ISSN: 2394-0026 (P)
ISSN: 2394-0034 (O)

Case Report

Bilateral Pheochromocytoma: An anesthetic challenge

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How to cite this article: Yogesh M Borse, Rashmi V. Bengali, Rajesh D. Subhedar, S.A. Thorat. Bilateral Pheochromocytoma: An anesthetic challenge. IAIM, 2015; 2(5): 200-204.

Available online at www.iaimjournal.com

Received on: 30-04-2015 **Accepted on:** 07-05-2015

Abstract

Pheochromocytoma is a rare catecholamine secreting neuroendocrine tumor. It is bilateral in 10% of cases. A 24 year lady with bilateral Pheochromocytoma operated for resection under epidural with general anesthesia. Diagnosis was confirmed with 24 hour urinary VMA levels, USG, CT scan and MIBG scan. Tumor resection was incomplete on left side so patient underwent reexploration. Preoperative BP controlled with Phenoxybenzamine and Prazocine. Intra-operative BP fluctuations managed with sodium nitropruside and esmolol. Post operative hypotension was tackled with higher doses of Dopamine and Noradrenaline due to down regulation of adrenergic receptors. Patient was discharged on oral steroids after 3 weeks.

Key words

Bilateral Pheochromocytoma, Anesthetic management, Adrenergic receptor, Down regulation.

Introduction

Pheochromocytoma is a catecholamine secreting neuroendocrine tumor that originates in adrenal medulla or chromaffin tissues along the paravertebral sympathetic chain [1]. These are rare tumors causing 0.1% of hypertensive patients. They are mostly seen in the 3rd to 5th decade of life. The “rule of 10” has been described for pheochromocytomas. 10% of tumors are extra adrenal, bilateral, malignant, and occur in children [2]. It presents most commonly as paroxysmal spells of headaches, sweating, palpitations, and hypertension. Rarely presents as pain in abdomen [3]. Early diagnosis with aggressive, management is required for reducing mortality. Anaesthetic management of pheochromocytoma is challenging task.

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Case report

A 22 year old 40 kg lady presented with headache, palpitations, sweating, pain in abdomen, easy fatigability, and irritability. On first visit her Pulse rate was 124/min and B.P. was 170/110 mm Hg. Urinary vanillylmandelic acid (VMA)/ 24 hour level was 53 mg (N< 13 mg). USG abdomen, Contrast CT Abdomen and Metaiodobenzylguanidine (MIBG) scan all were suggestive of suprarenal masses measuring 3.7x2.3 cm on Right side and 6x9 cm on Left side. (**Figure – 1, Figure - 2**) Patient put on Tab. Phenoxybenzamine 10 mg HS, Tab. Prazocin 2 mg BD, Tab. Propranalol 20 mg tds and Tab. Labetalol 100 mg BD. After one month BP came to 130/86 mm Hg, and heart rate to 90/min with no palpitations or sweating. She gained 1 kg weight and her sleep was normal with sense of wellbeing. Her Hb was 10 gm% and no albumin in urine. ECG, chest X-ray, serum electrolytes, coagulation profile, liver function tests, renal function tests and arterial blood gas (ABG) analysis were within normal limits. Fundoscopy revealed Grade II changes and 2D Echo suggestive of concentric left ventricular hypertrophy. On preoperative examination, her pulse was 84/minute, regular; BP was 130/86 in supine and 100/70 mm Hg in standing position. Phenoxybenzamine was stopped 36 hours before surgery. Tab. Nifidepin 10 mg Hs and 10 mg Diazepam was given in night and morning. Tab. Labetalol and Prazosin given 4 hours before surgery and 1000 ml Ringer lactate over 12 hours. Tab. Ranitidine 150 mg given at 7 am. Preloading was done with 200 ml Ringer. Patient premedicated with Inj. Fentanyl 50 mg, Inj. Midazolam 1.5 mg, Inj. Hydrocortisone 100 mg and Inj. Labetalol 5 mg IV. Thoracic epidural inserted and test dose of 3 ml Lignocaine 2% with 1:200000 adrenaline given. Non-invasive blood pressure (NIBP), invasive blood pressure (IBP), central venous pressure (CVP), ECG, SpO2 and temperature monitors applied.

