⁶ Recently, analysis of the arterial pressure waveform during mechanical ventilation was used to measure the negative component (D-down) of the respiratory systolic pressure variation in 13 patients undergoing pheochromocytoma surgery.⁴⁷ During tumor removal, the stability of D-down and the lack of correlation between systolic arterial pressure variation and D-down suggested that reduced preload associated with hypovolemia was not a major mechanism of hypotension after pheochromocytoma removal.⁴⁷ Furthermore, D-down values recorded before adrenal vein ligation in patients in whom arterial hypotension occurred after tumor removal overlapped those recorded in patients who remained normotensive.⁴⁷ This suggested that the predominant mechanism of severe hypotension after tumor resection likely is a decrease in arterial tone and that severe hypotension may occur in normovolemic patients.⁴⁷ This situation has been documented widely in the anesthetic literature in patients undergoing spinal and epidural anesthesia.⁴⁸ After a long period of intense fluid administration after central neural blockade using local anesthetics, current accepted recommendations focus on vasopressor administration.⁴⁸

Available published data clearly showed that regardless of whether patients have received fluid loading and/or regardless of the volume of fluid infused, intraoperative and postoperative courses did not differ in patients undergoing pheochromocytoma removal, as extensively reviewed.⁴⁵

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Does Preoperatively Increased Arterial Pressure Level Change Surgical Care in Patients Undergoing Pheochromocytoma Removal?

Plouin et al⁴⁹ reported that high preoperative arterial pressure was correlated independently with intraoperative and postoperative complications in patients undergoing pheochromocytoma removal. However, the emphasis in that article was on non-hypertension-related complications such as allergy; pneumothorax; postoperative bleeding or pancreatitis; intraoperative surgically related liver, kidney, lung, or bladder wounds; or a pulmonary artery catheter-related complication, which would not have been associated with increased arterial pressure. Recently, Lafont et al⁵⁰ reported perioperative data of normotensive patients with incidentally discovered pheochromocytomas.⁶ Most did not receive preoperative hypotensive drugs. These normotensive patients with incidentally discovered pheochromocytomas, who did not receive preoperative hypotensive drugs, were comparable with hypertensive pheochromocytoma patients in terms of hemodynamic instability during surgical resection.⁵⁰ Irrespective of preoperative arterial pressure level or preoperative drug administration, the mortality rate was zero in these series, similar to the most recent series.^{45,49,50}

Limited Number of Patients Scheduled for Pheochromocytoma Removal Must Receive Hypotensive Drugs Preoperatively

Aggressive medical control of arterial pressure with anti-hypertensive drugs is regarded as a major factor in accounting for the dramatic improvement in maternal and fetal outcome in patients with pheochromocytoma diagnosed during pregnancy, as reviewed.⁴⁵ Moreover, in general, it probably can be stated that the recommendation is for hypotensive drugs to be administered preoperatively, as is current medical practice, to patients with symptoms such as headache or tinnitus, and if systolic blood pressure is > 180 mmHg and diastolic blood pressure is > 115 mmHg.⁷ Indeed, these situations expose patients to an early risk in daily life.^{45,51} On the other hand, recommendations issued from studies conducted in vascular and general surgery stated that increased risk of surgical mortality was reported only if preoperative arterial hypertension coexisted with preexisting heart disease, including coronary artery disease; factors likely to favor coronary artery disease such as hyperlipidemia or diabetes; and symptomatic left ventricular dilation or hypertrophy or aortic stenosis.⁵¹

Unfortunately, the patient's arterial pressure levels were not reported in the text by Gregory et al, but nausea and vomiting are consistent with an increased untolerated arterial pressure, thus warranting preoperative hypotensive drug administration.⁵¹

Does Hypotensive Drug Administration Change Surgical Care of Patients Undergoing Adrenalectomy for Pheochromocytoma, Irrespective of Their Hypotensive Effect?

