International Journal of Medical Research Professionals
P-ISSN: 2454-6356; E-ISSN: 2454-6364
DOI: 10.21276/ijmrp

IJMRP

Case Report..

A Novel Anesthetic Approach To Hemodynamic Stability In Pediatric Pheochromocytoma: A Case Report

Dareen Al-amoudi[1], Amjed Shah[1], Abdulkarim Alhassoun[1], Rana Saad Alkhaibari[1], Abdullah Taiyeb[1]

[1]Department of Anesthesia, King Faisal Specialist Hospital and Research Center, Jeddah, Saudi Arabia.

ABSTRACT

We are presenting an unusual case of a nine years old boy with an extra-adrenal mass, found to be in the pelvic area at the bifurcation of the aorta and profoundly attached to the spinal body. Patient referred to our hospital for uncontrolled hypertension and tonic seizure, which required a PICU admission to control. Further investigation including ultrasound and MRI abdomen prove the presence of a mass. The diagnosis confirmed for pheochromocytoma versus Paraganglioma by measuring the free plasma metanephrine and normetanephrine concentration. After a multidisciplinary discussion, the patient was scheduled for surgical excision, despite that the patient blood pressure was poorly controlled on multiple medications including sodium nitroprusside and phentolamine infusion preoperatively. The hemodynamic was effectively controlled intra-operative by using dexmedetomidine infusion with a caudal block; this combination showed to be very effective in controlling the hemodynamics, blunting the reflexes that associated with securing the airway using laryngoscopy, surgical stress with skin incision and also, manipulation of the tumour.

Keywords: Pheochromocytoma, Pain, Pediatric, Caudal, Dexmedetomidine, Regional.

***Correspondence to:**

Dr. Dareen Al-amoudi,
Department of Anesthesia,
King Faisal Specialist Hospital and Research Center,
Jeddah, Saudi Arabia.

Article History:

Received: 12-10-2021, **Revised:** 05-11-2021, **Accepted:** 24-11-2021

Access this article online	
Website: www.ijmrp.com	Quick Response code
DOI: 10.21276/ijmrp.2021.7.6.015	

INTRODUCTION

Abdominal mass in pediatric age group could have a wide range of differential diagnosis and can be originated from any organ including the stomach, pancreas, kidney, adrenal, appendix, ovary and the biliary tree. Could be malignant or benign. Secretory or non-secreting. Concentrating on Pheochromocytoma and Paraganglioma are rare neuroendocrine tumours in children. Tumours that arise from the adrenal medulla are termed as pheochromocytoma and secreting catecholamine. While, tumours with extra-adrenal origins are called Paraganglioma, and its either secreting catecholamine or non-secreting.

The incidence of pheochromocytoma and Paraganglioma are estimated about 0.2 % of hypertensive patients; it is essential to aggressively evaluate the patient with clinically suspect symptoms because surgical extirpation is curative in more than 90% of patients and complications are lethal in undiagnosed cases. Most 85% - 90% pheochromocytomas are solitary tumours localized to a single adrenal gland, usually the right.[1] Approximately 10% of adults and 25% of children have bilateral cysts. The tumour may originate in extra-adrenal site 10%, anywhere along the paravertebral chain; however, 95% of the tumours are located in the abdomen, and a small percentage are located in the thorax, urinary bladder, or neck. The malignant spread of these highly vascular tumours occurs in approximately 10% of cases.[1]

In approximately 5% of cases, this tumour is inherited as a familial autosomal dominant trait. It may be part of the polyglandular syndrome referred to as multiple endocrine neoplasias (MEN)IIA or IIB. Type IIA includes medullary carcinoma of the thyroid, parathyroid hyperplasia, and pheochromocytoma; type IIB consists of medullary carcinoma of the thyroid, pheochromocytoma, and neuromas of the oral mucosa.[1] Pheochromocytoma may also arise in association with von Recklinghausen neurofibromatosis or von Hippel-Lindau disease (retinal and cerebellar angiomatosis).[1] The pheochromocytoma of the familial syndrome is rarely extra-adrenal or malignant. The bilateral tumour occurs in approximately 75% of cases. When these patients present with a single adrenal pheochromocytoma, the chances of subsequent development of a second adrenal pheochromocytoma are sufficiently high that bilateral adrenalectomy should be considered. Every member of a MEN family should be considered at risk for pheochromocytoma.[1]

