Dexmedetomidine for anaesthetic management of phaeochromocytoma in a child with von Hippel-Lindau type 2 syndrome

INTRODUCTION

Phaeochromocytoma is a rare neoplasm in the paediatric population with fewer than 20% of diagnosed cases.[1] We describe the use of dexmedetomidine infusion for successful management of an 11-year-old male child with von Hippel–Lindau type 2 syndrome (VHL), who underwent bilateral adrenalectomy for phaeochromocytoma.

CASE REPORT

An 11-year-old boy weighing 20 kg presented with pain in the abdomen, excessive sweating and dyspnoea on exertion. Pulse rate was 120/min and blood pressure (BP) was 156/96 mm Hg. A computerized tomography of the abdomen revealed bilateral adrenal masses and a mass lesion in the head of pancreas. Plasma free metanephrine was 19.4 pg/ml (n < 90 pg/ml) and normetanephrine was drastically raised to 2488 pg/ml (normal <180 pg/ml). A diagnosis of bilateral adrenal phaeochromocytoma with pancreatic head mass in a case of VHL type 2 syndrome was made. Ophthalmological evaluation showed Grade I hypertensive retinopathy. Two-dimensional echocardiogram showed mild concentric left ventricular hypertrophy. Child was started on tablet prazocin 7.5 mg qid and amlodipine 2.5 mg bid till BP was reduced to 90/60 mm Hg followed by tablet metoprolol 50 mg bid which brought down the pulse

rate to 90/min. Oral prazocin was continued till the morning of surgery while other antihypertensives were discontinued one day prior. He was then posted for a bilateral adrenalectomy with Whipple's procedure.

Child was pre-medicated with intravenous (IV) midazolam 1 mg, IV fentanyl 2 μg/kg. We administered a loading dose of dexmedetomidine 1 μg/kg over 20 min followed by infusion of 0.3 μg/kg/h. IV hydrocortisone 2 mg/kg was administered. Pre-induction BP was 120/90 mm Hg and pulse rate was 96 beats/min. General anaesthesia was induced with IV propofol 3 mg/kg and muscle relaxation with IV vecuronium 0.1 mg/kg. Fentanyl 3 μg/kg IV and lignocaine 1.5 mg/kg IV were administered 90 s prior to tracheal intubation to blunt the sympathetic response. Anaesthesia was maintained on oxygen, air, isoflurane at a minimum alveolar concentration of 1.2. Nitroglycerin (NTG) infusion was started at 0.3 μg/kg/min. Left radial artery and right internal jugular vein were cannulated. Epidural catheter was secured at T9-T10 interspace. Following test dose of 2 ml 1% lignocaine with epinephrine, bolus of bupivacaine (0.25%) 8 ml was administered. A continuous epidural infusion of bupivacaine (0.125%) at rate of 3 ml/h was used. Intraoperative monitoring included pulse oximetry, electrocardiography, invasive BP, non-invasive BP, central venous pressure, capnometry, nasopharyngeal and skin temperature, hourly blood glucose and urine output. There was significant haemodynamic response to left sided tumour handling, which was short lived and controlled well by stepping up dexmedetomidine infusion to a maximum dose of 0.7 μg/kg/h and increasing NTG infusion to 0.7 μg/kg/min. After adrenal vein ligation, dexmedetomidine and NTG infusion were stopped. The BP fell to 60/40 mm Hg after left adrenal vein ligation which was managed with an infusion of norepinephrine and phenylephrine boluses. Hydroxyethyl starch, Ringer lactate and one packed red blood cell transfusion were used for intravascular volume expansion. Frozen section of the pancreatic head mass revealed lipid-rich clear cell pancreatic neuroendocrine tumour and bilateral adrenal masses were confirmed as phaeochromocytoma. At the end of surgery, trachea was extubated. Steroid supplementation was continued. He was later discharged with BP 100/80 mm Hg and pulse rate less than 100/min.

DISCUSSION

Paediatric phaeochromocytomas although rare, have an increased incidence of bilateral, multifocal and familial preponderance when compared to adults.[1] In children, 40–60% are inherited in an autosomal dominant pattern.[1] Hereditary phaeochromocytoma is associated with multiple endocrine neoplasia type 2, neurofibromatosis type 1, VHL syndrome, and familial paragangliomas.[2] The VHL phenotype includes phaeochromocytoma, paragangliomas, retinal angiomas, cerebellar haemangioblastoma, epididymal cystadenoma, renal and pancreatic cysts, pancreatic neuroendocrine tumours and renal cell carcinoma.[2] Symptoms include the classic triad of headaches, sweating and palpitations. Hypertension is the most common presenting sign. Measurements of free plasma metanephrines and normetanephrines are the most reliable biochemical tests available for diagnosis.[1]

Surgical resection is the treatment of choice for bilateral phaeochromocytoma. Anaesthetic challenges include the stress of intubation, surgical incision, and manipulation of the tumour mass which can precipitate a hypertensive crisis. Profound hypotension with shock can occur after tumour resection because of the rapid decrease in the level of circulating catecholamines.

