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Anesthetic Management of Pheochromocytoma in a Child- A Case Report

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Abstract

Background: Pheochromocytoma is a catecholamine secreting tumour which imposes multimodal challenges to the anaesthesiologist in its perioperative management. We report a case of a 12 year old boy with complaints of palpitations, headache, sweating and sleeplessness. Pheochromocytoma was diagnosed by 24 hours urine metanephrines, vanillylmandelic acid and MRI abdomen. The main stay in the surgical management of pheochromocytoma is preoperative preparation which determines the outcome. Patient was operated under general anaesthesia with epidural analgesia. Beta blockers, nitroglycerine, sodium nitroprusside and phenylephrine are required to manage the hemodynamic fluctuations and should be used appropriately. Conclusion: Successful management requires careful preoperative optimization, meticulous intraoperative planning, and hemodynamic management.

Keywords: Atenolol; Metanephrines; Pheochromocytoma; Sodium nitroprusside; Vanillyl Mandelic Acid.

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Introduction

Pheochromocytomas are rare neuroendocrine tumors originating from chromaffin cells of the adrenal medulla. Extra-adrenal paragangliomas arise from autonomic ganglion. It may arise in any portion of paraganglion system, though they most commonly occur below the diaphragm, usually in the organ of Zuckerkandl. Pheochromocytomas present with classical triad of a headache, palpitations and sweating. Cardiac arrhythmias and hypertensive crisis may occur during surgery due to high levels of circulating catecholamines. Surgical resection of the tumor is the mainstay of treatment. We describe a successful anaesthetic management of a case of pheochromocytoma in a child.

Case Report:

A 12 year old boy was admitted with complaints of palpitations, headache, sweating, sleeplessness and vague abdominal pain for five months. On examination patient was thin built and his Pulse rate was 102/min, Blood pressure was 210/100mmHg. System examination was normal. The history was suggestive of pheochromocytoma for which he underwent all investigations. ECG showed sinus tachycardia with heart rate of 112 beats /min with no evidence of PVCs. ECHO showed mild concentric LVH. Examination of optic

fundus was normal. Urine analysis was done which showed Metanephrine 79.8 µmol/day normal 0.5 to 5 µmol/day. Vanillylmandelic acid (VMA) in 24 hours urine collection was 63.3mg/day normal 0.0-13.6 mg/day. On further evaluation with MRI

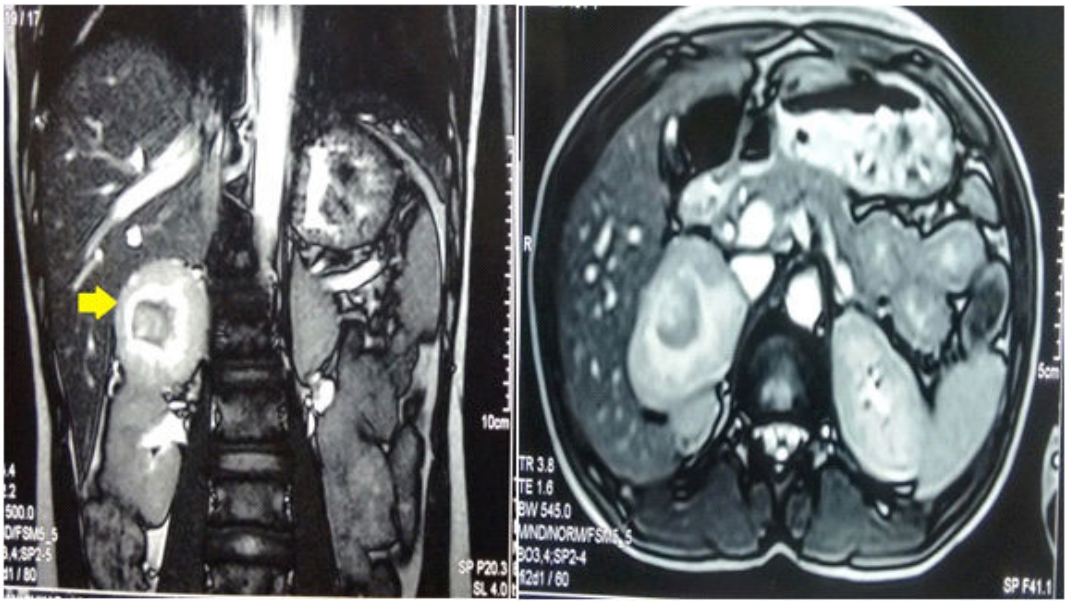

Fig. 1: Coronal and Axial T2- weighted sequence MRI of the abdomen demonstrates right heterogeneous adrenal pheochromocytoma. (Light bulb sign-yellow arrow).

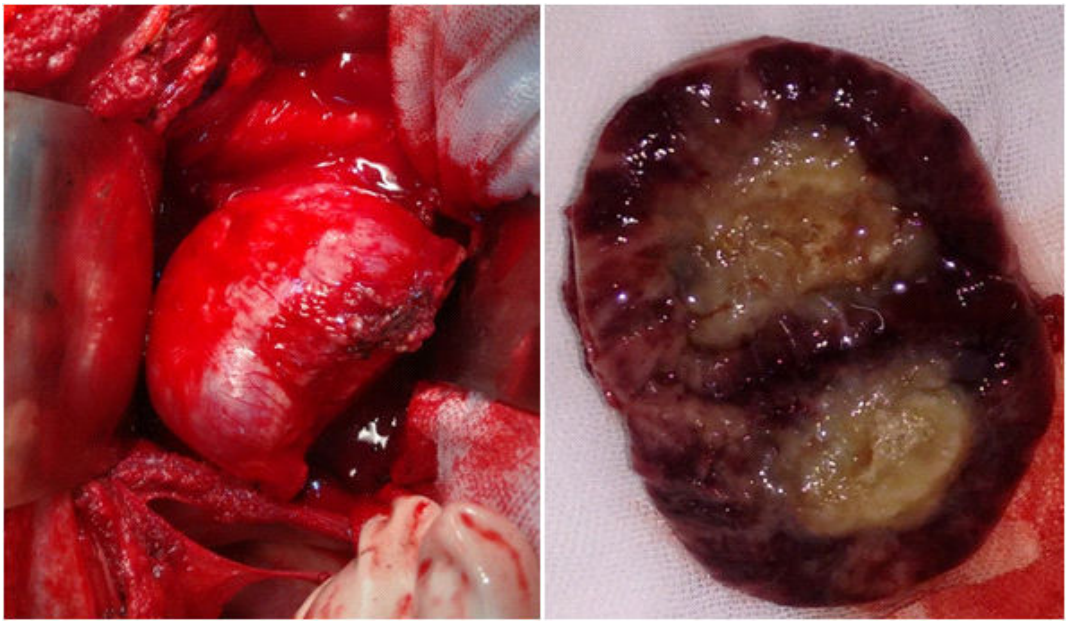

Fig. 2: Intra operative gross appearance and cut section of the tumour.

abdomen, it was confirmed as right adrenal mass which was 6×5cm in size. Patient was started on oral prozosin 2.5 mg twice daily, later patient was also started on oral Atenolol 50mg twice daily for three weeks.

In the preoperative assessment, pulse rate was 102/min and BP was 130/86 mmHg. Airway examination was normal. Patient was assessed under ASA III. On the day of surgery, pulse rate was 96/min and BP was 140/90mmHg. After obtaining informed written consent, patient was shifted to operation theatre and peripheral intravenous line secured in left forearm with 18G IV cannula. He was preloaded with fluid. Intra operative

monitoring was initiated. General anaesthesia was induced with propofol 50 mg intravenously and tracheal intubation facilitated with vecuronium 2.5mg. In right lateral position, epidural catheter was placed in T12-L1 space and fixed at 6 cm. 7 Fr size cental venous catheter placed through right internal jugular vein and CVP was monitored. Right radial artery cannulation was done with 20 gauge arterial catheter and IBP was recorded. Anaesthesia maintained with N_2O:O_2 66%:33%, sevoflurane 2%. Epidural analgesia was achieved with 0.25% bupivacaine 8ml and subsequent top-up doses. Intravenous crystalloids (normal saline and ringer lactate) were infused at 20ml/kg/hour with CVP monitoring. Till clamping of tumour vessels, HR was 76 to 97/min; SBP was 140 to 190 mmHg; DBP was 90 -110mmHg and SpO_2: 99 – 100%. Intraoperative hypertensive episodes were managed with sodium nitroprusside infusion at 0.5-1 µg/kg/minute till the ligation of suprarenal vein. After clamping and resection of tumour, HR was 90/minute; BP was 78/47 mmHg , Spo 2:99 %; Hypotension was managed with IV fluids and injection phenylephrine 100µg IV boluses. The total duration of surgery was 140 min and blood loss was 500 mL. Intraoperative blood sugar was within normal range. 2 units of whole blood were transfused. Urine output was adequate throughout the procedure. After completion of surgery, neuromuscular block was reversed, trachea extubated. Patient's hemodynamic status was stable during the entire postoperative period. Epidural analgesia was continued for 72 hours with 0.0625% bupivacaine and fentanyl 2µg/ml. Histopathology of the right adrenal gland confirmed the diagnosis of pheochromocytoma. Patient was discharged on 14th POD.

Discussion

Pheochromocytoma (PCC) is a catecholamine secreting tumor that originates in the adrenal medulla or in chromaffin tissues along the paravertebral chain extending from the pelvis to the base of the skull. More than 95% of all pheochromocytomas are found in the abdominal cavity and 90 % originate in the adrenal medulla. Pediatric PCC/PGLs appear to follow a pattern, which call "10%–90% rule" and is as mentioned below:

- 10%: Malignant
- 20%: Synchronous bilateral
- 30%: Extra−adrenal
- 40%: Familial
- 50%: Recur (second PCC/PGL) by 30 years
- 60%: Boys
- 70%: Sustained hypertension
- 80%: Germline mutations
- 90%: Secretory
- 100%: Germline + somatic?

Pheochromocytoma typically occurs in patients of 30–50 years age. Approximately 0.1% of the hypertensives suffer from this surgically correctable cause of hypertension which is often fatal if left untreated. Familial pheochromocytoma usually occurs as bilateral tumour, may be associated with MEN syndrome,Von Hippel Lindau syndrome or Neurofibroma-1 syndrome. The organ of zuckerkandl near the aortic bifurcation is the most common extra-adrenal site**.** Headache, sweating and palpitations are classic symptoms. Hypertension is the most frequent sign. Orthostatic hypotension can occur secondary to hypovolemia and impaired vasoconstrictor reflex response. Elevated blood glucose levels can occur secondary to catecholamine stimulation of glycogenolysis and inhibition of insulin release. Uncontrolled catecholamine release can result in malignant hypertension, cerebrovascuar accident, cardiomyopathy, congestive cardiac failure and myocardial infarction.[1,2] Diagnostic tools usually used are plasma metanephrine and 24-hour urine metanephrine. Urinary Vanilylmandelic acid test is used for screening.[5] CT / MRI provides tumour location and anatomical information.

Roizen's criteria for optimisation of the patient for surgical tumour excision are

1. Blood pressure <160/90 mmHg for 24 h prior to surgery;
2. No orthostatic hypotension with blood pressure <80/45 mmHg;
3. No ST or T wave changes for 1-week prior to surgery;
4. No more than 5 premature ventricular contractions in 5 min.[1]

Anesthetic goals are preoperative administration of alpha adrenergic blocker with or without beta blocker, correction of hypovolemia, avoidance of drugs or maneuvers that increases catecholamine levels and maintenance of cardiovascular stability.[7] In our case we used prazosin, a selective α1-competitive blocker. Prazosin interferes selectively with post synaptic α- Adrenergic receptor function. Prazosin causes less tachycardia and postural

hypotension than other α-adrenoreceptor blockers like phenoxybenzamine. Here we used atenolol as β-blocker. Alpha blockade helps to lower blood pressure, increase intravascular volume, prevents paroxysmal hypertensive episodes and improves myocardial performance and thus tissue oxygenation. Intraoperatively adequate plane of anaesthesia was maintained with sevoflurane and N_2O:O_2, analgesia maintained with epidural bupivacaine, fentanyl and avoiding hypovolemia with crystalloids with CVP monitoring and intraarterial BP monitoring. We managed intraoperative hypertensive episodes with sodium nitroprusside infusion. Patient was on continuous monitoring, we did not encounter arrhythmias in this patient. After ligation of tumour vessels and tumour removal, factors such as decreased catecholamine levels, residual alpha blockade, increased depth of anaesthesia, intraoperative fluid loss lead to precipitous hypotension. Adequate expansion of plasma volume with crystalloids plays important role in preventing precipitous fall in blood pressure. Vasopressor and inotropes are second line of management. In our case adequate fluid infusion before tumour vessel ligation helped in preventing precipitous hypotension. We used both fluids and directly acting alpha agonist phenylephrine 100 μg bolus during severe hypotension with immediate favourable response. After tumour resection due to loss of catecholamine dependent inhibition, insulin level rises and hypoglycaemia can occur. [1,2]

Conclusion

Hypotension is the most common cause of death in immediate postoperative period, hence postoperative haemodynamic monitoring and prevention of hypoglycaemia plays important role in preventing mortality. Anaesthetic management of pheochromocytoma is always a challenge to the anaesthesiologist. Thorough knowledge of pathophysiological changes, adequate pharmacological preparation of the patient and meticulous perioperative management helps in decreasing the mortality in a case of pheochromocytoma.

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