Boutros et al⁸ compared 31 patients receiving phenoxybenzamine or prazosin with 29 patients who did not receive any

hypotensive drugs. Preoperative blood pressures remained increased in most patients examined.⁸ No differences in perioperative morbidity or mortality were found between the groups, although the rise in intraoperative blood pressure was a little higher in the untreated group.⁸

Does Anesthetic Technique Matter? Is Epidural Anesthesia Contributive?

General anesthesia is favored in most reports, as reviewed.⁴⁵ No anesthetic protocol has been demonstrated to be better than the others.⁴⁵ Direct and continuous monitoring of arterial pressure via an intra-arterial catheter, allowing for immediate arterial pressure control, is recommended universally.⁴⁵ Epidural and spinal anesthesia have been used alone or as a complement to general anesthesia for pheochromocytoma removal.⁴⁵ However, sympathetic blockade associated with spinal anesthesia did not protect against the hemodynamic effects of catecholamine release during tumor manipulation.⁴⁵

With respect to the reported case reported here, irrespective of the expected postoperative analgesic benefit, the question arises as to whether abdominal pressure changes resulting from patient positioning, necessary for implementing epidural anesthesia, are likely to cause both catecholamine release and life-threatening arterial hypertension in patients with secreting pheochromocytoma.⁵² Moreover, in the case presented here, coagulopathy that could have resulted from both possible massive bleeding and transfusion could have been harmful after epidural puncture.⁵³

Aforementioned Findings Are Supported by the Pathophysiology of Pheochromocytoma

In patients undergoing pheochromocytoma removal, intraoperative increase in arterial pressure has been shown to be related directly to plasma norepinephrine and epinephrine release, which occurs (1) when moving the patient onto the table; (2) during induction of general anesthesia and tracheal intubation combined with intraabdominal pressure increases resulting from cough, light sedation, insufficient relaxation, and initiation of mechanical ventilation; (3) during the creation of pneumoperitoneum; and (4) when the tumor is manipulated directly.⁵² It is the opinion of the author of this commentary that none of these situations is likely to be balanced by hypotensive drug administration.⁵²

The so-called "improved preoperative preparation" in the case presented here should be changed to a more global concept, such as "intraoperative attention to patients' hemodynamics likely is to be more important than a specific hypotensive medication or unnecessary volume loading."

Is This Issue Adequately Addressed in Available Data?

Standard guidelines supporting the dogma of preoperative hypotensive drug administration and volume loading in patients with pheochromocytoma are derived from questionable data published in retrospective studies, sometimes

conducted over much more than 10 years, that did not include any control groups.^{8,45,47,49,50,52,54} However, as early as 1977, when comparing 2 personal historic studies of patients undergoing adrenalectomy for pheochromocytoma, Desmonts and Marty⁵⁴ concluded that administering fluid only intraoperatively, whenever necessary, was as efficient as preoperative fluid infusion in regard to hemodynamic control.⁵⁴ They also found that preoperative α -blockade was not necessary because available vasoactive drugs allowed intraoperative hemodynamic changes to be controlled immediately whenever necessary.⁵⁴

Many fast-acting intravenous hypotensive and hypertensive drug regimens indeed are available currently and have been reported to control intraoperative and postoperative changes in arterial pressure efficiently and consistently, resulting in a mortality rate close to zero in all recent series of pheochromocytoma removals, irrespective of preoperative care.⁴⁵ A critical analysis of the available data did not uphold the authors' contention that "patients with pheochromocytoma typically experience significant volume contraction and thus require preoperative intravascular volume resuscitation after starting α blockade to help minimize severe postoperative hypotension after resection of the tumor."

