Anesthesia Essays and Researches
Home
Current issue
Instructions
Submit article

Anesth Essays Res. 2016 Jan-Apr; 10(1): 107–110. PMCID: PMC4767069
doi: 10.4103/0259-1162.164730

Anesthetic management of a rare case of extra-adrenal pheochromocytoma

S. D. Pratibha, Vijay Katti, and Basvaraj Patil

Department of Anaesthesiology, BLDEU, Bijapur, Karnataka, India
Corresponding author: Dr. S. D. Pratibha, House No. 13, Behind KHB Colony, Sadashiv Nagar, Sholapur Road, Vijayapur, Bijapur, Karnataka, India. E-mail: pratibhakaradi@gmail.com

Abstract

Anesthetic management of pheochromocytoma is complicated and challenging. Extra-adrenal pheochromocytoma is a rare neuroendocrine tumor that produces, stores and secretes catecholamines. The main-stay in the management of pheochromocytoma surgeries is Preoperative preparation which has improved perioperative outcome. Modern anesthetic drugs with advanced monitoring have contributed to intraoperative stability. Resection of the tumor results in acute withdrawal of catecholamines, which may lead to severe hypotension. In perioperative period, adequate hydration should be maintained. Beta-blockers, nitroglycerine, sodium nitroprusside and phenylephrine are required to avoid hemodynamic fluctuations and should be used appropriately.

Keywords: Adrenergic beta antagonists, anesthesia, catecholamines, pheochromocytoma

INTRODUCTION

Pheochromocytoma is a neuroectodermal tumor arising from chromaffin cells of sympathetic nervous system, 90% of cases arises from adrenal medulla, but can be found anywhere chromaffin tissue exists. Pheochromocytomas are extra-adrenal in 10% of cases and have malignant potential in up to 40%.[1] Extra-adrenal pheochromocytoma is a rare neuroendocrine tumor that produces stores and secretes catecholamines.[2] It may arise in any portion of paraganglion system, though they most commonly occur below the diaphragm, usually in the organ of Zuckerkandl. These locations extend from the base of the skull to the anus, commonest site of extra-adrenal pheochromocytoma is a para-adrenal area, they can also occur at the aortic bifurcation, heart, brain, inferior mesenteric and iliac arteries. They are rarely associated with familial and hereditary pheochromocytoma except in Carney's syndrome. They pose the most fluctuating clinical course during anesthesia and surgical intervention. Cardiac arrhythmias and hypertensive crisis may occur during surgery due to high levels of circulating catecholamines. Clinical presentation, diagnosis and treatment are similar to adrenal tumors, but few associated medical conditions may pose a threat to life during the intraoperative period. Pheochromocytomas present with classical triad of a headache, palpitations and sweating. This is a case report of a patient with pheochromocytoma located anterior to inferior venecava (IVC) and just at the aortic bifurcation. We describe a successful management of a case of extra-adrenal pheochromocytoma posted for excision of the tumor. Anesthetic and surgical literature for pheochromocytoma is derived from case reports and small case series. Thus evidence from randomized prospective controlled trials is limited.

CASE REPORT

A 14-year-old male patient weighing 36 kg, height 150 cm and body mass index-16 was diagnosed as extra-adrenal pheochromocytoma.

Clinical presentation

The patient reported to the hospital with complaints of a headache, vomiting and weakness since 5 months which was on and off. The patient had a history of weight loss. No history of palpitations and syncopal attacks. On examination, pulse rate was 120 beats/min, blood pressure (BP) was found to be 160/110 mmHg in supine and 150/100 mmHg in standing position. Clinically Airway assessment, cardio-vascular system, respiratory system, per abdomen and central nervous system examinations were normal.

Differential diagnosis

Anxiety disorders, hyperthyroidism, hypoglycemia, insulinoma, paroxysmal supraventricular tachycardia.