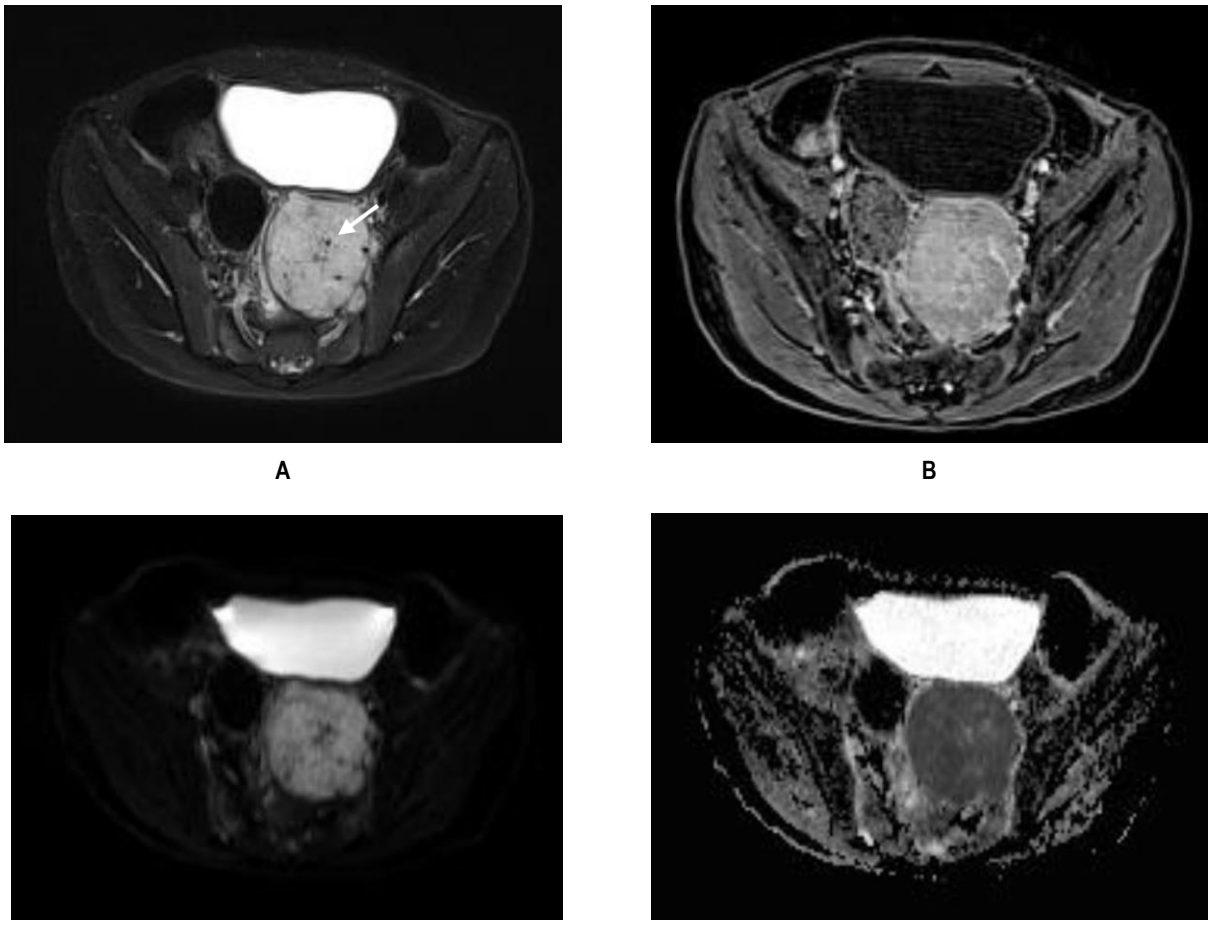
Managing a pheochromocytoma patient coming for surgery consider to be very challenging, especially in controlling the hemodynamics pre, intra and post-operatively. In our case, the tumour location also considers a risk because of its presence at the bifurcation of the aorta, which may lead to a massive bleeding intra-operative and making the hemodynamic more challenging to control.

CASE PRESENTATION

Nine years old boy, he was in his usual state of health until one month ago when he started to have an attack of seizure, headache, blurring of vision and vomiting. He turned to the emergency room where he presented with an episode of hypertension emergency (BP 225/140 mmHg) and palpitation. Brain MRI revealed a showed left side ganglionic infarct. Abdominal ultrasound showed a lesion near the common iliac vessels. Following that the patient got transfers to our hospital for further investigation and management.

When the patient presented to our hospital, the BP was reaching 200/140 mmHg, and the heart rate is 137 he required immediate admission to the PICU to control the BP. Started on antihypertensive medications which are phentolamine infusion at a rate of 4 mic/kg/min, propranolol 10 mg PO TID, amlodipine 5 mg PO BID and sodium nitroprusside infusion two mic/kg/min. Lab work was repeated including CBC, renal and liver function was normal, x-ray chest shows no signs of infiltration, ECG was normal except for sinus tachycardia HR > 120 bpm.

MRI abdomen, spine and ultrasound abdomen all are done, and it is become positive for an intraabdominal mass, well defined, encapsulated in the Para-sacral region attached to the spinal body at the level of the bifurcation of the aorta measuring 5.5 cm. The diagnosis for pheochromocytoma versus Paraganglioma confirmed by measuring the free plasma metanephrine and normetanephrine concentration. Pediatric surgery evaluated the patient, and after a multidisciplinary meeting, the decision was made to proceed for surgical resection of the tumour.



A **B**

C **D**

Figure 1. Extra-adrenal pheochromocytoma in a 9-year-old boy (a) Axial T2-weighted fat- saturated image demonstrates presacral hyperintense mass with central foci of low signal intense flow void (arrow), (b) Axial T1-weighted VIBE sequence fat-saturated post contrast image demonstrates avid enhancement, (c) Axial diffusion-weighted image demonstrates high signal intensity with corresponding low signal intensity in apparent diffusion coefficient (d) in keeping with diffusion restriction.

Despite trying to control blood pressure with alpha and beta blocker and preparing the patient for surgery, the blood pressure was not controlled. After knowing the patient weight, which is 20.7 kg, height 112 & BMI of 16.5. The anaesthesia plan to give him an anxiolytic using midazolam 2 mg IV and dexmedetomidine infusion at a rate of 1 mic/kg/hour for 10 minutes using intravenous access to decrease the fluctuation in the hemodynamic during induction of anaesthesia and intubation of the trachea using laryngoscope. American Society of Anesthesiologist (ASA) standard monitors was connected and displayed HR 135/min, BP 130/75 mmHg and spo2 100%. Ringer lactate infusion started at a rate of 10 ml/kg. He was

preoxygenated with 100% oxygen for 3 minutes, patient was induced with fentanyl 4 mic/kg IV, lidocaine 1% 1.5 mg/kg IV, propofol 3-5 mg/kg IV, and rocuronium 0,5 mg/kg IV followed by smooth laryngoscopy and quick securing of the trachea with 6 mm cuffed endotracheal tube. Triple lumen central venous catheter was inserted using ultrasound-guided in the right internal jugular. The right radial artery cannulated with 22 G catheter for continuous intra-arterial pressure monitoring and frequent sample, followed by a single shot caudal anaesthesia with (bupivacaine 0,25% 20ml plus 20 mics of Dexmedetomidine). Sevoflurane used as an inhalation agent to maintain surgical anaesthesia. Magnesium sulfate 10%, 10 ml IV 500 mg, calcium chloride 400 mg 100 mg/ml solution IV was given. Dexmedetomidine infusion for the whole surgery at a rate of 0.5 mic/kg/hour IV was running. About 1800 ml of crystalloid fluid was infused before ligation of the tumour, and total urine output is 500 ml. The hemodynamic status was stable without the need for any vasoactive drugs even during mass manipulation. At the end of the operation, the patient was extubated and found to be alert, pain-free and obeying commands. Blood gas analysis was in the normal range. Post-operatively he was shifted to the PICU for close monitoring and pain control.

DISCUSSION

Catecholamines secreted tumour are rare tumours in pediatric age group, and its ether pheochromocytomas that arise from the Chromaffin cells in the adrenal medulla or extra-adrenal origins is termed paragangliomas, that arise from the paraganglia of the sympathetic chain of the nerves system.

