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Anesthetic Management of a Patient with Von Hippel-Lindau Syndrome: A Case Report

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Von Hippel-Lindau syndrome, anesthetic management, perioperative care, catecholamine, surgical intervention.

ABSTRACT:
Von Hippel-Lindau (VHL) syndrome poses significant challenges in anesthetic management due to its diverse clinical manifestations, including vascular tumors and endocrine abnormalities. We present a case of a 25-year-old male with VHL syndrome undergoing pancreatic lesion and right adrenal mass resection. Preoperative optimization involves α and β blockers to manage potential catecholamine-induced hypertensive crises. Anesthesia induction comprised of fentanyl, propofol, and vecuronium, with lignocaine to blunt intubation response. Maintenance of anesthesia with oxygen-nitrous oxide-isoflurane with an activated epidural infusion. Intraoperative monitoring focused on detecting catecholamine surges, with vasodilators and vasopressors administration as required. Postoperative care emphasizes the monitoring of persistent hypertension and hemodynamic instability. The patient's perioperative course was uncomplicated, highlighting the importance of meticulous preoperative assessment and intraoperative vigilance. This case underscores the need for tailored anesthesia strategies and multidisciplinary collaboration to optimize outcomes in VHL syndrome surgical interventions, addressing the unique challenges posed by this complex condition.

1. INTRODUCTION

Von Hippel Lindau disease is a hereditary autosomal dominant disorder characterized by a predisposition to develop distinctive tumors in the retina, brain stem, spinal cord, and cerebellar hemispheres (capillary hemangioblastomas) [1]. About 60% of patients with Von Hippel Lindau disease have cerebellar tumors, while 14–50% have brain stem tumors. Surgical removal is performed for accessible lesions [2]. Despite being benign, these tumors may be linked to pheochromocytoma and may induce symptoms as a result of pressure or bleeding. This study presents a case report detailing the anesthetic management of a patient with Von Hippel-Lindau (VHL) syndrome undergoing surgical resection of pancreatic lesions and a right adrenal mass, highlighting the importance of tailored anesthesia strategies and multidisciplinary collaboration for optimizing perioperative outcomes in complex genetic disorders.

CASE PRESENTATION
A 25-year-old man presented with a three-week history of excessive sweating, diarrhea, and palpitations. Physical examination revealed a heart rate of 80/min, blood pressure of 176/114 mmHg, and left ventricular hypertrophy on ECG. Further investigations, including abdominal ultrasound, indicated pancreatic cysts and a right adrenal mass. Elevated plasma metanephrine levels (190 pg/mL) and urine fractionated metanephrine (2.3 mg/24 hrs) confirmed the diagnosis of pheochromocytoma. The patient had a history of evisceration due to left-eye retinal detachment and angle-closure glaucoma. PET-CT imaging revealed multiple lesions in the head and body of the pancreas, right adrenal gland, prevertebral region, and right jugular

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foramen. Preoperative optimization involved initiating treatment with T.Prazosin (2.5 mg BD, increased to 5 mg BD) and T. Propranolol (20 mg BD for 7 days), adhering to Roizen's criteria before surgery. Preoperative vitals were stabilized, with a heart rate of 64/min and blood pressure of 136/80 mmHg. Anesthesia induction included midazolam for anxiolysis, radial artery cannulation for blood pressure monitoring, and an epidural catheter placed at the L1-L2 space. General anesthesia was induced with fentanyl and propofol, followed by muscle relaxation with vecuronium. Anesthesia was maintained with a combination of oxygen, nitrous oxide with FiO2 of 0.5, and isoflurane to maintain an age-appropriate minimum alveolar concentration (MAC) of 1–1.2. At the same time, an epidural infusion was activated before the incision and continued throughout the surgery. Management of catecholamine surges and post-resection hypotension was performed with nitroglycerin, sodium nitroprusside, and noradrenaline. The procedure lasted 3 hours and 20 minutes, with stable hemodynamics throughout. Extubation was uneventful, and the patient was monitored in the ICU for two days before discharge on postoperative day 8.