In this respect, drug selection mainly results from local practices. With regard to the text by Gregory et al, it is of note that in contrast to calcium inhibitors and alpha- and beta-blockers, magnesium sulfate, which was administered to the reported patient, did not reduce intraoperative arterial pressure consistently in patients undergoing pheochromocytoma removal.⁵⁵ Furthermore, the dramatic intraoperative association of the 4 drugs administered to the patient described in this report was not, to this author's knowledge, reported in any series. Also, the author of this commentary is unaware of the resistance to norepinephrine ever being reported in patients with pheochromocytoma, provided that an adequate dose is administered. Arginine vasopressin administration only has been recommended to treat refractory shock in intensive care units.⁵⁶

Specific Comments on the Reported Respiratory Failure Are Warranted

It is the opinion of the author of this commentary that the diagnosis of respiratory failure resulting from circulatory failure is questionable because of the following: (1) respiratory failure was associated with an early recorded hypercarbia, which is unusual in patients with hemodynamic pulmonary edema, except if massive alveolar flooding is recorded⁵⁷; (2) hypercarbia persisted further under mechanical ventilation, which is an additional unusual factor in the case of hemodynamic pulmonary edema⁵⁷; (3) this adverse event was recorded in a young patient with a preoperative and intraoperative healthy heart and euvoemia recorded at the end of the procedure, as assessed using TEE; and (4) respiratory failure occurred after intraoperative hemorrhage, massive transfusion, and hypertensive crisis had been tolerated as assessed using TEE, thus confirming the diagnosis of a healthy heart unlikely

to account for the respiratory failure.⁵⁷ In addition, the diagnosis of cardiogenic pulmonary edema cannot be based on standard chest x-rays.⁵⁷ Also, to support the diagnosis of late circulatory failure, a late additional TEE assessment is lacking, which could have been carried out at the bedside and astonishingly was not done.⁵⁷ A basic measurement, namely "pro-brain natriuretic peptide measurement," also is lacking, which could have supported the diagnosis of heart failure.⁵⁷ The primary assumption of this commentary's author would have been "too-early extubation in a patient with extended atelectasis related to a long surgical procedure."⁵⁸

Because a chest x-ray was reported to show pulmonary edema, at least the following 3 additional diagnoses should have been discussed: (1) nonspecific respiratory distress syndrome,¹⁴ (2) lung injury directly related to blood transfusion,⁵⁹ and (3) Takotsubo-like myocardial injury resulting from intraoperative catecholamine release.³⁰ In like manner to that previously indicated, prolonged hypercarbia made the diagnosis of Takotsubo-like cardiomyopathy very unlikely.⁶⁰ Contributive TEE and biologic assessments were lacking to properly discuss the latter possible diagnosis.^{57,60}

Conclusion

This report could have offered the opportunity of broadening the debate on the perioperative care of patients undergoing pheochromocytoma removal and on postoperative respiratory failure. Indeed, it is possible presently to regard preoperative and intraoperative care of patients with pheochromocytoma differently.⁴⁵ This timely reappraisal suggests that preoperative hypotensive treatment of patients with life-threatening complications of high blood pressure, such as congestive heart failure, stroke, coronary artery disease, arrhythmia, and blood pressure > 180/115 mmHg, and patients with pheochromocytoma diagnosed during pregnancy is warranted.^{45,51} In contrast, systematic preoperative administration of fluids and hypotensive drugs is not supported by any evidence-based study.⁴⁵ On the other hand, postoperative hemodynamic assessment could have enabled postoperative respiratory failure to be studied further.^{57–60} However, adequate symptomatic treatment of adverse preoperative, intraoperative, and postoperative symptoms by a committed team ready to implement sophisticated circulatory support resulted in a fortunate outcome.

Commentary 2*

This case conference described an interesting scenario in which a malignant pheochromocytoma with vascular extension was resected successfully in a complex and multidisciplinary fashion. Unlike a typical pheochromocytoma that can be managed laparoscopically, this extensive malignant tumor involved the IVC, necessitating an extensive laparotomy for adrenalectomy, IVC thrombectomy, and subsequent IVC reconstruction. In this commentary, the authors focused on

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the following 4 aspects in the management of malignant pheochromocytoma with local extension: the diagnostic approach, the pharmacologic approach, the contributions of echocardiography, and the role of extracorporeal circulation in the setting of major vascular extension.