Investigations

Revealed Hb - 12.4 g%, total white blood cell count - 12,200 cells/cumm, platelet count - 2.25 lakh/cumm, random blood sugar - 116 mg/dl, serum creatinine - 0.6 mg/dl, serum sodium - 140 mEq/L, serum potassium - 5.4 mEq/L, serum calcium 9 mg/dl. Chest X-ray and thyroid function tests were normal.

Electrocardiogram (ECG): Sinus tachycardia with tall T waves. Echocardiography showed concentric left ventricular hypertrophy, no valvular pathology, no cardiac shunt lesion, normal pulmonary artery pressure, left ventricular ejection fraction - 60%.

Ultrasonography abdomen revealed heterogenous echotexture mass lesion measuring 48 mm × 45 mm × 30 mm with cystic areas noted in right pre- and the para-vertebral region just above the umbilicus suggestive of pheochromocytoma. Computed tomography abdomen showed well defined heterogenously enhancing retroperitoneal para-aortic soft-tissue density mass lesion with nonenhancing central hypodense areas, anterior to IVC at the level of inferior mesenteric artery origin and just at the aortic bifurcation showing evidence of extra-adrenal pheochromocytoma.

Urinary metanephrine: 702.72 μg/24 h (normal: 32–167 μg/24 h).

Preoperative management

Patient was started on tablet prazosin 2.5 mg OD for 4 weeks and tablet atenolol 25 mg OD for 3 weeks.

On the day before surgery, pulse was 84 beats/min and BP - 120/80 mmHg. Tablet alprazolam 0.5 mg, tablet prazosin and tablet atenolol were continued till the morning of surgery. Written informed consent was taken and adequate blood was arranged.

Intraoperative management

Combined general and epidural anesthesia was planned. Anesthetic agents and other drugs like injection sodium nitroprusside (SNP), nitroglycerine (NTG), phenylepherine, esmolol, isoprenaline and amiodarone were kept ready. Noninvasive BP, pulse oximetry, ECG and capnography were connected. Before induction vital parameters recorded-BP was 110/70 mmHg and heart rate was 98 beats/min. The peripheral line was secured in the left forearm with 18 gauge cannula. The patient was premedicated with intravenous (IV) fentanyl 20 μg, ondansetron 2 mg and midazolam 1 mg. central venous pressure (CVP) line was secured in right internal jugular vein. Epidural catheter with 18 gauges was inserted at L1-L2 space. After preoxygenation for 3 min patient was induced with IV injection propofol 80 mg, injection vecuronium 5 mg were given. Injection lidocaine 100 mg preservative free was given 1 min before initiating direct laryngoscopy, injection esmolol 20 μg IV given 30 s before intubation to attenuate the pressor response and intubated with 7 mm cuffed oral endotracheal tube. Vital parameters were maintained. SNP 100 μg/h infusion was started after intubation. Anesthesia was maintained with isoflurane, oxygen and nitrous oxide and injection vecuronium. 4 ml of injection bupivacaine 0.25% and injection fentanyl 2 μg/kg was given through epidural route. During surgery, CVP increased from 6 to 12 cm of H_2O with fluid infusion 10–15 ml/kg/h of crystalloids and colloids. During dissection of tumor, BP increased to 225/187 mmHg, which was controlled with an additional dose of SNP 0.5 mcg/kg/min and injection nitroglycerine 0.5 mcg/kg/min infusion [Figure 1]. Just before ligation of the tumor vasodilators were tapered to avoid precipitous fall in BP. There were minimal fluctuations in BP except after ligation of the tumor BP decreased to 70/40 mmHg which was treated with injection phenylephrine 2 mcg/kg. Urine output was adequate and temperature was normal [Figure 2]. Intraoperative blood glucose and arterial blood gas were normal. Surgery lasted for 2 h.

Patient was extubated after giving reversal with IV injection neostigmine (1.5 mg) and injection glycopyrrolate (0.2 mg) patient was fully awake and was shifted to Surgical Intensive Care Unit and vitals were monitored.

Postoperative period

Epidural top up doses was given with injection bupivacaine 0.125% for postoperative analgesia, the postoperative period was uneventful [Table 1]. No neurological deficits were observed in the postoperative period. Later the patient was discharged from the hospital after 1-week. The histopathological report confirmed the diagnosis of pheochromocytoma.

DISCUSSION

Pheochromocytomas are important to anesthesiologist because 25–30% of hospital deaths occur in patients during induction of anesthesia or during an operative procedure. Intubation, laryngoscopy, pneumoperitoneum and manipulation of the adrenal gland can induce catecholamine secretion and consequently lead to hypertensive crisis.[3] Resection of the tumor results in acute withdrawal of catecholamines which leads to hypotension causing reduced plasma volume expansion due to chronic vasoconstriction.[4] In our case, we used fluids and phenylephrine during severe hypotension to maintain adequate hydration. Plasma expanders are preferred than crystalloids. Beta blockers, NTG, SNP, phenylephrine should be used to avoid hemodynamic fluctuations. Estimated prevalence among the hypertensive population who are diagnosed as pheochromocytoma is 0.1–0.6%.[5] Undiagnosed pheochromocytoma bears high perioperative mortality of 27%. Operative mortality was 30–40% before the need for preoperative evaluation and preparation was appreciated.[4] Pheochromocytoma with adrenal localization is observed in fourth and fifth decades and mostly second in females whereas with extra-adrenal localization found in second and third decades and mostly in males.[6]

Currently, the test of choice in imaging is I-labeled meta-iodobenzylguanide scintigraphy to overcome the limitations of anatomic imaging. For diagnostic imaging combining PET with CT scan will provide a definitive tool in near future. Combined methods will increase the accuracy of tumor localization and be useful in monitoring the response to treatment. Recent recommendations in testing are a measurement of catecholamine metabolites in plasma, urine or both which gives 100% sensitivity whereas, urinary vanillylmandellic acid assays are only 64% sensitive.[7]

In extra-adrenal pheochromocytoma, symptoms can be divided into two types. The first type symptoms are due to an excess of catecholamine and are similar to their adrenal counterpart. The second type symptoms are due to the specific location of the tumors and may help in localizing the tumors. When β-blockade is needed to control symptoms, prazosin, terazosin or doxazosin can be used to circumvent some of the disadvantages of nonselective β-blocker.[8] In our case, prazosin an α-blocker that does not produce reflex tachycardia and has shorter half-life was used which will contribute to less hypotension in pre- and post-operative period. Use of α-blockers in the preoperative period and during preparation phase before the procedure decreased pheochromocytoma resection related mortality.[9] Phentolamine a nonselective α-adrenergic blocker can be used which can control the paroxysmal rise in BP which can be due to the stress of anesthesia and during tumor resection.

Preoperative assessment is the corner stone in management of pheochromocytoma and was done prior to surgery according to Roizen's Criteria:

- BP < 160/90 mmHg for 24 h before surgery
- No orthostatic hypotension with BP < 80–45 mmHg
- ECG should be free of any ST-T changes for a week and
- No PVC's more than 1 in 5 min.

Drugs to be avoided in patients with pheochromocytoma are cocaine, droperidol, ketamine, ephedrine, halothane, metoclopramide, morphine, succinylcholine, pancuronium, curare and atracurium. If neurological deficits occur in the postoperative period, it can be attributed to exposure of the patient to paroxysmal rise in BP leading to high cerebral perfusion pressure and cerebrovascular resistance. Chronic exposure to catecholamines leads to receptor down regulation in which case vasopressors may be useful because its

effects are mediated through noncatecholamine. Lifelong follow-up is advised as extra-adrenal pheochromocytoma may recur and metastasize. Persistent hypertension after removal occurs approximately in 25% of cases, this is probably due to the residual tumor, metastatic disease or intraoperative injury to the renal artery or the kidney. Plasma catecholamines or urinary metanephrines should be measured 2 weeks after surgery and every 3 months for 1st year then annually even in normotensive patients. BP should be monitored at monthly intervals for 1st year and then twice a year thereafter. A critical innovation that has created the opportunity for progress in anesthetic management is used of electronic automatically recorded BP and heart rate measurements by anesthesia information management systems to study intraoperative hemodynamics during pheochromocytoma resection. This dense data collection will allow for careful definitions of "BP variability" so that larger case series between institutions may be effectively compared.

CONCLUSION

The management of patients with extra-adrenal pheochromocytoma remains a challenge for the anesthesiologist, despite the advent of new drugs and techniques. Our role in the successful outcome of such surgeries begins from adequate preoperative preparation, intense intraoperative monitoring and careful follow-up during the postoperative period. Prognosis is usually good if the tumor is detected early to avoid major complications related to catecholamine excess.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest

REFERENCES

1. Emerson CE, Rainbird A. Use of a 'hospital-at-home' service for patient optimization before resection of phaeochromocytoma. Br J Anaesth. 2003;90:380–2. [PubMed: 12594154]

2. Prys-Roberts C. Phaeochromocytoma – recent progress in its management. Br J Anaesth. 2000;85:44–57. [PubMed: 10927994]

3. Tauzin-Fin P, Sesay M, Gosse P, Ballanger P. Effects of perioperative alpha1 block on haemodynamic control during laparoscopic surgery for phaeochromocytoma. Br J Anaesth. 2004;92:512–7. [PubMed: 14766711]

4. Bajwa SS, Bajwa SK. Implications and considerations during pheochromocytoma resection: A challenge to the anesthesiologist. Indian J Endocrinol Metab. 2011;15(Suppl 4):S337–44. [PMCID: PMC3230104] [PubMed: 22145137]

5. Bruynzeel H, Feelders RA, Groenland TH, van den Meiracker AH, van Eijck CH, Lange JF, et al. Risk factors for hemodynamic instability during surgery for pheochromocytoma. J Clin Endocrinol Metab. 2010;95:678–85. [PubMed: 19965926]

6. Shinn HK, Jung JK, Park JK, Kim JH, Jung IY, Lee HS. Hypertensive crisis during wide excision of gastrointestinal stromal cell tumor (GIST): Undiagnosed paraganglioma – A case report. Korean J Anesthesiol. 2012;62:289–92. [PMCID: PMC3315663] [PubMed: 22474560]

7. Chen H, Spiel RS, O'Dorisio MS, Vinik AI, Lloyd RV, Pacak K, et al. 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. [PMCID: PMC3419007] [PubMed: 20664475]

8. Sherif L, Hegde R, Shetty K, Gurumurthy T, Jain P. Anesthetic management of a rare case of extra adrenal pheochromocytoma – A case report. Internet J Anesthesiol. 2008;21:1–21.