The main objectives of perioperative optimization are the control of BP, control of heart rate and arrhythmias and control of secondary symptoms. This is done by α-adrenergic blockade followed by β-blockade.[1] The efficacy of therapy should be judged by the reduction of symptoms and stabilization of BP and heart rate for age and height.

Currently, intraoperative hypertension control is often attained by nitroprusside, nicardipine, NTG, magnesium, clonidine and/or by deepening the anaesthesia.[3] Epidural analgesia may blunt the sympathetic response to surgical incision. However, it does not protect against catecholamine release during tumour manipulation. Dexmedetomidine is a highly selective α2-adrenoceptor agonist having sedative and analgesic properties[3] and a novel agent for management of hypertensive surges and reducing anaesthetic requirements.

We decided that the combination of dexmedetomidine, NTG, isoflurane and epidural analgesia might make additional hypotensive agents unnecessary. Plan A was to use these agents and manage the post-resection hypotension with norepinephrine infusion as it was a primarily normetanephrine secreting tumour. Plan

B included the addition of magnesium sulphate infusion in case of failure of above agents. For control of post-resection hypotension, we proposed to use phenylephrine bolus injections and epinephrine and/or dopamine infusions in case of non-response to norepinephrine.

The combination of dexmedetomidine, NTG, isoflurane and epidural analgesia was effective to achieve safe anaesthesia in our patient. BP surges during tumour manipulation were short-lived and well controlled by merely stepping up the infusion dose of dexmedetomidine. There was no event of dangerous bradycardia. Post-adrenal vein ligation, hypotension was a concern in spite of terminating NTG and dexmedetomidine infusions. Norepinephrine infusion had to be started. Three phenylephrine boluses were required and intravascular volume status was maintained.

Similar results have been published by few groups. Bryskin and Weldon used dexmedetomidine and magnesium sulphate for the perioperative management of a child undergoing laparoscopic resection of bilateral phaeochromocytoma.[4] The use of dexmedetomidine and remifentanil for an adolescent undergoing resection of phaeochromocytoma has been successfully reported by Jung *et al*.[5] Dexmedetomidine and sevoflurane were successfully used by Khetarpal *et al*. in an adult undergoing resection of phaeochromocytoma.[6]

CONCLUSION

Dexmedetomidine based anaesthesia along with pre-operative receptor blockade may reduce the need for vasoactive drugs in the intraoperative management of phaeochromocytoma. There may be post-resection hypotension which can be safely managed with standard ionotropic agents and restoration of intravascular volume status.

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REFERENCES

1. Waguespack SG, Rich T, Grubbs E, Ying AK, Perrier ND, Ayala-Ramirez M, *et al*. A current review of the etiology, diagnosis, and treatment of pediatric pheochromocytoma and paraganglioma. J Clin Endocrinol Metab 2010; 95:2023-37.
2. Chen H, Sippel RS, O'Dorisio MS, Vinik AI, Lloyd RV, Pacak K, North American Neuroendocrine Tumor Society (NANETS). The North American Neuroendocrine Tumor Society consensus guideline for the diagnosis and management of neuroendocrine tumors: Pheochromocytoma, paraganglioma, and medullary thyroid cancer. Pancreas 2010; 39:775-83.
3. Domi R, Laho H. Management of pheochromocytoma: Old ideas and new drugs. Niger J Clin Pract 2012;15:253-7.
4. Bryskin R, Weldon BC. Dexmedetomidine and magnesium sulfate in the perioperative management of a child undergoing laparoscopic resection of bilateral pheochromocytomas. J Clin Anesth 2010;22:126-9.
5. Jung JW, Park JK, Jeon SY, Kim YH, Nam SH, Choi YG, *et al*. Dexmedetomidine and remifentanil in the perioperative management of an adolescent undergoing resection of pheochromocytoma-A case report. Korean J Anesthesiol 2012;63:555-8.
6. Khetarpal M, Yadav M, Kulkarni D, Gopinath R. Role of dexmedetomidine and sevoflurane in the intraoperative management of patient undergoing resection of phaeochromocytoma. Indian J Anaesth 2014;58:496-7.

Access this article online	
Quick response code	Website: www.ijaweb.org
	DOI: 10.4103/0019-5049.156891