Extra-adrenal paragangliomas either functional or nonfunctional, the functional tumour is a hormonally active that secreting of catecholamines hormone.[2,3] This activity can lead to a life-threatening situation due to the cardiovascular, neurological or metabolic crisis. Clinical manifestation is seen due to the high level of circulatory catecholamine that is resulting in arteriolar and venous vasoconstriction that leads to hypertension in 60-90% , hypertension its typically sustained in children rather than paroxysmal in adult.[4] It can be complicated as Malignant hypertension with its associated risk of (e.g., increased intracranial pressure and encephalopathy), headache in 50-80% , Episodic sweating, tachycardia, or palpitations in 50 to 60 % , abdominal pain 30%, Other symptoms and signs that occur less frequently include constipation, dizziness, blurred vision, papilledema, pallor and in severe cases Convulsion pulmonary oedema and cardiomyopathy can occur.[3,5]

Importantly, attention deficit hyperactivity disorder (ADHD) is more common in children with pheochromocytoma and Paraganglioma than in the general population in pediatric patients with hypertension and ADHD symptomatology[3,6], an evaluation to rule out pheochromocytoma or Paraganglioma is warranted prior to treatment with stimulant medications, which may exacerbate hypertensive crises.[6]

It is clinically silent In 13 % of the patients in spite of high levels of circulating catecholamines due to receptor ‘downregulation’ this clinically silent leading to lethal intraoperative complications and outcomes with the being of tumour manipulation during surgery.[2]

Surgical resection for Paraganglioma is considered to be very challenging for the anesthesiologist, especially in controlling the hemodynamics pre, intra and post-operatively.[7]

Primary Preoperative main objective is optimization and managing the patient hemodynamics this done by pharmacological control, alpha-adrenergic blockade and volume expansion that aimed at minimizing the physiologic impact of catecholamine release.[8]

In the case of dysrhythmia and tachycardia, Beta-adrenergic blockade can be used After adequate alpha-adrenergic, Beta-adrenergic blockade should never be started first as it will result in blockade of vasodilatory peripheral beta-adrenergic receptors with unopposed alpha-adrenergic receptor stimulation can lead to a further elevation in blood pressure.[4]

The efficacy of therapy should be judged by the hemodynamic stabilization for age and reduction of symptoms.[4]

Anaesthetic challenges include precipitate of hypertensive crisis with intubation, painful stimulation with surgical incision and surgical manipulation of the tumour and in the other hand hypotension with shock can occur after tumour resection because of the rapid decrease in the catecholamines level in the blood. Using of short-acting drugs for maintaining patient hemodynamic stable before and after tumour resection.[4,7]

Goal of anesthesiologist to avoid any medication or maneuvers which produce a catecholamine surge e.g. stress, anxiety, pain, hypoxia and hypercapnia[4], that could be avoidable by using premedication anxiolytic and optimal surgical condition and suppression of the response to endotracheal intubation with increase the depth of anaesthesia and tight pain control using regional technique are measure taken to prevent wide swings in blood pressure changes[4,9], maintaining patient hemodynamic using of intraoperative short-acting vasoactive drug at time of the tumour handling and devascularization.[4]

Combined of general anaesthesia with regional (caudal) Is a preferred technique for intra and post op. pain management.[10]

Premedication anxiolytic using midazolam and Dexmedetomidine started preoperative to provide sedative effect especially in a pediatric patient, ketamine should be avoided since it can cause histamine release.[4] The drug that causing histamine release is ketamine, atracurium, morphine and suxamethonium. Metoclopramide, ephedrine and chlorpromazine these can produce catecholamine surge should not be given.[4]

Dexmedetomidine is highly selective α2-adrenoceptor agonist having sedative and analgesic properties that reducing anaesthetic requirements. And for management of hypertensive surges.[9] And Intravascular Dexmedetomidine infusion for the whole surgery at a rate of 0.5 mic/kg/hour IV was running.

The general anaesthesia induction with lidocaine, fentanyl, Propofol, rocuronium followed by smooth intubation.

Rocuronium provides more suitable conditions for rapid tracheal intubation did not result in significant changes in plasma histamine concentrations has less effect on mean arterial blood pressure and heart rate.[11] Inhalational anaesthesia Sevoflurane it depresses sympathetic neurotransmission by reducing norepinephrine release, and sensitivity from the omental vessel make it a suitable inhalation agent for pheochromocytoma.[8]

Followed by a single shot caudal anaesthesia with (bupivacaine 0,25% 20ml plus 20 mics of Dexmedetomidine). Caudal epidural analgesia is one of the commonly performed regional blocks in pediatric age group, 0ne of the limitations of caudal anaesthesia is the short duration of action.

With a single injection of local anaesthetic solution that can be overcome by using a caudal catheter to infusing local anaesthesia

or repeating dose but with the risk of infection make it less favored, a mixture of 0.25% bupivacaine with 0.5–1 μg/kg dexmedetomidine has shown to increase duration and quality of caudal analgesia and required significantly less number of rescue analgesics as compared to plain bupivacaine The side effects of neuraxial dexmedetomidine administration include hypotension and bradycardia. Intrathecal or epidural Dexmedetomidine the mechanical of analgesic action its results from direct stimulation of pre- and post-synaptic α2 adrenoreceptors in the dorsal grey matter of spinal cord and inhibiting the release of nociceptive neurotransmitters.

