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CASE REPORTS

The Blood Pressure Paradox in Familial Pheochromocytoma: A Case Report of General Anesthesia Management in Oral Surgery Following Bilateral Adrenalectomy

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Pheochromocytoma is an endocrine tumor characterized by abnormal catecholamine production. We report the anesthetic management of a 47-year-old woman with a history of familial bilateral pheochromocytoma and prior bilateral adrenalectomy, who underwent cyst enucleation under general anesthesia. The patient was on maintenance corticosteroid therapy and presented with suspected local recurrence. Given her adrenal insufficiency and history of hypertension, perioperative care required meticulous hemodynamic monitoring, stress-dose steroid supplementation, and avoidance of exogenous epinephrine. Anesthesia was induced with sevoflurane and remifentanyl, and intraoperative hypotension was successfully managed with ephedrine. Phentolamine was prepared in anticipation of potential catecholaminergic crises. The procedure proceeded uneventfully with stable hemodynamics and no complications. This case underscores the unique anesthetic challenges in managing patients with coexisting adrenal insufficiency and the risk of catecholamine excess. Careful perioperative planning is essential to ensure safety and hemodynamic stability.

Introduction

We report a case of safe general anesthesia management in a patient with a history of familial pheochromocytoma who had undergone bilateral adrenalectomy, resulting in the absence of endogenous adrenocortical and adrenomedullary hormone production. Through meticulous perioperative care-including stress-dose corticosteroid supplementation and appropriate circulatory management, anesthesia was conducted safely and without complications. Informed consent for publication was obtained from the patient both verbally and in writing.

Case presentation

The patient was a 47-year-old woman, 160 cm in height and weighing 85.4 kg (BMI 33.2), scheduled to undergo cyst enucleation of the jaw under general anesthesia. Her medical history was significant for familial bilateral pheochromocytoma. She had undergone left adrenalectomy at the age of 21, followed by right adrenalectomy and resection of a perihilar tumor at the age of 34. Since then, she had been under regular follow-up with the

urology department every three months. At the age of 45, a suspected local recurrence was identified on CT imaging, showing a slowly enlarging lesion at the site of the previous right adrenal resection. A 24-hour urine catecholamine test revealed mildly elevated norepinephrine at 426 μg/day (reference: 31–160 μg/day), with epinephrine at 16 μg/day (3.0–41.0 μg/day) and dopamine at 1037 μg/day (280–1100 μg/day). Her other past medical history included hypertension, with blood pressure ranging from 126–141/74–85 mmHg. She was on maintenance steroid therapy with prednisolone 5 mg and hydrocortisone 10 mg daily. Preoperative laboratory results were within normal limits. To prevent intraoperative hypotensive episodes, oral doxazosin mesylate was initiated one month prior to surgery and titrated to 16 mg/day by the time of the procedure. The interincisal distance was three fingerbreadths, with a Mallampati score of II. There was no limitation in neck mobility, and no predictors of difficult airway were identified. The patient was classified as American Society of Anesthesiologists (ASA) physical status class II. Written informed consent was obtained from the patient.

General anesthesia was induced using fentanyl 50 μg, remifentanil at 0.3 μg/kg/min, propofol 120 mg, oxygen at 6 L/min, and sevoflurane at 2%. Following loss of consciousness, rocuronium 60 mg was administered, and endotracheal intubation was performed orally with a 7.0-mm spiral tube (Shiley™, Covidien Japan, Tokyo). Anesthesia was maintained with air at 2 L/min, oxygen at 1 L/min, sevoflurane at 2%, and remifentanil at 0.1 μg/kg/min, using volume-controlled ventilation (tidal volume 8 mL/kg, respiratory rate 10/min). The FiO_2 was set at 0.4. Arterial blood gas analysis revealed pH 7.37, PaO_2 84 mmHg, $PaCO_2$ 44 mmHg, SaO_2 97.2%, HCO_3^- 25.4 mEq/L, BE −0.1 mEq/L, tHb 10.6 g/dL, Na^+ 136 mmol/L, K^+ 4.1 mmol/L, Ca^{2+} 1.18 mmol/L, and glucose 104 mg/dL.

Although the patient had undergone bilateral adrenalectomy, imaging suggested a local recurrence with gradual progression and mildly elevated urinary norepinephrine. In consultation with the oral surgery team, epinephrine-containing local anesthetics were avoided, and 5.4 mL of epinephrine-free dental mepivacaine was administered. Considering the potential for pheochromocytoma recurrence, phentolamine was also prepared for intraoperative hypertensive crises. Intraoperatively, blood pressure remained stable around 105/55 mmHg, and dropped to 75/45 mmHg during minimal surgical stimulation, which was corrected with 4–8 mg boluses of ephedrine, maintaining BP at 85–90/50–55 mmHg.

At the end of surgery, oxygen was administered at 6 L/min and other anesthetics were discontinued. The TOF count showed 4 twitches with a T4/T1 ratio of 80%, prompting administration of 200 mg sugammadex for reversal. Emergence was smooth, with appropriate response and limb movement confirmed prior to extubation. After extubation, FiO_2 was set at 0.4, and repeat arterial blood gas analysis showed: pH 7.32, PaO_2 159 mmHg,

$PaCO_2$ 49 mmHg, SaO_2 97.9%, HCO_3^- 25.2 mEq/L, BE −1.2 mEq/L, tHb 10.6 g/dL, Na^+ 137 mmol/L, K^+ 4.1 mmol/L, Ca^{2+} 1.17 mmol/L, and glucose 107 mg/dL, without clinically concerning abnormalities. Intraoperative urine output was 125 mL, and blood loss was 15 mL. For postoperative analgesia, an additional 1.8 mL of mepivacaine was administered, followed by 50 mg flurbiprofen axetil IV. The patient reported no PONV and experienced manageable postoperative pain. Surgical duration was 59 minutes, and anesthesia time was 1 hour 41 minutes. Upon discharge from the operating room, vital signs were stable: BP 118/47 mmHg, HR 89 bpm, RR 12/min, SpO_2 98% (on 3 L/min oxygen).

Discussion

Pheochromocytoma is an endocrine tumor characterized by excessive catecholamine secretion. Sympathetic stimulation induces abnormal secretion of norepinephrine and epinephrine, which can lead to sudden hypertension and tachycardia, making perioperative hemodynamic management challenging. In the present case, the major concerns were the risk of severe hypotension or circulatory collapse due to general anesthesia following bilateral adrenalectomy, and conversely, the possibility of a hypertensive crisis triggered by surgical stress or epinephrine use, due to residual or recurrent pheochromocytoma.

In general, if one adrenal gland remains functional, hormonal secretion can be compensated. However, this patient had undergone bilateral adrenalectomy, eliminating endogenous secretion of both adrenocortical and adrenomedullary hormones. The adrenal glands consist of two anatomically and functionally distinct regions: the cortex and the medulla.[1] The adrenal cortex secretes glucocorticoids and mineralocorticoids.[2] Glucocorticoids play essential roles in the perioperative period, including increasing blood glucose levels via hepatic gluconeogenesis, suppressing inflammatory responses, and mediating the stress response. Mineralocorticoids regulate fluid volume and blood pressure by promoting sodium reabsorption and potassium excretion in the renal distal tubules.[3] The adrenal medulla secretes catecholamines, including epinephrine, norepinephrine, and dopamine, with epinephrine being the predominant component.[2] Epinephrine increases heart rate, myocardial contractility, and blood glucose, thereby contributing to the acute stress response. Norepinephrine causes vasoconstriction, leading to increased blood pressure. Bilateral adrenalectomy eliminates endogenous cortisol and aldosterone secretion, increasing the risk of adrenal crisis, hypoglycemia, infection, hypotension, and hemodynamic instability.

Physiologically, 10–20 mg of cortisol is secreted daily, which is equivalent to 2.5–5 mg of prednisolone. Patients receiving ≥20 mg/day of prednisolone (or equivalent) for more than 3 weeks within the past year are considered to have suppressed adrenal function and require perioperative steroid coverage.[4, 5] During surgical or anesthetic stress, cortisol secretion normally increases;

however, in cases of adrenal insufficiency, this compensatory mechanism is impaired, potentially leading to acute adrenal crisis. Therefore, steroid coverage was deemed necessary in this case. While there is no high-quality evidence supporting a standardized approach to steroid supplementation, various regimens have been proposed using different types and doses of corticosteroids. Some studies have reported no difference in postoperative complications regardless of whether steroid coverage was administered.[6] Supplementation should be tailored to the degree of surgical stress. In this minor surgical case, intravenous administration of 25 mg hydrocortisone or 6–8 mg dexamethasone is recommended.[4,5,7] Inadequate steroid coverage may result in hypotension and poor responsiveness to vasopressors, whereas excessive supplementation can cause immunosuppression, delayed wound healing, and poor glycemic control. In this case, the patient's routine dose of 5 mg prednisolone (equivalent to 20 mg hydrocortisone), was supplemented with 10 mg hydrocortisone preoperatively and 6.6 mg dexamethasone intraoperatively (equivalent to 150–200 mg hydrocortisone).[5] No abnormalities in blood glucose or temperature were observed postoperatively, and wound healing was uneventful.

In this patient, a suspected local recurrence had been noted on CT imaging. The lesion in the right adrenalectomy bed had shown gradual enlargement, and urinary norepinephrine levels were mildly elevated, suggesting possible residual or recurrent pheochromocytoma. Although the surgical field was intraoral, unexpected catecholamine surges could be induced by body movement or abdominal pressure during general anesthesia, such as during patient transfer or surgical assistance.[8-10] In addition to endogenous catecholamine production from surgical stress, exogenous epinephrine use could provoke a hypertensive crisis due to catecholamine hypersecretion from residual tumor tissue. Since the patient had comorbid hypertension and was receiving 16 mg of doxazosin mesylate, a nonselective α-blocker, epinephrine-containing local anesthetics were avoided in consultation with oral surgeons, and epinephrine-free mepivacaine was used instead.

A balanced general anesthesia using inhalational agents was selected for this case. Although few randomized controlled trials have demonstrated clear superiority of one technique over another, sevoflurane has been widely used in pheochromocytoma resection and is associated with stable hemodynamics, lower airway irritability, and reduced arrhythmogenic potential compared to desflurane.[11] Furthermore, a study comparing total intravenous anesthesia (TIVA) and balanced anesthesia in 172 patients with pheochromocytoma reported a higher incidence of intraoperative hypertensive events in the TIVA group.[12] Based on these findings, we considered inhalational anesthesia to be the safer choice and opted for balanced anesthesia rather than TIVA.

Although local recurrence in familial pheochromocytoma is rare, reported recurrence rates vary, with Harrison et al. citing a recurrence rate of 7.1%.[13] Fatal hypertensive crises during general anesthesia have also been reported in patients with pheochromocytoma.[14] Therefore, it is critically important to minimize pain and stress during surgery and ensure meticulous perioperative hemodynamic monitoring and management.

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