Figure 1: Node enhancement in PET CT-Scan

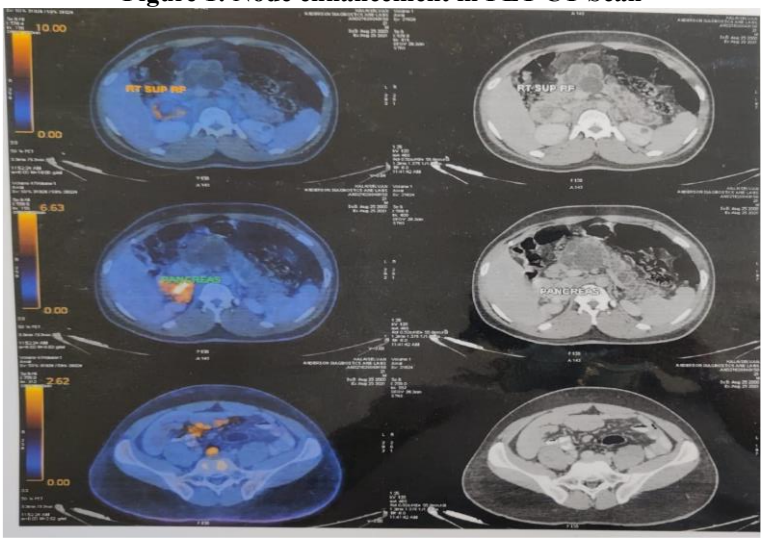
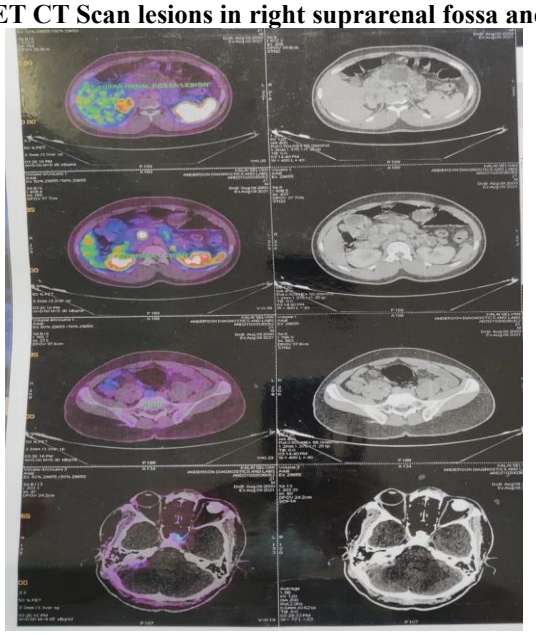


Figure2: PET CT Scan lesions in right suprarenal fossa and pancreas



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2. DISCUSSION

Pre-operative evaluation of Pheochromocytoma patients is a crucial component of the management [3, 4]. Even while alpha-blockade isn't always thought of as absolutely necessary, some researchers suggested appropriate alpha1-receptor blockade [5-7]. In order to guarantee complete blocking, we employed a high-dosage alpha-adrenergic blocker in our patient prior to surgery, with the development of postural hypotension as our goal.

The preoperative conditions listed below are advised by Roizen et al. before pheochromocytoma surgery: Blood pressure should be < 160/90 mmHg for 24 hours before surgery, postural hypotension should be > 80-45 mmHg, the ECG should show no ST-T alterations for a week, and there shouldn't be any PVCs more frequently than once every five minutes. In our instance, we closely followed the guidelines set forth by Roizen. Phenoxybenzamine is typically used to achieve preoperative alpha-adrenergic blockade. However, a newer generation of selective alpha-blockers has been used, which has several advantages over phenoxybenzamine: they have a shorter half-life, do not cause reflex tachycardia, and allow for rapid dosage adjustments, which should reduce preoperative and postoperative hypotension [8]. It is possible to think about terazocin and prazocin. Preoperative beta-blockade is typically not required unless the patient has a tumour that secretes adrenaline. This is because the typical preoperative alpha-adrenergic inhibition does not aggravate cardiac alpha2 receptors. The cause of hypotension after tumour resection could be haemorrhage, insufficient intravascular volume replacement, or the aftereffects of preoperative alpha-receptor blockade [9]. In the current instance, severe hypotension happened at the ligation of the tumor's vascular supply when the right adrenal gland was removed. Since these patients need a lot of fluid following tumour resection, fluids ought to be the first line of management for them. It is thought that controlling hypotension with fluid replacement contributes to a reduction in perioperative mortality [10]. Vasopressors don't work when a person is hypovolemic. It is preferable to utilize phenylephrine and norepinephrine as vasopressors if necessary [11]. This case report demonstrates the effective management of a patient with Von Hippel-Lindau syndrome undergoing surgical resection, highlighting the importance of preoperative optimization and vigilant intraoperative monitoring. While offering valuable insights into tailored anesthesia strategies for complex cases, its limitations include the lack of long-term follow-up data and the inherent constraints of being a single-case study, warranting further research to validate its findings.

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