Case Report

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Anaesthetic management of pheochromocytoma in a child

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ABSTRACT

Pheochromocytomas are an unusual tumour in paediatric age group. A 6 year old hypertensive boy with left sided pheochromocytoma was scheduled for excision of tumour. He had presented with complaints of pain in abdomen, excessive sweating, headache and raised blood pressure. Diagnosis was confirmed by CT scan (abdomen) and raised 24 hour urinary catecholamine levels. Preoperative blood pressure was controlled with prazosin (ά-adrenergic blocker) and propranolol (β-adrenergic blocker). The anaesthetic technique used was general anaesthesia with caudal catheter. Child was later discharged on oral antihypertensive.

Key words: Pheochromocytoma, paediatric hypertension

INTRODUCTION

Pheochromocytoma, a catecholamine secreting tumour, arises from the chromaffin cells. The most common site of origin is adrenal medulla. Only 10% occur in children in whom they present most frequently between 6 and 14 years of age occurring more often on right side than left side. Pheochromocytoma may also be associated with other syndromes such as neurofibromatosis and von Hippel –Lindau disease and as a component of multiple endocrine neoplasia syndromes (MENII A and B). This case report describes the perioperative anaesthetic management of a child with left sided pheochromocytoma.

CASE REPORT

A six year boy, weighing 14kg, was admitted in paediatric surgery ward with complaints of severe abdominal pain, headache, excessive sweating since 1 yr. He was diagnosed to have left sided pheochromocytoma by raised levels of urinary catecholamine, ultrasound and CT scan (left adrenal mass 4.7 X 2cms). He was on tab. enalapril5 mg. His physical examination revealed a pulse rate of 112 min-1 and BP of170/110 mmHg. Other investigations like complete blood count, plasma glucose, urea, electrolytes were within normal limits. His antihypertensive medication was changed for adequate control of BP with tab.prazosin 2 mg thrice daily and tab. propranolol 10 mg bd. Airway examination was normal. Patient was taken for left sided adrenalectomy when BP was <140/90 mmHg on preanaesthetic examination.

Patient was kept nil orally for 6 hours on maintenance fluid to avoid dehydration and morning doses of oral antihypertensive were continued. Inj. Glycopyrrolate 0.004 mg/kg (i.m) and inj. midazolam 0.05mg/kg (i.m) premedication was administered. I.V line was established. Standard monitors (SpO_2, NIBP, ECG) were attached. His preoperative H.R was 105 min-1, B.P. was 134/98 mmHg (mean 110 mmHg).Vasodilators and inotropes were kept ready. After preoxygenation, inj. Fentanyl 2µg/kg, was given, along with Inj. Labetalol 1mg/kg 90 seconds before intubation (as loading) and anaesthesia was induced with inj. propofol2mg/kg slow titrated, vecuronium bromide 0.1mg/kg, N2O : oxygen (50 : 50) and isoflurane (0.5 -1%), trachea was intubated with 5.5 mm cuffed endotracheal tube. Blood pressure surge of 180/110 mmHg intraoperatively was controlled by infusion of labetalol 0.2-2 mg/kg/hr depending on titration. A 20G

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epidural catheter was inserted by caudal route upto T10 level and 5 ml of 0.25% bupivacaine was administered after a test dose of plain lignocaine. Maintenance of anaesthesia was done with 60% N_2O in Oxygen with isoflurane and intermittent doses of vecuronium 0.02mg/kg. Intraoperative BP fluctuations during dissection and tumour manipulation were controlled with infusion labetalol .2-2 mg/kg/hr 25mg in 100 ml of 5% D i.e. 11.2 ml/hr-112 ml/hr and heart rate remained stable throughout. Removal of the tumour led to the precipitous fall of B.P. which was restored by crystalloids, colloids, noradrenaline infusion and inj. hydrocortisone2mg/kg. Total estimated blood loss was about 600 ml and total fluids administered intraoperatively were 1.5 L of crystalloid, 250 ml of blood. Urine output was monitored and maintained at 1 mlkg-1hr-1, throughout the procedure. At the end of surgery the vitals of the patient were stable on noradrenaline infusion at 0.01-0.05mcg/kg/min and respiratory efforts being adequate the extubation was planned, reversal done with inj. Neostigmine 0.05mg/kg and inj. Glycopyrrolate 0.01 mg/kg and the child was transferred to ICU for strict monitoring of blood pressure and blood glucose levels. Infusion labetalol was kept ready in case the blood pressure rose postoperatively. Analgesia was achieved with 3 ml of 0.125% bupivacaine and paracetamol suppository. On 2nd post op day, patient became hemodynamically stable, and noradrenaline infusion was tapered and discontinued. Child was shifted toward after caudal catheter removal. On 4th postoperative day, he was given oral dexamethasone 2 mg bd. The child showed improvement in symptoms and blood pressure was normalized after 10 days after which he was discharged and called for follow up.