General anesthesia instituted with Inj. Propofol 2 mg/kg slowly and intubated under effect of Inj. Vecuronium 0.1 mg/kg body weight with cuffed endotracheal tube no 7. Anesthesia was maintained on Oxygen, Nitrous oxide and isoflurane. Analgesia achieved with epidural infusion of 0.25% Bupivacaine. Vitals maintained as per following MAP - 70-90 mm of Hg, HR - 80-100/min, CVP - 4-6 mmHg. Transient hypertension 200/130 mm Hg during subcortical tumor resection on right side was present. Intra-operative BP fluctuations were managed with Inj. Sodium nitropruside (50-100 µgm/min) and Inj. Labetlol 5 mg IV. CVP maintained at 4-6 mmHg with 1500 ml Ringers solution, Haesteril 3% and blood 300 ml. Urine output was 720 ml at end of surgery. Hourly blood sugar was monitored and was between 80 to 160 mg/dl. Patient was extubated uneventfully after reversal with Inj. Neostigmine 0.04mg/kg and Inj. Glycopyrrolate 0.01 mg/kg. Inj. Buprenorphine 0.1 mg was given through epidural catheter for postoperative analgesia. Blood sugar was 84 mg%. After 30 min patient became drowsy with B.P. 160/110 mm and H.R. 100/min so kept on ventilator on SIMV+PS mode. Tab. Nifedipine through Ryles Tube was given. B.P. continued to rise so Prazosin 2 mg was given as status persisted after 1 hour. 2 hours later BP rose to 170/120 mm and HR - 100/min in spite of normal blood sugar, ABG electrolytes consciousness, no pain. B. P. was controlled with Phentolamine and Sodium nitropruside drip. Portable USG revealed 4x6 cm mass behind splenic flexure. Patient re explored and except for transient rise of BP during tumor handling, patient had uneventful course. At end of surgery, CVP 8 mm Hg, BP 110/70 mm Hg, HR 110/min without any drug was present. Overnight ventilation continued. After 6 hours systolic BP dropped to 70 mm Hg, CVP 3 mm and HR 128/min. CVP remained same for 1 hour in spite of 1 litre ringer lactate and 500 ml 3% hestar. Dopamine 10 mcg/kg/min started but

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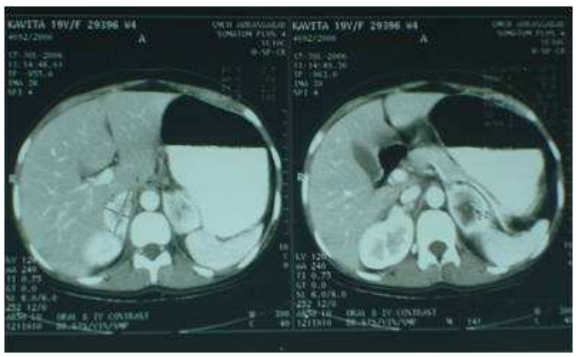
systolic BP remained 80 mm Hg. ABG was normal with S. Potassium dropped to 3 meq/lit. Noradrenalin 3mcg/min started, 300 ml blood given then BP came to 110/70 mm, CVP – 7 mm, HR - 120/min. After 3 hours SPO2 dropped to 85%, RR increased, Rhonchi appeared and CVP 12 mm, BP 80/60 mm, HR 140/min recorded. ABG showed Respiratory Acidosis with Potassium 2.5 meq/lit. Patient kept on volume control ventilation With PEEP and 100% O2. Potassium replacement done 10 meq/ hr. Dobutamine added, noradrenaline reduced to 1 micrgm/min, Inj. frusemide 80 mg given and fluids restricted to 40 ml/ hr to keep CVP 4-6 mm. DNS, potassium supplementation and hydrocortisone continued. Vitals became stable within 60 min. ECHO showed Good LV. Urine output was 1 L with CVP 8 mm and BP 110/70 mm Hg. Patient regained consciousness and ABG, BSL and serum electrolytes came to normal. Ventilation continued on SIMV with PS mode after 12 hours. Urine output was 1800 ml/24 hour. Dopamine omitted after 18 hours and extubated 24 hours after re exploration. BP was 130/100 mm so Tab. Nicardia retard 20 mg BD continued for 7 days. Hydrocortisone tapered over 5 days and patient shifted on oral steroids. Mild wound infection managed with antibiotics. She was discharged after three weeks with stable BP without anti-hypertensives.

Figure – 1: CT scan showing bilateral suprarenal mass.



Discussion

Only 10% of pheochromocytomas are bilateral. Bilateral pheochromocytomas are commonly seen in childhood but rarely seen in adults [4]. Pheochromocytomas are catecholamine secreting tumors. Adrenal pheochromocytoma predominantly secrets norepinephrine and less epinephrine, whereas extraadrenal pheochromocytoma secrets only epinephrine. Clinical manifestations like palpitations, tremors, blanching and sweating are due to paroxysmal release of epinephrine and rarely dopamine. Release of norepinephrine results in severe vasoconstriction leading to decreased circulatory volume [5]. 24 hour Urinary catecholamine estimation confirms the diagnosis, but their metabolites VMA gives 15% false positive results [6]. CT scan and MRI helps to localize the tumour. MIBG scan is useful to detect extra-adrenal sites [6, 7].

Figure – 2: MIBG scan.

Mainstay of anaesthetic management is preoperative pharmacological control of circulatory adverse effects of catecholamines and restoration of blood volume. In our case we optimized patient with selective α1 blockers like phenoxybenzamine, prazocine and α β blocker

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labetalol for 1 month. β blockers are necessary to prevent dysrhythmias and tachycardia. Phenoxybenzamine is longer acting so we stopped it 3 days prior to operation. Selective α1 blockers cause postural hypotension that should be kept in mind. Epidural with general anesthesia is preferred technique [8, 9]. Premedication agents with histamine release are to be avoided so we used fentanyl and midazolam. Successful anesthesia management depends on combating the effects of stress response, surgical stimulation, tumor handling and devascularization. We used invasive blood pressure monitor which is recommended for noting minute to minute variations [7]. Propofol , fentanyl, N_2O, and isoflurane are safe agents so we preferred these drugs. Veccuronium is preferred muscle relaxant by virtue of excellent hemodynamic stability and non release of histamine so we used it [8]. BP fluctuations can be managed with sodium nitroprusside, NTG, Phentolamine and esmolol [9, 10, 11]. Heart rate can be controlled with esmolol, labetalol and Dexmeditomidine [11]. Hemodynamic fluctuations in bilateral tumors resection are more pronounced and prolonged. We used sodium nitropruside and labetalol to tackle the fluctuation. Hypotension after excision of pheochromocytoma may not respond to IV fluids. Adreno receptor down regulation may be important factor so we required higher dose of vasopressor [9]. Critical balance of fluids and vasopressors is needed to avoid pulmonary edema. Hypokalemia in postoperative period might be due to unopposed action of insulin, stress of surgery and frusemide. Glucocorticoid and mineralocorticoid support is necessary for bilateral adrenalectomy [7]. Our patient discharged on oral steroids with written instructions for emergency management of addisonian crises.

Conclusion

The thorough understanding of pathophysiology of tumor, pharmacology of adrenergic receptor agonist and antagonists, vigilant monitoring, adequate fluid, electrolyte balance, availability of rapidly acting vasodilator and β blockers have reduced the mortality to 3%. We should confirm the size of the tumor removed. Intra-operative sonography should be considered if there is any doubt. Multidisciplinary approach is quintessential in making the anaesthesia management of Pheochromocytoma resection a safe game.

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ISSN: 2394-0034 (O)

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Source of support: Nil **Conflict of interest:** None declared.