Introduction

Von Hippel Lindau Disease is an autosomal dominant disorder, which increases susceptibility to benign and malignant tumours, including pheochromocytoma. This disease has an incidence of 1 in 36,000 live births. Pheochromocytomas are tumours arising from adrenal or extra-adrenal chromaffin cells that usually secrete massive amounts of catecholamines (mainly norepinephrine). This tumor causes the classic triad of headaches, sweating and palpitations associated with hypertension in 90% of patients.

Case Presentation

A 13 year old female patient with history of Von Hippel-Lindau (VHL) disease was admitted to our Institution because of cephalgia, hypertension and had a normal physical examination. She received multiple antihypertensive medications including sodium nitroprusside and finally achieved adequate blood pressure control with prazosin and metoprolol. Pheochromocytoma was suspected because of history of VHL disease and family history of pheochromocytoma. Elevated plasma catecholamines and urinary metanephrines as well as radiologic finding of a right adrenal gland tumour confirmed the diagnosis.

Preoperative Evaluation

- No target organ damage, Spinal Hemangioblastomas (T10-12)
- Normal preoperative vital signs, physical and neurological examination
- Normal chemistry panel, Complete Blood Count and coagulation panel
- No electrocardiographic abnormalities or Radiological findings
- Weight 43.5 Kg, Height 153 cms

Anesthetic Management

- Technique: combined general/regional epidural anesthesia (L1-2).
- Basic and Invasive monitors (arterial line and CVP), TOF, Urinary output, esophageal temperature
- Hemodynamic stability during anesthetic induction
- Surgical manipulation: esmolol (50-200 mcg/kg/min) and nitroglycerin (0.5-4 mcg/kg/min)
- After resection: ephedrine 10 mg IV and IV dextrose 0.5 g/kg (capillary glucose 43 mg/dl)
- Accidental spleen laceration and intraoperative hemorrhage (2 L)
- Splenectomy and distal pancreatectomy
- Fluid resuscitation with colloid, cristalloid and red blood cell transfusion
- Low dose norepinephrine was required for a brief period and suspended after volume replacement
- Restoration of acid-base balance
- Successful extubation and transfer to the ICU (no inotropic support)
- Epidural analgesia: ropivacaine 0.125% 6 ml/hr

Table 1. Intraoperative arterial and venous Blood Gas Analysis

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<th>Time</th>
<th>pH</th>
<th>pCO2</th>
<th>pO2</th>
<th>HCO3</th>
<th>Na</th>
<th>Ca</th>
<th>K</th>
<th>CL</th>
<th>CI</th>
<th>Hb</th>
<th>Ha</th>
<th>Na</th>
<th>CO2</th>
<th>lactate</th>
<th>sO2</th>
<th>PaO2</th>
<th>PaCO2</th>
<th>PaCl</th>
<th>Paict</th>
<th>PaOict</th>
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<td>31.9</td>
<td>99</td>
<td>117</td>
<td>130</td>
<td>4.5</td>
<td>38,1</td>
<td>94</td>
<td>76</td>
<td>207</td>
<td>121</td>
<td>116</td>
<td>3.6</td>
<td>133</td>
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<td>99</td>
<td>103</td>
<td>1,2</td>
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<tr>
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</table>

Keypoints

- Conduct a thorough preoperative evaluation in order to identify patients with diseases related to pheochromocytoma (such as VHL).
- Evaluate the presence of target organ damage related with pheochromocytoma or comorbid diseases, mainly in the cardiovascular and central nervous system, that will change the anesthetic management.
- Preoperative control of hypertensive crises is crucial to reduce perioperative morbidity and mortality and it usually includes alpha adrenergic blockade.
- Invasive monitoring of arterial pressure waveforms is mandatory as well as a central venous catheter, in order to evaluate hemodynamic changes during induction, tumour manipulation and after resection and provide an early treatment.
- After tumoral resection patients may require vasoactive medication and caloric intake because of increased risk of hypoglicemia.
- Utilization of regional anesthetic techniques may improve perioperative analgesia and optimize blood pressure control, mainly in open procedures.
- Spinal hemangioblastomas are frequently found in patients wit VHL disease but there are many case reports of the use of epidural anesthesia in this setting without any record of complications.
- Hypertension is frequently found in the postoperative setting and is usually transient.
- Postoperative care should be provided in intermediate or intensive care units.

References


Postoperative Evolution

- Adequate pain control.
- Antihypertensive medication (Prazosin TID).
- Hospital discharge at day 8.

Table 2. Postoperative arterial and venous Blood Gas Analysis