A Case Report

Anaesthetic Management of a case of Bilateral Pheochromocytoma with Cardiomyopathy

Rupali Namdev Gorgile
IMSR, Mayani, Satara, India

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ABSTRACT
A 22 year old male patient with bilateral pheochromocytoma presented with dilated cardiomyopathy, hypertension and tachycardia with history of Congestive cardiac failure (CCF). Confirmation of diagnosis was done by CT scan and raised catecholamines levels. Preoperative BP and HR were controlled with phenoxybenzamine, Angiotensin converting enzyme (ACE) inhibitors and Beta blockers. The anaesthetic technique used was general anaesthesia with epidural analgesia. Extubation was done after 12hrs with the help of Inj.Dexmedetomidine infusion.

Key Words: Pheochromocytoma, Dilated cardiomyopathy, Anaesthetic management, Dexmedetomidine

INTRODUCTION
Pheochromocytoma is a rare catecholamine secreting tumor arising from chromaffin cells in the adrenal medulla or extra adrenal sites such as urinary bladder, thorax and paraganaglion of sympathetic nervous system. These tumors are not common, occurring in 0.1% of hypertensive population only 2, and typically occurs in patients who is 30-50 years of age11. A “rule of 10” has been applied to these tumors. 10% bilateral, 10% malignant and 10% are extra-adrenal11. The hallmark is an increase in systemic vascular resistance with or without tachycardia. 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 produce receptor mediated effect on myocardium causing pressure induced cardiomyopathy leading to left ventricle dilatation, hypokinesia and eventually cardiac failure11. The definitive management of cardiomyopathy associated with pheochromocytoma includes medical management with alpha-adrenergic blockers, ACE inhibitors, beta-1 adrenergic blockers followed by excision of tumor.

CASE REPORT
This case was done in cancer hospital. A 22 year old male, weighing 55kg was admitted in hospital for removal of tumor. On pre-operative checkup, he gave history of intermittent headache since 1 year, breathlessness on exertion, weakness, sweating, palpitation since 6 months, he had history of CCF 3 months back with LVEF 23%, type II diastolic dysfunction and biventricular failure on 2DECHO. During investigations
bilateral pheochromocytoma was diagnosed on CT scan with marked elevation in levels of normetanephrine 697pg/ml(440pg/ml), metanephrine 274pg/ml(220pg/ml) and urinary vanillyl mandelic acid(VMA) 27mcg/24hrs(1.6-4.20).

Since then he was on T.Ivabradine 5mg bd, T.Digoxin 0.25mg od, T.Prolomet R (Metoprolol+Ramipril) 25 od, T.Tide (Torsemide) 5mg and referred here for further management. On examination, his general condition was moderate, with presence of puffiness of face and pallor. His heart rate was 110/min, BP -130/80mmHg in sitting &140/100mmHg in supine position with normal systemic examination except for presence of basal crepts bilaterally .There was inverted T wave with ST depression in all leads on ECG with mild cardiomegaly on CXR with haziness in lung field. His fresh 2DECHO showed dilated LV, mild MR, LV dysfunction, rim of pericardial effusion, mild generalized hypokinesia, EF-35%. His other investigations like complete blood count, renal profile, liver profile, serum electrolytes, and blood sugar level were normal. He was advised T. Phenoxybenzamine 10mg bd, T.Inderal (Propranolol) 10mg bd.T.0.25mg hs with above drugs and called for review check up after 3weeks in preanaesthesia check up department. After 3weeks his general condition improved with his heart rate(HR) 98/min, Blood pressure(BP)-128/80mmHg and 2DECHO revealed EF40-45% with rest findings same as previous 2DECHO. Inj. Hydrocortisone 100mg tds was started since 3days. A day prior to surgery under local anesthesia central line was put. All drugs were taken with sips of water except for T. Phenoxybenzamine on the day of surgery. Monitors were attached and vitals were HR-98/min, BP-150/98mmHg, and SpO2:-97%,CVP:-0cm ofH2O, so 500ml of NS was infused fast. Inj. Dopamine 400mg in 500ml D5. Inj.Sodium Nitroprusside(SNP) 5mg in 500ml D5 infusion covered with black paper were kept ready. Emergency drugs, defibrillator was also kept ready. Then under local anesthesia epidural catheterization was done at T12-L1 interspace and 10ml of 0.25% bupivacaine was given. Left radial artery was also cannulated under LA with arterial blood pressure(ABP) 180/100mmHg so 0.01% infusion of SNP was started in titrated dose to make ABP around 130/80mmHg, simultaneously preoxygenation was done with 100% O2 for 3min.Then he was premedicated with Inj.Emset, Inj.midazolam1mg, Inj. fentanyl 100mcg Inj. hydrocortisone 100mg and induced with Inj.propofol100mg, Inj. vecuronium bromide 6mg. 75mg xylocard was given one minute before laryngoscopy to obtund pressor response and intubation was done with cuffed ETT no 9mm and EtCO2 monitor was connected. Maintenance of anaesthesia was done with O2:N2O(50:50), isoflurane (0.4%-0.8%) and Inj.vecuronium bromide with positive pressure ventilation on volume control mode .Continuous hemodynamic monitoring was done. Intraoperative surge of BP >180/114 and rise in heart rate >120 was controlled by 0.01% infusion of SNP in titrated dose and bolus of inj.metoprolol 2mg respectively. After removal of tumor on right side
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there was no fall in BP but it remained around 150/90 mmHg.
Tumor handling on left side also causes surge in blood pressure to 166/100mmHg and heart rate to 120/min which was controlled by SNP and metoprolol same as above. Patient received 500ml of 6% hydroxyethyl starch, normal saline, ringer lactate to maintain CVP around 5-6 cm of H2O. Removal of tumor on left side resulted drop in systolic blood pressure to 100mmhg and CVP dropped to 3cm of H2O. Infusion of SNP was discontinued and 500ml of 5% dextrose, DNS, hydroxyethyl starch, one unit of blood was infused to restore CVP and blood pressure.
Infusion of inj. dopamine was started at the rate of 5mcg/kg/min and inj. hydrocortisone 100mg was given. Throughout the procedure, analgesia was maintained with 8ml of 0.25% bupivacaine at 45min interval. Urine output was also maintained. At the conclusion of surgery, patient was shifted to surgical intensive care unit with endotracheal tube and put on ventilator with SIMV mode with TV-500ml and RR-12/min. Infusion of inj. Dexamethasone 200mcg in 50ml NS at the rate of 1mcg/kg for 10 min followed by 0.3 - 0.6mcg/kg/hr was started for sedation. Next day morning patient was extubated through gradual weaning from SIMV-CPAP-T piece. Post-operatively patient’s vitals were stable with blood pressure around 120/70mmHg and HR 70/min. He was received antibiotics, Inj. Clexane 0.4mg subcutaneously for DVT prophylaxis, Inj. Hydrocortisone 100mg tds for 3days. Patient was discharged after three weeks with T. Hisone (Hydrocortisone) for steroid cover, T. Tide (Torsemide) 5mg od, T. Digoxin 0.2mg od.