This effect correlates with the concentration of Dexmedetomidine in the cerebrospinal fluid but not that in the plasma.[10,12]

For intraoperative blood pressure control Magnesium sulfate 10%, 10 ml IV 500 mg, was given. Magnesium sulfate can control hemodynamic disturbances by decreasing catecholamine release and reducing peripheral resistance on an arteriolar but with minimal effects on venous or pulmonary capillary wedge pressure. As will has the ability to antagonist the catecholamine-induced arrhythmias.[5]

Various multimodal techniques have been designed to achieve safe anaesthesia in our patient; the combination of sevoflurane, Magnesium sulfate, Dexmedetomidine and caudal analgesia was effective.

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 Magnesium sulfate and dexmedetomidine infusions. Hemodynamic status was stable without the need for any vasoactive drugs. At the end of the operation, and intravascular volume status was maintained.

What we did different in this case is using of dexmedetomidine as an infusion peri, intra, and post-operatively with a single shot caudal anesthesia. We found it's a very effective in controlling the hemodynamic and blunting any sudden elevation in the blood pressure that associated with the airway instrumentation, and especially during manipulating the tumor. Using dexmedetomidine intra-operatively with good optimization before the surgery can reduce the need for vasodilators drug.

REFERENCES

1. Barash PG CB, Stoelting RK, Cahalan MK, Stock MC, Ortega R Clinical anesthesia: Seventh edition: Wolters Kluwer Health, 2013. 1792 p.

2. Tomulic K, Saric JP, Kocman B, Skrtic A, Filipcic NV, Acan I. Successful management of unsuspected retroperitoneal paraganglioma via the use of combined epidural and general anesthesia: a case report. Journal of medical case reports. 2013;7(1):58.

3. William F Young J, MD, MSc. Pheochromocytoma and paraganglioma in children. In: Alberto S Pappo MMEG, MD Alison G Hoppin, MD, editor. Waltham, MA: UpToDate.

4. Jain S, Vats A, Arora K. Anaesthetic management of pheochromocytoma in a child. Central Journal of ISA. 2017;1(1):36-8.

5. James MF, Cronjé L. Pheochromocytoma crisis: the use of magnesium sulfate. Anesthesia & Analgesia. 2004;99(3):680-6.

6. Batsis M, Dagalakis U, Stratakis C, Prodanov T, Papadakis G, Adams K, et al. Attention deficit hyperactivity disorder in pediatric patients with pheochromocytoma and paraganglioma. Hormone and Metabolic Research. 2016;48(08):509-13.

7. Khetarpal M, Yadav M, Kulkarni D, Gopinath R. Role of dexmedetomidine and sevoflurane in the intraoperative management of patient undergoing resection of phaeochromocytoma. Indian Journal of Anesthesia. 2014;58(4):496.

8. Khetarpal M, Kulkarni D, Gambhir R, Rao SM. The effective use of sevoflurane during resection of phaeochromocytoma. Ind J Anaesth. 2005;49(2):137-9.

9. Dias R, Dave N, Garasia M. Dexmedetomidine for anaesthetic management of phaeochromocytoma in a child with von Hippel–Lindau type 2 syndrome. Indian journal of anaesthesia. 2015;59(5):319.

10. Goyal V, Kubre J, Radhakrishnan K. Dexmedetomidine as an adjuvant to bupivacaine in caudal analgesia in children. Anesthesia, essays and researches. 2016;10(2):227.

11. Elbradie S. Neuromuscular efficacy and histamine-release hemodynamic changes produced by rocuronium versus atracurium: a comparative study. J Egypt Natl Canc Inst. 2004;16(2):107-13.

12. Fukushima K. The effect of epidurally administered dexmedetomidine on central and peripheral nervous system in man. Anesth analg. 1997;84:S292.

Source of Support: Nil. **Conflict of Interest:** None Declared.

Cite this article as: Dareen Al-amoudi, Amjed Shah, Abdulkarim Alhassoun, Rana Saad Alkhaibari, Abdullah Taiyeb. A Novel Anesthetic Approach To Hemodynamic Stability In Pediatric Pheochromocytoma: A Case Report. Int J Med Res Prof. 2021 Nov; 7(6): 71-74. DOI:10.21276/ijmrp.2021.7.6.015