



Anaesthetic Management of a Case of Pheochromocytoma with Dilated Cardiomyopathy-A Case Report

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Abstract: Pheochromocytoma is a rare catecholamine secreting tumour arising from chromaffin cells in the adrenal medulla or in the extra adrenal sites such as urinary bladder, thorax and paraganglion of sympathetic nervous system. The hallmark, regardless of specific catecholamine secreted, is an increase in systemic vascular resistance and blood pressure. The intraoperative management of hyperadrenergic state associated with pheochromocytoma presents a formidable anaesthetic challenge. Catecholamines and their oxidation products cause a direct toxic effect on the myocardium. They may also produce receptor mediated effect on myocardium causing pressure induced cardiomyopathy leading to left ventricle dilatation, hypokinesia and eventually cardiac failure. The definitive management of cardiomyopathy associated with pheochromocytoma includes medical management with alpha-adrenergic blockade, ACE inhibitors, and with or without beta-1 adrenergic receptor blockers followed by excision of tumour. We report the anaesthetic management of a patient with pheochromocytoma and dilated cardiomyopathy posted for open adrenalectomy using proseal laryngeal mask for airway management. The use of proseal laryngeal mask airway (PLMA) to bypass the pressor response of laryngoscopy and ETI is highlighted.

Keywords: Pheochromocytoma; Proseal Laryngeal Mask Airway; Sympathetic nervous system; General anaesthesia.

Detection of pheochromocytoma is imperative since it has lethal potential and is one of the few truly curable forms of hypertension. Ten percent of pheochromocytomas are inherited and can



be a part of multiple endocrine neoplastic syndromes. Both sexes are equally affected with peak incidence occurring in 3rd to 5th decade of life. 80% of pheochromocytomas are located in the adrenal medulla. Right gland is involved more often than left. Adult pheochromocytomas are solid, highly vascular tumours usually 3 to 5 cm in diameter and average 100 g in weight¹. α - adrenergic effects predominate with norepinephrine secreting tumours, and patients present with systolic, diastolic hypertension with reflex bradycardia. β - adrenergic effects predominate with epinephrine secreting tumours and patients usually have systolic hypertension, diastolic hypotension and tachycardia. Headache, sweating, pallor and palpitations are classic signs and symptoms. Orthostatic hypotension is also a common finding and is secondary to hypovolemia and arterial vasoconstriction reflex responses. A catecholamine induced cardiomyopathy can also occur. A global reduction in myocardial pump function results from the net reduction in viable myofibrils and the downregulation of β -receptors. The aetiology is multifactorial and includes catecholamine induced permeability changes in sarcolemmal membranes leading to excess calcium influx, toxicity from oxidized products of catecholamine and damage by free radicals. This pheochromocytoma associated cardiomyopathy normally reverses over a period of months after tumour resection or aggressive medical management². Pheochromocytoma patients may develop cardiac hypertrophy with congestive heart failure (CHF) secondary to sustained hypertension. CHF may be due to coronary artery disease, dilated cardiomyopathy, chronic pressure or volume overload. Ventricular arrhythmias are common in patients with LV dysfunction. Decreased ejection fraction, a hallmark of chronic LV systolic dysfunction, is closely related to increase in the diastolic volume of the left ventricle³.

Case Report: A 31 year old female, weighing 60 kg, presented in the emergency department with complaints of shortness of breath, orthopnea, paroxysmal nocturnal dyspnoea, worsening exercise tolerance and pedal oedema. On examination, she was conscious with a heart rate of 84/min, RR 28/min and blood pressure 130/84 mm of Hg. Chest was bilaterally clear with no air entry at the bases and cardiac sounds were normal. X-ray chest revealed cardiomegaly with bilateral blunting of costophrenic angles suggestive of pleural effusion. ECG showed nonspecific ST segment and T wave changes. She was diagnosed as a case of cardiac failure. USG examination of abdomen revealed right adrenal mass with mild ascites. Echocardiography revealed dilated left atrium, dilated left ventricle and LVEF of 30%. There was systolic dysfunction, mild mitral regurgitation and tricuspid regurgitation. Oxygen inhalation by ventimask, ACE inhibitors, diuretics and β blockers were started and she gradually improved.

