Case Report

Anesthetic Management of a Patient with Undiagnosed Paraganglioma

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Abstract

Pheochromocytomas are catecholamine secreting tumours that arise from the chromaffin cells located within the adrenal medulla. Paragangliomas are also catecholamine secreting tumours arising from extra-adrenal chromaffin cells located along the sympathetic paravertebral ganglia of the pelvis, abdomen, and thorax. The anaesthetic management of both Pheochromocytoma and paraganglioma are same. The anaesthetic care of patients with pheochromocytoma and paraganglioma (PPGL) presenting for surgical resection presents a challenge for the anaesthesiologist, but this challenge will be greatly increased when the tumour is unexpected.

Keywords: Nitroprusside, paraganglioma, pheochromocytoma, undiagnosed

Introduction

Anesthesia and surgery in unsuspected pheochromocytoma patients have a high incidence of mortality.[1] Most of the extra-adrenal paraganglioma are rarely diagnosed preoperatively for their rarity and nonspecific signs and symptoms.[2]

The unexpected encounter of paraganglioma in the operating theaters, delivery rooms, and other circumstances carries a mortality rate of 25%–50%. In this case, the patient presented with an undiagnosed extra-adrenal pheochromocytoma, which led to a life-threatening hypertensive crisis at the time of tumor resection.

Case Report

A 20-year-old male patient presented with an intra-abdominal swelling provisionally diagnosed as retroperitoneal tumor was posted for explorative laparotomy and resection. Preoperative laboratory investigations were within normal limits.

On physical examination, the height of the patient was 167 cm and weight was 68 kg (body mass index 24.1). Airway assessment was found to be normal. On auscultation, bilateral lungs were clear, with normal heart sounds. ECG showed normal sinus rhythm. The preoperative blood pressure (BP) was 110/60 mmHg, and the heart rate (HR) was 60 beats/min (bpm). Other system examinations were found to be normal.

Computed tomography abdomen showed a solid mass in paraaortic area at the site of aortic bifurcation [Figures 1-4]. Provisional diagnosis of a retroperitoneal tumor was made.

The patient was taken up for surgery under general anesthesia with epidural analgesia after written informed consent. Standard monitoring with electrocardiogram, pulse oximetry, and noninvasive BP was done (PR 66/min, BP 120/78 mmHg, and SPO_2 99%). 20G epidural catheter was placed in T9-T10 space. The patient was premedicated with injection glycopyrrolate 0.2 mg, fentanyl 100 µg, and midazolam 1 mg intravenous (IV). Anesthesia was induced with propofol 2 mg/kg, vecuronium bromide 1 mg/kg IV, sevoflurane 1.5%, and nitrous oxide and oxygen. Patient intubated orotracheally with 8.5-sized cuffed endotracheal tube. Anesthesia was maintained with sevoflurane (1.5%–2%), nitrous oxide and oxygen (2:1), and muscle relaxation

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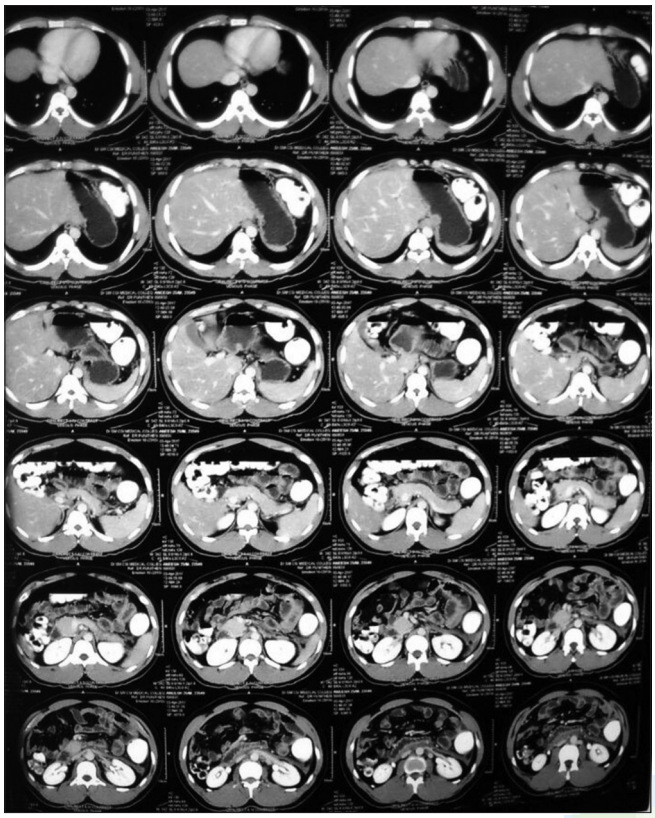