Diagnostic Approach to Pheochromocytoma

Pheochromocytomas are catecholamine-secreting tumors that arise primarily from the chromaffin cells of the adrenal medulla.^{20,61} Less commonly, extra-adrenal pheochromocytomas (paragangliomas) can arise from sympathetic ganglia.^{20–62} The annual incidence of these tumors is approximately 2-to-8 cases per 1 million, with 80% to 85% being pheochromocytomas and the rest paragangliomas.^{2,63} Pheochromocytomas can metastasize in about 10% of patients, invading the liver and beyond.^{2,63} Factors associated with tumor malignancy in this setting include large tumor size and genetic susceptibilities.^{2,62,63} Although the diagnosis of pheochromocytoma typically is based on classic clinical signs and symptoms that develop secondary to catecholamine release, there is a wide range of clinical presentations because this catecholamine production is highly variable.^{2,62} In fact, the initial clinical presentation of pheochromocytoma can be as a perioperative hypertensive crisis.⁶⁴

Suspicion of a pheochromocytoma should prompt measurement of catecholamine levels, such as total metanephrines in the urine, as was done in this case conference (Fig 4).^{20–62} Metanephrine and normetanephrine represent breakdown products of epinephrine and norepinephrine, respectively.⁶² Consequently, this tumor was secreting large amounts of epinephrine and norepinephrine, which is a typical catecholamine profile for pheochromocytomas at clinical diagnosis.^{2,62} The measurement of total metanephrine levels in the plasma and urine is accurate not only for screening but also for diagnosis, with a reported sensitivity and specificity of more than 90%.^{2,62,63} The prolific production of this pheochromocytoma resulted in ready detection of catecholamines, obviating the requirement for further diagnostic maneuvers such as a 24-hour urine metanephrine determination and/or the

performance of a clonidine suppression test.^{20,62,65} The failure of an adequate clonidine dose to reduce levels of epinephrine or norepinephrine by at least 50% is highly sensitive and specific for pheochromocytoma.⁶⁵ Although testing for metanephrines is a diagnostic technique of choice in suspected pheochromocytoma, it is important to remember that multiple factors still can interfere with this technique, such as age, sex, diet, medications, body position, cigarette smoking, and comorbidities.^{2,62–65} In challenging diagnostic cases, replacement of disruptive medications may be warranted to optimize the precision of the biochemical testing.^{3,62–65} The biochemical diagnosis confirms the diagnosis of pheochromocytoma, rendering a tissue diagnosis from percutaneous adrenal biopsy unnecessary.⁶⁶ Given that many adrenal masses may not have diagnostic biochemical markers, adrenal biopsy in these settings often is required for definitive diagnosis.⁶⁶

Once the diagnosis of pheochromocytoma has been reached, detailed imaging is required to determine tumor location and the local and distal extent. The imaging options include computed tomography, MRI, MIBG scanning, and positron emission tomography.^{2,62,63} In this patient, detailed MRI demonstrated not only the right adrenal origin of this tumor but also staged its significant vascular extension into the infrahepatic IVC. In the assessment of vascular extension by pheochromocytoma, MRI typically is superior to computed tomography.^{2,62,63} The clinical contribution of MIBG scanning in this setting is to detect extra-adrenal tumor burden, which was especially important in this patient due to the malignant features of the tumor.^{2,62,63} The MIBG tracer is concentrated in the tumor due to its avid uptake of precursor amines required for catecholamine synthesis.⁶² Although MIBG scanning requires 24 to 48 hours, it has a high reported specificity of 95% to 100%.^{2,62,63} In this patient, MIBG scanning confirmed the absence of distal metastasis, suggesting that radical surgical resection might be curative in this young man. Furthermore, positron emission tomography scanning with a tracer such as fluorodopamine had a limited role here because its main indication is for tumor localization when biochemical testing is diagnostic but conventional imaging detects no anatomic catecholamine source.^{2,62,63} In this patient, the tumor was massive both anatomically and biochemically. The options for biochemical control of catecholamine production and their systemic effects in this challenging clinical presentation are the subject of the next section.

Pharmacologic Approach in Pheochromocytoma

Once the pheochromocytoma has been diagnosed, characterized, and staged, the next step is to achieve catecholamine control for symptom relief and preoperative stabilization.⁶⁷ There are 2 major pharmacologic choices in this setting: suppression of catecholamine production with metyrosine and peripheral catecholamine blockade with alpha- and beta-blockers (Fig 4).^{2,62,63} Given the catecholamine burden in this patient, it is no surprise that both choices were used. Metyrosine is a competitive inhibitor of the enzyme tyrosine hydroxylase, the rate-limiting step in catecholamine synthesis

Fig 4. The synthesis and metabolism of catecholamines. (From Kinney et al²⁰.)

(Fig 4).^{2,62,63} As a result, titrated therapy ultimately decreases tumor catecholamine stores so that the risk of a catecholamine crisis is reduced substantially, especially in the setting of massive tumors that are metabolically active.^{2,62,63} A recent clinical trial (n = 174) demonstrated that the absence of metyrosine from the preoperative medical regimen in planned surgical resection of pheochromocytoma was significantly associated with a greater risk for intraoperative hemodynamic instability (for heart rate $p = 0.034$; for systolic blood pressure $p = 0.020$) and cardiovascular complications (absolute increase 15.8%, $p = 0.034$).⁶⁸ The investigators concluded that suppression of catecholamine synthesis with metyrosine likely improved perioperative outcomes in this setting.⁶⁸ The major limitation with this agent is the side effect profile, including extrapyramidal signs due to acute dopamine deficiency.^{2,62} In this patient, therapy with metyrosine was deliberately acute so as to maximize catecholamine suppression and minimize side effects just before the surgical resection.

Because most tumors produce catecholamines with alpha-adrenergic effects, such as norepinephrine, α -blockade has become a mainstay of peripheral catecholamine blockade.^{2,62} Although the long-acting nonselective phenoxybenzamine has been the classic pharmacologic approach, its clinical niche recently has been challenged by the shorter-acting α -1 specific agents such as doxazosin, prazosin, and terazosin.^{2,62–65,69} Recent clinical trials have demonstrated that these selective agents are less protective against intraoperative hypertension than is phenoxybenzamine.⁷⁰ On the other hand, their shorter half-lives likely are protective against persistent hypotension after tumor removal.^{2,20,62} The current evidence base does not provide enough guidance about which α -blockade approach is superior but rather highlights the advantages and disadvantages of each approach. Regardless of the selected alpha-blocking agent, it should be titrated precisely to achieve optimal catecholamine control in time for surgery, as outlined by the classic Roizen criteria.⁷¹ The titration of beta-blockade in this patient was essential, given the generous production of epinephrine by this large tumor. Additional roles for magnesium and calcium channel blockade in this setting have been highlighted in the *Journal*, including the newer short-acting clevidipine.²¹

As highlighted in this case conference, the pharmacologic approach shifted dramatically with the clamping of the tumor venous supply because this begins the period of acute catecholamine withdrawal after massive chronic excess. The severe vasoplegia from acute catecholopenia in this patient was mitigated by the following maneuvers: a shorter-acting α -1 blocker in the preoperative period, short-acting intraoperative vasodilators before tumor removal, and titrated vasopressin after tumor removal. The rationale and roles for vasopressin in this setting also have been reviewed recently in the *Journal*.²¹

Echocardiography in the Management of Pheochromocytoma

Pheochromocytoma can affect the heart in many ways, including the consequences of severe hypertension, acute type-