She was subjected to CT examination of abdomen and 24 hours urinary catecholamines evaluation. CT findings confirmed the presence of right adrenal mass of size 5.3 x 3.9 cm (Fig.-1) There was a marked elevation in levels of norepinephrine 457.77 (12.10–85.50) $\mu\text{g}/24$ hours, epinephrine 59.20 (1.70 – 22.40) $\mu\text{g}/24$ hours and VMA 25.63 (1.60 – 4.20) $\mu\text{g}/24$ hours consistent with the diagnosis of pheochromocytoma. However, dopamine levels were normal 149.96 (52–480) $\mu\text{g}/24$ hours. She was admitted for preoperative preparation of catecholamines secreting pheochromocytoma with dilated cardiomyopathy for adrenalectomy. Monitoring revealed HR

100/minute, BP 140/100 mm Hg, and RR 24/min. Fundus examination showed Grade-I hypertensive changes. ECG was suggestive of LVH and sinus tachycardia. Repeat ECHO revealed improved LV systolic functions with LVEF of 35%. Tab. phenoxybenzamine 20 mg BD was added. A day prior to surgery right internal jugular vein and left radial artery were cannulated and lumbar epidural catheter was placed. Phenoxybenzamine was discontinued a day prior to surgery as it has a long half life and can cause postoperative hypotension. Rest of the medications were continued. Tab alprazolam 0.5mg was given the night before and 2 hrs prior to surgery.

Patient was wheeled to OR. NIBP, EKG, SpO₂, IBP, CVP and temperature monitoring were started. 10 ml of 0.125% bupivacaine with 20 µg of fentanyl was given through the epidural catheter. At induction, the heart rate was 102/min, IBP 130/84 mm of Hg, CVP 8 cm of H₂O and SpO₂ 100%. Anaesthesia was induced with inj fentanyl 100 µg, propofol 140 mg, vecuronium 6 mg iv, Inj esmolol 50 mg iv was administered to obtund the pressor response. Airway was secured with proseal laryngeal mask airway (PLMA) size 3. Continuous hemodynamic monitoring revealed a transient surge in blood pressure to (150/100 mm Hg) and heart rate 125/min which was controlled with inj esmolol 30 mg and was followed by a continuous infusion at the rate of 100µg/kg/min. Anaesthesia was maintained with O₂ and N₂O (33:67) in isoflurane 1%. Operative manipulation of tumour resulted in rise in heart rate to 130 per min and blood pressure 190/120 mm of Hg which required sodium nitroprusside infusion at a rate of 0.3µg/kg/min to control it. Ligation of right adrenal vein and removal of tumour resulted in decrease in systolic BP to 70 mm of Hg and CVP dropped to 4 cm of H₂O. Esmolol and nitroprusside infusion were discontinued. 500 ml of hydroxyethyl starch and ringer lactate each were infused and inotropic support was started with dopamine infusion at the rate of 10µg/kg/min and haemodynamics were stabilized. At the completion of surgical procedure, the patient was reversed with inj neostigmine 2.5 mg and inj glycopyrrolate 0.4 mg and PLMA was removed. She made an uneventful recovery and was shifted to HDU for postoperative management.

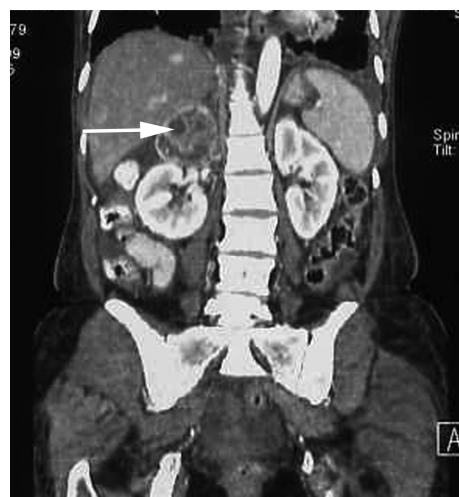


Fig 1: Arrow showing heterogenous mass at upper pole of right kidney reaching up-to renal hilum anteriorly showing heterogenous enhancement – Adrenal mass