DISCUSSION

Hypertension due to pheochromocytoma in children tends to be more sustained rather than paroxysmal in adults.[1,2] Clinical manifestations are seen due to release of epinephrine and norepinephrine resulting in constriction of arteriolar and venous segments and thereby decreasing the circulating blood volume. During attacks, the patient complains of headache, palpitation, abdominal pain, dizziness, pallor, vomiting and sweating. In severe cases, convulsions, pulmonary edema, cardiac and hepatic enlargement, hypertensive encephalopathy can occur. Child inspite of good appetite fails to gain weight. Diagnosis is usually confirmed by raised urinary catecholamine levels and their metabolites but VMA gives15% false results and measurement of 24 hour free urinary catecholamine is the best confirmatory test. In contrast to adults, the predominant catecholamine in children is norepinephrine and total urinary catecholamine excretion exceeds 300 microgram/hr. Localisation of tumour is accurately done by CTscan, MRI, MIBG.[3, 4]

Primary preoperative aim should be good pharmacological control of adverse effects of circulatory catecholamine and restoration of blood volume by ά-adreno receptor blockade,adequate control of blood pressure, heart rate and arrhythmias.β blockers are unnecessary if tumour isepinephrine secreting but required in cases of significant dysrrythmias or tachycardia.[4, 5] β-blocker should be started only after adequate alpha block as the blockade of β 2 vasodilatory receptors lead to worsening of hypertension and removal of β1 stimulation can precipitate heart failure. Here, prazosin was used preoperatively in our case for selective ά-1 blockade and propranolol for controlling tachycardia.[5]

Goals of anaesthetic management should aim at avoiding drugs or maneuvers which produce a catecholamine surge - stress, anxiety, pain, hypoxia, hypercarbia, providing optimal surgical conditions and suppression of the responses to endotracheal intubation with deep plane, surgical stimulation, tumour handling and devascularisation, maintain cardiovascular stability with short acting drugs, maintain normovolemia and hemodynamics after tumour resection.

General anaesthesia combined with regional anaesthesia is a preferred technique.[6, 7]

Pre medication should be based on preferences of the anaesthesiologist but drugs causing histamine release are to be avoided[5] - Atracurium, Morphine, Suxamethonium (produce a catecholamine surge by muscle fasciculation), Metoclopramide, Ephedrine, and Chlorpromazine (produce hypertensive response). We have used inj. Midazolam intramuscular as premedicant in our case. Monitoring (invasive blood pressure and central venous pressure) in our case could not be performed as per recommendations.[4] The anaesthetic agents preferred for induction were propofol, fentanyl, isoflurane, N_2O and labetalol to blunt response to endotracheal intubation. Isoflurane was used because it provides good relaxation and mild analgesia without sensitizing the heart to catecholamines. Vecuronium is a preferred as muscle relaxant because of its hemodynamic stability and non release of histamine.[6] Intraoperative hemodynamic variations can normally be controlled by combining regional anaesthesia with general anaesthesia using inhalational andnarcotics[7-9] but surges during tumour handling need potent vasodilators. Caudal-epidural anaesthesia with general anaesthesia was used to expand the vascular bed and provide pre-emptive and post

operative analgesia. We used infusion Labetalol, an alpha and beta adrenergic blocking drug, that produces dose-related fall in blood pressure without reflex tachycardia and without significant reduction in heart rate. The experience of others in Europe and Asia has shown labetalol to be effective in lowering blood pressure and relieving symptoms in pheochromocytoma patients[10-12] Labetalol is particularly useful when given intravenously for the rapid control of blood pressure; and has provided satisfactory coverage during surgery at the time of removal of the tumour. There have been a few reports of paradoxical hypertensive response after labetalol administration in patients with pheochromocytoma.[13-17]

Transabdominal resection is recommended in children because of extra-adrenal prevalence.[18] Post operative concerns included avoidance of hypotension, hypertension (increase 50 % for few days, as elevated catecholamine levels are present for 7-10 days) and hypoglycaemia.

CONCLUSION

Management of anaesthesia in pheochromocytoma is very challenging. A thorough etiopathological knowledge of the condition and meticulous perioperative hemodynamic control are the keystone for successful management in these cases.

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