A aortic dissection, and catecholamine cardiomyopathy.²⁰ A recent analysis of the cardiac phenotype in patients with pheochromocytoma using cardiac MRI demonstrated that cardiac abnormalities such as myocarditis, fibrosis, and myocardial dysfunction are common.⁶⁹ The detected fibrosis was at times focal and at times diffuse.¹⁴ The detected myocardial dysfunction was not only both systolic and diastolic, but did not always readily resolve after tumor removal.⁶⁹ Furthermore, it should be noted that the cardiac phenotype in pheochromocytoma frequently can resemble Takotsubo cardiomyopathy with left ventricular regional wall motion abnormalities.⁷² Although at times similar to Takotsubo cardiomyopathy, cardiomyopathy in pheochromocytoma is excluded specifically in the Mayo Clinic criteria for Takotsubo cardiomyopathy.⁷² These aspects have been discussed recently in the *Journal* in great detail.⁷² Given the frequency of cardiac involvement by pheochromocytoma, a baseline transthoracic echocardiogram on presentation will detect major abnormalities readily, which can inform management decisions.^{2,20,62,63}

In this case conference, the patient did not appear to have clinical cardiac involvement in the preoperative phase. TEE was selected for intraoperative assessment of cardiac function, intravascular volume status, and vascular spread by the tumor. As stated in the presented case, there is no specific perioperative protocol that has been demonstrated to be superior for management of pheochromocytomas. Specifically, the role for TEE has to be evaluated on a case-by-case basis. During resection of massive pheochromocytomas, there may be tremendous swings in hemodynamics with tumor manipulation and excision. Furthermore, in this case of malignant pheochromocytoma, there was significant vascular extension into the IVC with risk for pulmonary embolization. The possibility of pulmonary embolization highlighted the risk for acute cardiovascular collapse. In all these hemodynamic scenarios, expert comprehensive TEE could have provided immediate anatomic and hemodynamic data to guide intraoperative anesthetic and surgical decision-making beyond the capabilities of a pulmonary arterial catheter.^{73,74} The choice of TEE in this patient was in full agreement with the recent TEE guidelines from the American Society of Anesthesiologists, Society of Cardiovascular Anesthesiologists, and the American Society of Echocardiography, given the outlined cardiovascular risks.^{73,74}

The limitations of central pressure monitoring have been well established.^{73–75} Furthermore, Sandham et al⁷⁵ demonstrated no survival benefit for placing a pulmonary arterial catheter for goal-directed therapy in high-risk surgical patients. A meta-analysis (n = 5,051, 13 randomized controlled trials) showed no benefit in mortality, number of days hospitalized, inotrope exposure, or vasodilator therapy when using a pulmonary arterial catheter for critically ill patients in the ICU.⁷⁶ Direct cardiac imaging using TEE frequently can bypass these limitations.^{73,74}

Several studies suggested TEE may be beneficial in non-cardiac surgical patients with hemodynamic instability, explaining its strong endorsement for this setting in recent guidelines.^{77–79} These clinical studies implied that TEE may

be a more sensitive tool, not only for quickly and accurately diagnosing the etiology of hemodynamic instability, but also for providing a means to monitor the effects of early intervention.^{77–79}

Although TEE offers major value in this setting, it is accompanied by several disadvantages. First, an experienced clinician trained and certified in TEE is required to guide the performance and interpretation of the comprehensive examination.⁷⁴ Second, although uncommon, there are a number of contraindications to TEE, such as major esophageal pathology.⁷⁴ Third, there also is significant cost associated with the purchase and maintenance of the echocardiography hardware. Overall, TEE can be performed rapidly to provide qualitative and quantitative data in real time to guide intraoperative decision-making at all stages of the procedure, as illustrated so clearly in this complex case.

Extracorporeal Circulation in the Management of Pheochromocytoma

The presence of an IVC tumor thrombus prompted the authors of the case report to cannulate the patient for possible extracorporeal circulation (ECC) in the form of VVB to facilitate control of the IVC and/or CPB/ECMO for cardiopulmonary stabilization in case of massive pulmonary embolization of tumor.⁸⁰ The VVB arrangement is achieved by cannulation caudal to the renal veins for drainage and cannulation above the SVC for return of the bypassed blood, as illustrated in this patient. The role of VVB is well documented for resection of tumors complicated with IVC thrombus.^{81,82} The advantages of VVB in this setting include avoiding significant decreases in preload during IVC clamping, reduction of blood loss, and improved surgical exposure.⁸¹