Figure 1: Computed tomography abdomen-Axial view

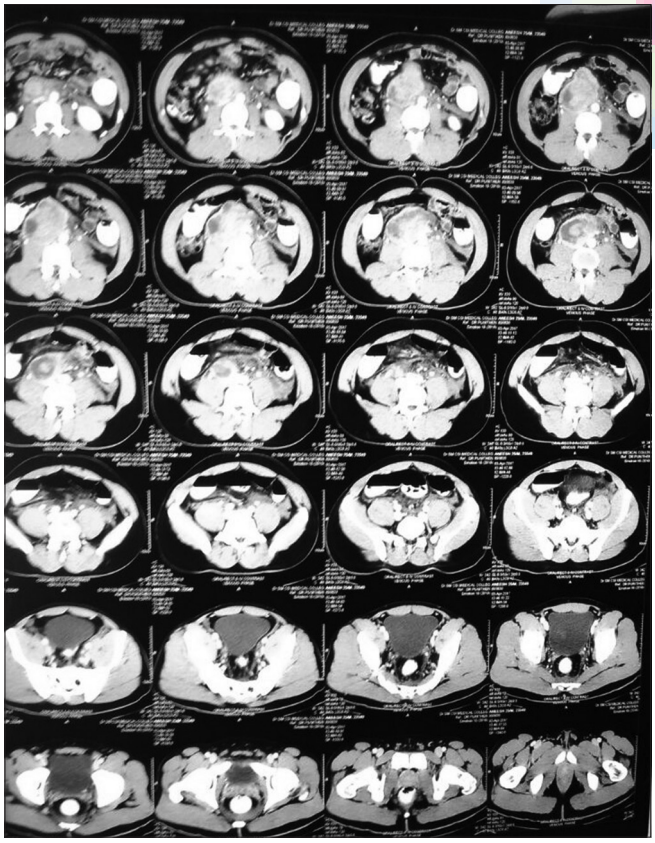

Figure 3: Computed tomography abdomen

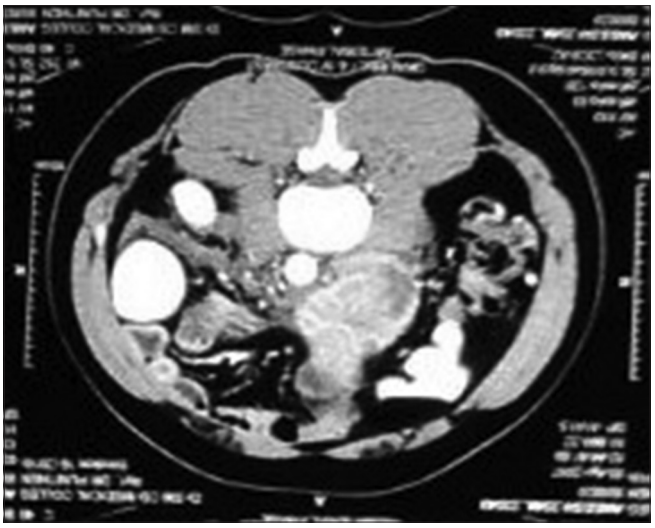

Figure 2: Computed tomography solid cystic mass in paraaortic region

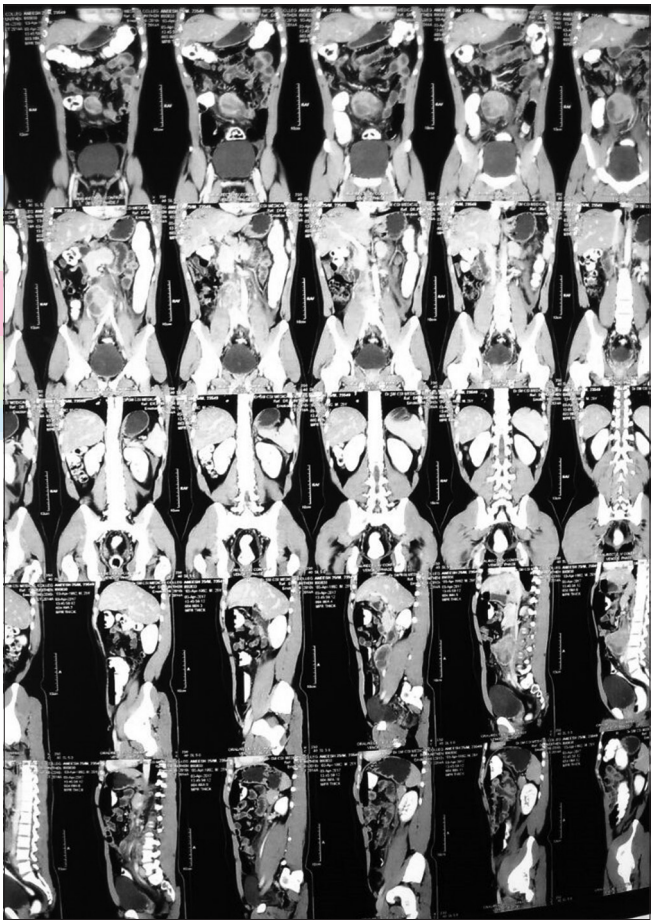

Figure 4: Computed tomography abdomen-sagittal and coronal view

maintained with vecuronium bromide IV. Initially, for the first 20 min of the surgical procedure, the vitals remained stable. However, during manipulation of the retroperitoneal mass, the BP raised to 170/100 mmHg. Anesthesia was deepened by increasing sevoflurane to 3%. Graded epidural