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DISCUSSION
Primary preoperative goal should be good pharmacological control of adverse effects of circulatory catecholamines and restoration of blood volume in heart failure patient with the help of alpha and beta blockers and at the same time avoid drug induced myocardial depression, increase in ventricular afterload, preload and tachycardia. Phenoxybenzamine is a non-competitive, non-selective (both alpha1 and alpha2) adrenergic blocker given in the dose of 10-20mg twice daily. This is the preferred drug for preoperative preparation and our patient was given the same drug11. Metyrosine inhibits
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catecholamine synthesis and is recommended in patients who are refractory to phenoxybenzamine or in whom beta adrenergic blockade is contraindicated. Correction of hypovolemia is also important in reduction of morbidity. If tachycardia, arrhythmia or myocardial infarction is present despite adequate alpha blockade, use of beta blockers is recommended. They are used to reverse the harmful effects of sympathetic nervous system activation in heart failure. They improve the EF and decrease ventricular remodelling. American college of Cardiology and American Heart association guidelines recommend B-blockers as integral part of the therapy for heart failure. The main aim of anaesthetic management is to provide optimal surgical conditions and suppress the response of endotracheal intubation, surgical stimulation, tumor handling, devascularization and smooth recovery with smooth extubation. Combined epidural and general anaesthetic technique is preferred, so we used same. Monitoring in our case was performed as per recommendation. Lignocaine 1.5mg/kg IV was given 1 minute before laryngoscopy to attenuate the stress response. To control BP and tachycardia, we used IV infusion of SNP in titrated doses and metoprolol IV boluses. As patient had history of CCF and underwent major surgery we avoided extubation on operating table and extubated the patient after 12 hours with the help of dexmedetomidine. Dexmedetomidine is an alpha2 adrenoceptor agonist which gives better haemodynamic stability, anxiolysis, hypnosis and analgesia without respiratory depression, so it helps in smooth recovery and extubation which was needed in our case. There is case study of comparison of post-operative ICU sedation between dexmedetomidine and propofol in mechanically ventilated patients which reported that dexmedetomidine is a safe and acceptable sedative agent with better haemodynamic stability and analgesic properties without respiratory depression. There is also case report of usage of IV infusion of dexmedetomidine and magnesium sulphate in perioperative management of pheochromocytomas. Hence we used infusion of dexmedetomidine postoperatively in ICU till patient get extubated. Steroid cover is mandatory for patients undergoing bilateral adrenalectomy. We also used hydrocortisone in preoperative, intraoperative and postoperative period.

In conclusion, the proper anaesthetic management of pheochromocytoma is a truly rewarding challenge. Mortality has been reduced in recent years due to better knowledge of these tumors and more adequate pretreatment regimens, better intra and post-operative management. Dexmedetomidine has a better safety profile for ICU sedation with analgesic properties without respiratory depression.

REFERENCES:


