A rare case of hypertrophic obstructive cardiomyopathy posted for adrenalectomy for pheochromocytoma

INTRODUCTION

Phaeochromocytoma is a tumour of the adrenal medulla that secretes excessive amounts of catecholamine. The patients present with fluctuating blood pressure, sweating and palpitations.[1] Pre-operative management consists of control of blood pressure and restoration of intravascular volume.[2]

Hypertrophic obstructive cardiomyopathy is characterized by asymmetric hypertrophy of the myocardium resulting in left ventricular outflow tract obstruction. Decrease in venous return and systemic vascular resistance or increase in myocardial contractility worsens the outlet obstruction. Management of anaesthesia in these patients entails maintenance of haemodynamic parameters and management of specific complications like hypotenion, dysrhythmias and congestive heart failure.[3]

The presence of both of these conditions together poses a unique problem because the opposite management strategies like the use of vasodilators for control of blood pressure in pheochromocytoma intraoperatively can lead to sudden collapse in hypertrophic obstructive cardiomyopathy; also, presence of both the conditions increase the chances of cardiac failure and ischaemia. Hence, we present here a case of hypertrophic obstructive cardiomyopathy posted for adrenalectomy.

CASE REPORT

A 56-year-old female weighing 45 kg came with complaints of palpitations, sweating, headache and episodes of dizziness since 4 years. On examination, she had a blood pressure of 180/106 mmHg and Mallampatti class I, while the rest of the examination was normal. The vinyl mandelic acid levels were 21.4 mg/24 h. Computed tomography of the abdomen showed a mass at the right adrenal of size 8 cm×7 cm. The iodine-131-meta-iobenzylguanidine scan confirmed the tumour. The electrocardiogram showed severe left ventricular hypertrophy. 2d echocardiography showed an asymmetrical hypertrophy of the septum of 23.9 mm, severe left ventricular outlet obstruction with pressure gradient of 104 mmHg and diastolic dysfunction. Blood sugar level was normal. She was posted for adrenalectomy.

To control her blood pressure, she was cautiously started on Tab. Phenoxybenzamine and Labetalol, which was gradually increased to the current dose of 100 mg and 40 mg BD, respectively.

She was posted for surgery after control of blood pressure to 140/90 mmHg. Haematocrit before surgery was 30.

After appropriate written informed consent, she was taken into the operation theatre. A pulseoximeter and defibrillator were attached. Intravenous fluids were started to preload the patient. An epidural was inserted at the T9-10 levels for post-operative analgesia with opioids only.

Pre-medication consisted of Inj. Glycopyrrolate 225 μg IM, Inj. Midazolam 1 mg IV, Inj Fentanyl 50 μg IV, Inj. Clonidine 60 μg IV, Inj. Hydrocortisone 100 mg and Inj. Dexamethasone 8 mg along with antiemetics and antacids.

The pulse was 80/min and blood pressure 140/96 mmHg. An arterial line and central venous line was inserted.
She was induced with Inj. Pentothal sodium 400 mg and Inj. Rocuronium 45 mg IV. One minute prior to intubation, Inj. Lignocaine 90 mg was given IV. After intubation, anaesthesia was maintained with O₂:N₂O 50% mixture and Isoflurane with Inj. Vecuronium as the skeletal muscle relaxant. The vitals remained stable till this time.

When the tumour was reached, the blood pressure suddenly shot up to 300/130 mmHg. Immediately, a bolus of Inj. Esmolol followed by infusion at the rate of 300 μg/kg/min was started and the blood pressure came down to 160/96 mmHg. The dose of Esmolol was reduced gradually.

After clamping of the adrenal vein, the blood pressure immediately started falling and dropped to 60 mm Hg systolic. Esmolol infusion was stopped and Inj. Noradrenalin was started at 0.5 μg/kg/min. The blood pressure increased to 80 mmHg, but did not increase further; hence, another pressor Inj. Dopamine 3 μg/kg/min was cautiously started. Fluids were administered to improve the left ventricular filling. Gradually, the blood pressure came up to 100/60 mmHg. The blood sugar level was checked and found to be 120 mg/dL. Rest of the course was uneventful.

At the end of the surgery, the blood pressure came to 120/78 mmHg. The patient was reversed using Inj. Glycopyrrolate 450 μg and Inj. Neostigmine 2.5 mg IV and was extubated. Intraoperative urine output was 600 mL.

The patient was shifted to the surgical intensive care unit and monitored for vitals and blood sugar levels. The infusions were gradually tapered. Post-operative analgesia was maintained with epidural Fentanyl. Rest of the post-operative course was uneventful.

**DISCUSSION**

Pheochromocytoma is a tumour of the chromaffin cells of the medulla that secretes catecholamines, leading to hypertension, palpitations and headache. These patients are highly prone to arrhythmias, like atrial fibrillation and ventricular tachycardia. Anaesthetic challenges include control of blood pressure and intravascular volume perioperatively to prevent sudden increase in blood pressure and blood loss, prevention of arrhythmias and management of hypotension, which occurs after the venous clamping due to the decrease in the catecholamines.

Hypertrophic obstructive cardiomyopathy can be a genetic disorder but is also seen in some cases of pheochromocytoma due to excessive adrenergic stimulation and hypertension.

The anaesthetic challenges include prevention and management of perioperative arrhythmias and maintenance of adequate preload, afterload and heart rate to relieve left ventricular outflow tract obstruction. The presence of both of these conditions together poses unique problems.

The control of the blood pressure pre-operatively requires use of alpha and beta blockers. The vasodilatation hence caused can lead to sudden collapse in hypertrophic obstructive cardiomyopathy due to outflow obstruction.

Hence, Tab. Phenoxybenzamine and Labetolol were started cautiously and the dose was gradually increased.

Handling of the tumour produces sudden increase in the blood pressure, which can be reduced using Inj. Nitroprusside, but in this case vasodilatation was contraindicated and hence Esmolol was preferred.

After resection of tumour, the blood pressure tends to go down suddenly due to withdrawal of the catecholamines. Use of pressors like Dopamine, which also increases the heart rate, can lead to decreased cardiac filling and increased outflow obstruction. Hence, we started with Noradrenalin, and used Dopamine cautiously.

An epidural catheter was reserved for pain relief post-operatively with the use of opioids alone. Use of local anaesthetics was avoided as the resultant vasodilatation could lead to sudden outflow obstruction of the left ventricle. One study showed that a slight hypotension caused by epidural anaesthesia had a devastating effect on the patient.

Adequate monitoring for the blood sugars was done as, post-operatively, pheochromocytoma patients are likely to go into hypoglycaemia.

About 10% of these tumours can be extraadrenal. Some could be coexisting with the adrenal tumours and missed in the diagnosis. The classical drop in the blood pressure seen post-removal of the adrenals is not seen here due
to the continued secretion of the catecholamines by the extraadrenal tumours. Surgical removal is the treatment of choice. These tumours have a greater chance of recurrence and hence follow-up is required.[7]

Several cardiac pathologies like failure, cardiomyopathy and ischemic damage are seen in pheochromocytoma.[8] As there is a chance of reduction in the outflow obstruction after tumour resection, we are following-up the case.[4]

CONCLUSION

Pheochromocytoma itself is a huge anaesthetic risk, which is compounded many fold by the presence of severe hypertrophic obstructive cardiomyopathy. Anaesthetic drugs can exacerbate the life-threatening cardiovascular effects of catecholamines. Knowing the pathophysiology well before using various drugs available in our armamentarium can reduce the complications to a great extent.

Shweta R Yemul-Golhar, Pradnya M Bhalerao, Yogesh Gavali, Kalpana V Kelkar
Department of Anaesthesia, B. J. Medical College, Pune, Maharashtra, India

Address for correspondence:
Dr. Shweta R Yemul-Golhar,
505, Daisy, Silverdale Colony, B. T. Kawade Road,
Ghorpadi, Pune - 411 001, Maharashtra, India.
E-mail: Shweta.golhar@gmail.com

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Announcement

Dr. TN Jha and Dr. KP Chansoriya travel grant
From the year 2011, the Dr. TN Jha and Dr. KP Chansoriya travel grant will be awarded to the participants from 15 states. All the states can select their candidate during their annual conference and send them with the recommendation of the Secretary. Only one candidate is allowed from each state. In case, if two states have a combined annual meet but separate as per the records, have to select one candidate from each state. If more than 15 states recommend the candidates for the award, selection will be made on first come first served basis.

Dr. M V Bhimeshwar
Hon. Secretary - ISA