with 1% Xylocard (lignocaine) was given up to a dose of 10 ml over 10 min. Despite the measures to deepen the plane of anesthesia, the BP further shot up to 220/120 mmHg. Surgical team was informed and asked to stop manipulating the tumor. Injection nitroglycerin infusion was started at 0.5 μg/kg/min, and gradually the dose was increased to 1 μg/kg/min but nitroglycerin proved ineffective. Surgery resumed, and subsequently sodium nitroprusside (SNP) infusion at dosage of 0.5 μg/kg/min titrated to 1.0 μg/kg/min was started. BP remained a wide range (170/96 mmHg) despite increased anesthetic depth and antihypertensive theraphy. Therefore, we highly suspected an unrecognized ectopic pheochromocytoma and injection phentolamine 5 mg bolus was readied.

However, within 5 min of starting SNP infusion, the surgeon clamped the feeding vessel of the tumor. Once the feeding vessel was clamped, the BP gradually reduced to 120/70 mmHg and then suddenly plummeted to 80/40 mmHg. Nitroprusside and nitroglycerin infusions were stopped. A tumor mass measuring 10 cm × 8 cm was dissected out of the abdomen. An additional peripheral 14G IV line was secured and 500 ml hydroxyethyl starch and 1 Lt. Ringer lactate was rapidly infused. Injection phenylephrine 5 μg/kg bolus was given and 0.5 μ/kg/min infusion started. BP increased to 100/60 mmHg, and HR was maintained around 78/min. Intraoperative blood loss was found to be around 450 ml. Central venous line was secured in the right internal jugular vein under ultrasound guidance, and central venous pressure monitoring was done.

Rest of the intraoperative period was uneventful. After completion of surgery, the patient was extubated. The patient was conscious and oriented with stable hemodynamics without supports, maintaining an adequate oxygen saturation of 97%–98% with oxygen through venturi mask.

The patient was shifted to the Intensive Care Unit (ICU) for observation. Postoperative epidural infusion with injection ropivacaine was started in the ICU and titrated according to BP. His BP, pulse rate, and oxygen saturation remained within normal limits, and no episode of arrhythmia occurred. The patient was shifted to the ward after 48 h observation in ICU. Histopathology report confirmed the tumor as paraganglioma.

Discussion

Paragangliomas are indistinguishable from pheochromocytomas at the cellular level, and they may be collectively referred to as pheochromocytoma and paraganglioma (PPGL). Therefore, the anesthetic concerns of patients in PPGL are the same.

Classic symptom triad of these patients is headache, palpitations, and diaphoresis. Hypertension may be either a sustained or paroxysmal.

Predominant presentation in case of epinephrine-secreting and dopamine-secreting tumors is orthostatic hypotension, while sustained hypertension is typical of norepinephrine-secreting tumors.[3]

Arterial vasoconstriction and associated ischemia can cause chest pain and abdominal pain.

The historically quoted "Rule of 10s" (10% of PPGL are extra-adrenal, 10% are malignant, 10% are bilateral, and 10% are familial) is not correct because at least 32% of PPGL are familial.[4] In familial disease, more than 80% are bilateral or in multiple sites. The incidence of a malignant pheochromocytoma is found to be 10%–17%.[5]

Hemodynamic instability remains a common occurrence in patients with PPGL even though they are adequately prepared preoperatively with alpha blockers for 7 days and restoration of adequate intravascular volume.[6]

Hypotension following devascularization in any PPGL may be due to abrupt catecholamine deficiency, inadequate intravascular volume (due to pooling or absolute loss from surgery), or even catecholamine receptor downregulation.[7]

The initial treatment should include discontinuation of vasodilator therapy, optimization of intravascular fluid volume, and aggressive titration with norepinephrine and/or phenylephrine. If hypotension is refractory to the previous measures, vasopressin should be considered.

A transient hypoglycemic phenomenon can occur postoperatively due to sudden catecholamine deficiency and rebound hyperinsulinemia.

To conclude, for any patient with retroperitoneal tumor, while analyzing differential diagnosis, think of a clinically silent but hormonally active PPGL that can be the cause for sudden hemodynamic instability in the intraoperative period. This case emphasizes the need for vigilance and preparedness for such unexpected encounters which were crucial in managing the cases.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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