The indication for possible VVB in this case conference was that the caval spread of tumor had reached the liver. The caval spread of tumor thrombi traditionally has been classified into 4 levels.⁸¹ This classification is important to understand because it provides a platform for understanding the surgical approach to resection and the roles of ECC. Level I is defined as tumor thrombus that is confined to the renal vein. Level II is defined as tumor thrombus that is confined to the infrahepatic IVC.⁸¹ Levels I and II tumor thrombi typically are resected via a cavotomy without ECC.⁸¹

Level III is defined as tumor thrombus that has reached the retrohepatic IVC or the suprahepatic IVC without reaching the right atrium.⁸¹ Level III tumor thrombi are further subdivided into 4 types. Level IIIa (intrahepatic) thrombus, as in this patient, extends into the retrohepatic IVC but terminates below the major hepatic veins. Level IIIb (hepatic) thrombus extends into the retrohepatic IVC to reach the ostia of the major hepatic veins; it may cause venous occlusion and the Budd-Chiari syndrome.⁸¹ Level IIIc thrombus (suprahepatic but infradiaphragmatic) extends above the hepatic veins in the retrohepatic IVC but terminates below the level of the diaphragm. Level IIId (suprahepatic but infracardiac) thrombus extends into the IVC above the hepatic veins and

diaphragm into the intrapericardial IVC but not into the right atrium. Level IV thrombus (cardiac) extends into the right atrium.⁸¹

In this patient, level IIIa tumor thrombus was resected without VVB, although it was available readily. Test clamping of the IVC was tolerated, and the surgical team also was prepared to limit bleeding with the Pringle maneuver. Levels IIIb, IIIc, IIId, and IV tumor thrombi typically require ECC (VVB or CPB) to permit complete extraction in a relatively bloodless field. In selected patients, CPB with deep hypothermic circulatory arrest may be required to achieve complete removal of tumor thrombus.^{81,82} Although comprehensive preoperative imaging facilitates the surgical plan, intraoperative TEE may play an integral role in guiding the decision not only to initiate ECC but also which type (VVB or CPB).

The advantages of VVB over CPB include a lesser degree of systemic anticoagulation, no requirement for hypothermia, and a lesser systemic inflammatory response.⁸¹ In a large series comparing VVB with CPB for IVC tumor thrombectomy, 41 patients were reviewed, 13 of which underwent VVB and 28 underwent CPB.⁸² In that analysis, Granberg et al⁸² demonstrated that VVB compared with CPB significantly reduced anesthetic time, surgical time, bypass time, median blood loss, transfusion burden, and length of hospitalization.

Although VVB may be superior to CPB in this setting, the risks of percutaneous cannulation for VVB can be serious.^{83,84} A case series of 320 patients who underwent right internal jugular vein cannulation reported a 2.2% complication rate, including hemomediastinum, air embolism, low flows, hypotension, and atrial fibrillation.⁸³ A second case series of 312 patients reported a 1.6% complication rate, including hemothorax and 1 cardiac arrest leading to death.⁸⁴ Although there is a low complication rate, it remains essential to use ECC (VVB or CPB) only when necessary. In this patient, the level IIIa extent of the tumor thrombus raised the significant possibility of VVB that was planned for during the intraoperative phase. As outlined, the resection was completed without VVB because test clamping of the IVC was tolerated hemodynamically. Furthermore, the resection was completed without CPB or ECMO because there was no cardiopulmonary embolization of tumor. This multidisciplinary team had integrated the roles of ECC (both VVB and CPB) into the conduct of this staged procedure for hemodynamic rescue as needed.

Conclusion

This presented case of malignant pheochromocytoma with vascular extension is a rare occurrence. The authors are to be congratulated for their multidisciplinary and systematic approach to this complex case during all phases that resulted in a successful outcome, despite a challenging perioperative course. The expert integration of anesthetic management into this multidisciplinary intervention is readily evident throughout the perioperative period.

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