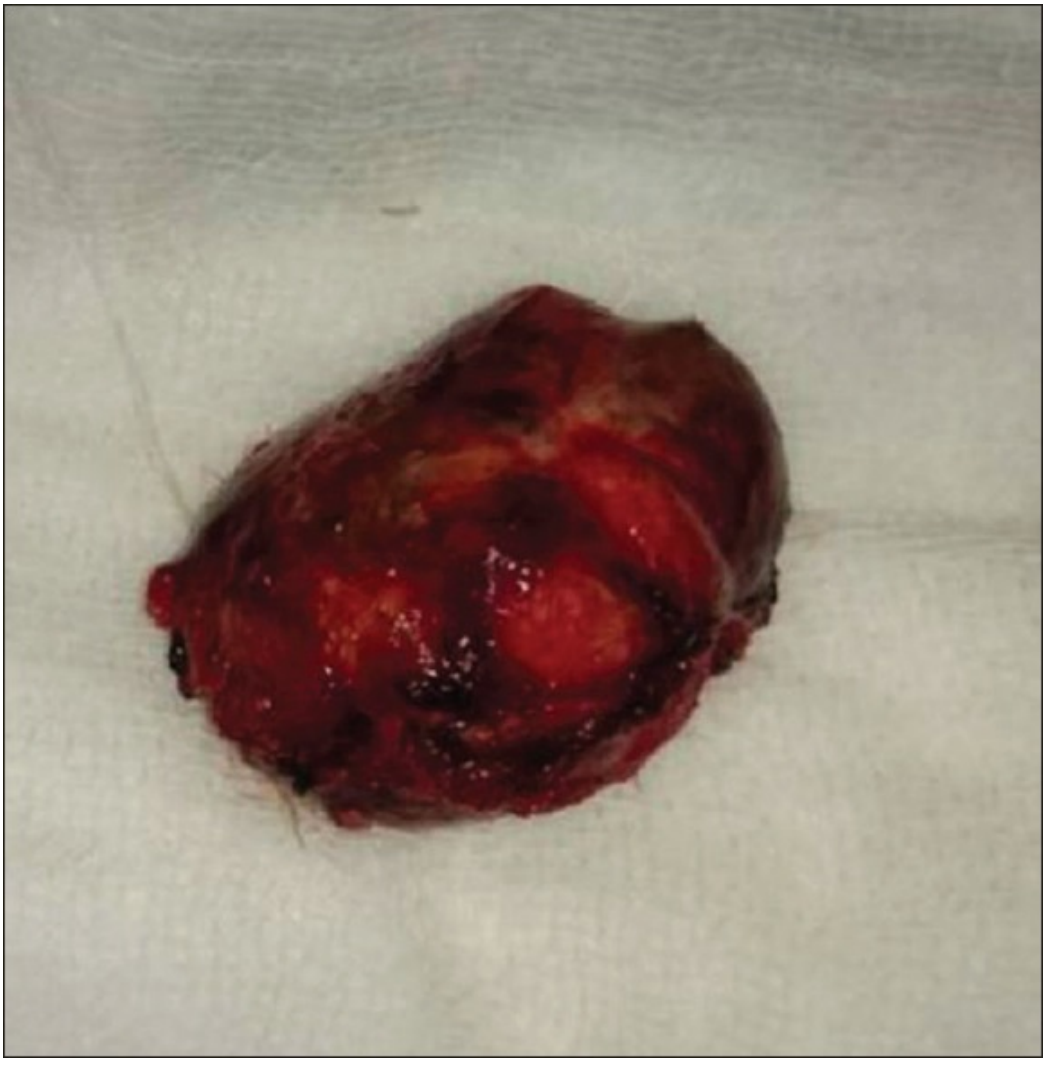
9. Lentschener C, Gaujoux S, Tesniere A, Dousset B. Point of controversy: Perioperative care of patients undergoing pheochromocytoma removal-time for a reappraisal? Eur J Endocrinol. 2011;165:365–73. [PubMed: 21646289]

Figures and Tables

Figure 1

Parameters during dissection

Figure 2



Tumor after dissection

Table 1

Case report	Present article	Reference article
	Anesthetic management of a rare case of extra-adrenal pheochromocytoma	Sherif *et al.* Anesthetic management of a rare case of extra adrenal pheochromocytoma - A case report. The Internet Journal of Anesthesiology. 2008 Volume 21 Number 1
Anesthetic drugs	Glycopyrrolate, midazolam, propofol, fentanyl, xylocard, esmolol, isoflurane, vecuronium	Midazolam, xylocard morphine, thiopentone sodium, fentanyl, propofol, vecuronium
Vasodilators/ vasopressors	SNP, nitroglycerine, phenylephrine	Halothane, SNP, NTG
Monitoring	NIBP, pulseoximetry, ECG and capnography	IBP, SpO_2, ECG
Postoperative analgesia	Epidural top up doses with bupivacaine 0.125%	Epidural top up doses with bupivacaine

SNP=Sodium nitroprusside, NTG=Nitroglycerin, ECG=Electrocardiogram, NIBP=Noninvasive blood pressure, IBP=Invasive blood pressure

Comparison of the present article with a published case report