Discussion: Adequate preoperative preparation of patient with pheochromocytoma is mandatory to prevent severe intraoperative cardiovascular complications. The mainstay of preparation is α -adrenergic blockade with or without β -blockade. Phenoxybenzamine, prazosin, doxazosin, terrazosin



are commonly used α -adrenergic blockers. Despite premedication with phenoxybenzamine and a β -adrenergic receptor blocker, a significant percentage of patients experience considerable intraoperative hemodynamic lability. Larger tumour size, prolonged duration of anaesthesia and increased levels of preoperative urinary epinephrine, norepinephrine, metanephrines and vanillylmandelic acid (VMA), are significant risk factors for the adverse perioperative events or complications.

Phenoxybenzamine is a non-competitive, non-selective (both α_1 and α_2) adrenergic blocker given in the dose of 10–30 mg/day. This is the preferred drug for preoperative preparation. However, postural hypotension, nasal congestion and dry mouth are the common side effects. Metyrosine, controls the rate limiting step of catecholamine production, by inhibiting tyrosine hydroxylase which catalyses the conversion of tyrosine to diphenylalanine. Metyrosine is recommended in patients who are refractory to phenoxybenzamine or in whom β -adrenergic blockade is contraindicated⁴. Correction of hypovolemia is also important in reduction of morbidity. If tachycardia, arrhythmia, or myocardial infarction is present despite adequate α blockade, use of β -blockers is recommended. They are used to reverse the harmful effects of sympathetic nervous system activation in heart failure. β -blockers improve the EF and decrease ventricular remodelling. American college of Cardiology and American Heart Association guidelines recommend β -blockers as integral part of the therapy for heart failure⁵. Potential objection to use of β -blockers are incompletely α -blocked patient, CHF, pulmonary oedema, bronchospasm and 2nd or 3rd degree heart block. Esmolol results in rapid decrease in systolic blood pressure without effect on diastolic pressure. While β -blockers might be expected to depress myocardial contractility and cardiac index function, in the face of a markedly elevated after-load, the use of short acting agent with rapid reversibility on discontinuation can minimize the duration of myocardial depression⁶.

On laryngoscopy or intubation there is mechanical irritation of stretch receptors situated in the respiratory tract leading to reflex haemodynamic responses in the form of increase of heart rate and blood pressure. Reflex haemodynamic response to laryngo-tracheal stimulation by endo-tracheal intubation does not cause any lasting damage in the normotensive patients. But the possible cardiac or cerebral complications (myocardial infarction, cerebrovascular accident) may result in hypertensive patients due to this exaggerated hypertensive response⁷. Previous laryngeal mask studies indicate only minor hemodynamic responses to classic LMA insertion with a 0 to 20% increase in heart rate and mean arterial blood pressure⁸. Hemodynamic responses to PLMA insertion were similar to those of the classic LMA in a randomized comparative trial of 280 patients anesthetized with a standard technique⁹. Two non-randomized studies of 335 patients with varying anaesthetic techniques reported hemodynamic variables change with PLMA less than 10%^{10,11}. An abstract reports hemodynamic response to insertion/removal of the PLMA as significantly less than for tracheal intubation/extubation¹². Literature suggested a minimal haemodynamic response to PLMA insertion, which was significantly less than for tracheal intubation hence we decided to insert PLMA.



The combination of epidural analgesia and drugs resulted in minimal fluctuations in blood pressure and heart rate during surgery. Optimal sympathetic blockade and de-afferentation was achieved with preoperative placement of epidural anaesthetic. Placement of PLMA for securing airway caused less hemodynamic surge and a smooth induction. Multiple pharmacotherapeutic interventions have been proposed to minimize intraoperative cardiovascular complications. No technique can be termed as ideal. However, a combination of preoperative preparation with α and/or β -adrenergic blockers, epidural anaesthesia followed by general anaesthesia, aggressive intraoperative hemodynamic monitoring with β -blockers and sodium nitroprusside infusion followed by inotropic support after tumour excision with adequate postoperative pain relief led to optimal management in our patient.

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