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Case Report

Undiagnosed Retroperitoneal Paraganglioma, an Anaesthesiologist's Challenge

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Abstract

Majority of the phaeochromocytomas are seen in the adrenal medulla, with a few being extra-adrenal. We present a case of a forty-seven year old hypertensive man who was taken up for a retroperitoneal tumour excision. On manipulation of the tumour there was severe rise in blood pressure and heart rate along with ST-T changes on the electrocardiogram, which suggested the possibility of a catecholamine producing tumour. Haemodynamics were controlled effectively with antihypertensive drugs, and the tumour was removed, after which the patient required inotropic support to control hypotension. Histopathology confirmed the presence of paraganglioma. A patient with a retroperitoneal tumour should be suspected of having a paraganglioma, even in an asymptomatic case. Management of such cases poses a challenge to the anaesthesiologist, even with the advent of new drugs.

Keywords: Anaesthesia, hypertensive crisis, paraganglioma, phaeochromocytoma, retroperitoneal.

1. Introduction

Phaeochromocytoma is a neuroectodermal tumour arising from the chromaffin cells of the sympathetic nervous system. 90% of cases, phaeochromocytoma is located in the adrenal medulla, the remaining 10% are extra-adrenal (paragangliomas) arising from the cells of the sympathetic ganglia, distributed along the paravertebral axis (Organ of Zuckerkandl), carotid body, prostrate and even the heart.[1,2] The unexpected encounter of phaeochromocytoma intraoperatively can lead to life threatening complications, and have been associated historically with a mortality rate of 40%.[1] We present an undiagnosed case who developed severe cardiovascular events on manipulation of the tumour.

2. Case report

A forty-seven year old, 80kg male patient, known hypertensive on tablet Telmisartan-Hydrochlorthiazide 40mg/12.5mg combination since four years, was admitted to the hospital for an elective retroperitoneal tumour excision, his chief complaint being pain in the abdomen since eight days. An abdominal computerised tomography (CT) scan showed a large retroperitoneal mass from the level of the uncinate process of the pancreas to the aortic bifurcation, measuring approximately 7.58 x 11.5 x 12.4cm. He had no other past medical or surgical history. Blood tests, electrocardiography (ECG), chest radiograph, two dimensional echocardiography (2D ECHO) were normal.

The starting heart rate (HR) on ECG was 61/min, non-invasive blood pressure (BP) was 154/92mmHg and oxygen saturation (SpO_2) was 100%. A 20G peripheral canula was *insitu* and 500mL of Ringer's Lactate (RL) was started. Intravenous (i.v.) induction was performed with fentanyl 1µg/kg, propofol 1.5mg/kg, atracurium 0.5mg/kg and intubation with number 8 cuffed endotracheal (ET) tube. Haemodynamics remained stable during induction and anaesthesia was maintained with 50% air in oxygen with sevoflurane 2vol%, and i.v. atracurium infusion at 25mg/h.

During tumour resection, the BP surged to 250/120mmHg with a rise in HR to 115/min. The plane of anaesthesia was deepened by administering fentanyl 1.5µg/kg i.v., propofol 50mg i.v., sevoflurance 3vol% and

nitrous oxide (N_2O) 50%. Suspecting an extra-adrenal phaeochromocytoma, we discussed with the surgeon and decided to continue since the tumour was well encapsulated and resectable, subject to haemodynamic control. Two 18 guage (G) peripherals, left radial 20G arterial line, right internal jugular vein central line (triple lumen – 7 French) were secured, and blood was sent for grouping and cross-matching. I.v. esmolol boluses amounting total to 100mg were given which brought down the heart rate to 90/min, followed by an infusion at 0.5mg/kg/h. I.v. nitroglycerine (NTG) infusion was started at 0.1µg/kg/min and was titrated till 0.4µg/kg/min which brought BP down to 180/100 mmHg. The ECG showed ST-T depression of -1.5, with fall in the SpO_2 to 89%. Chest auscultation confirmed no ET tube displacement and no adventitious sounds. Furosemide 20mg i.v. was administered. N_2O was stopped, fraction of inspired oxygen (FiO_2) was made 80%, and an arterial blood gas (ABG) was performed, which showed [Table 1].

Table 1: Arterial blood (Intra-operative)

pH	PaO_2	$PaCO_2$	$EtCO_2$	SaO_2	HCO^3	Hb	HCT	Na	K
7.18	73.5	52.6	31	89.7	19	15.7	48.2	133	3.3

Fifty mL sodium bicarbonate 7.5% and potassium chloride 20mEq/L were administered i.v. Phenoxybenzamine bolus of 2mg i.v. was administered, followed by an infusion at 0.5mg/kg/h. The SpO_2 rose to 95-96%, BP dropped to systolic of 150mmHg, HR dropped to 85/min and ST-T depression improved to -0.9. The blood glucose was 396mg/dL, 10 IU of Human Actrapid Insulin was administered i.v. and an infusion started at 4IU/h. The surgery was commenced and tumour removed.

On tumour removal, the BP dropped to 82/48mmHg. All infusions were stopped, sevoflurane was reduced to 1vol%, and crystalloids (RL, normal saline (NS) and plasmalyte) and colloid (volulyte) were rapidly infused. Noradrenaline infusion was started at 0.2µg/kg/min i.v. and titrated according to the BP.

On completion of the surgery, the patient had a HR of 110/min, BP of 101/55mmHg, and SpO2 of 99%. Noradrenaline infusion had been tapered to 0.1µg/kg/min. Two litres of RL, 1 litre of normal saline, 1.5 litres of plasmalyte and 1 litre of volulyte were in total given during surgery. The ABG showed [Table 2].

Table 2: Arterial blood gas (Post-operative)

pH	PaO_2	$PaCO_2$	$EtCO_2$	SaO_2	HCO^3	Hb	HCT	Na	K
7.24	97.3	43.4	27	97.3	18.2	11.4	35	133	4

The blood glucose was 298mg/dL. The blood loss was approximately 500mL and the urine output was 1200mL. The duration of the surgery was four hours. The patient was shifted intubated and sedated to the ICU. He was ventilated overnight and extubated the next day. Noradrenaline infusion was tapered down and stopped. He was shifted to the wards on the third postoperative day and discharged on the ninth postoperative day. Histopathology confirmed diagnosis of benign phaeochromocytoma.

3. Discussion

Phaeochromocytomas are rare tumours with a prevalence of 0.1-0.5% in the general population[1], being responsible for less than 0.1% of all cases of hypertension[3,4], and are more common in men.[2] Signs and symptoms are generally due to excess catecholamine secretion, most commonly noradrenaline, presenting with the classic triad of headache, tachycardia and sweating.[2,5] Other clinical features include sustained hypertension resistant to conventional treatment, pallor, anxiety attacks.[5] Few cases have atypical presentations, like normotension, psychiatric disorders, weight loss, polyuria, hyperglycemia, polycythemia, stroke and cardiomyopathy making the diagnosis difficult.[2,6] In such cases, the paraganglioma is discovered incidentally during CT or magnetic resonance imaging (MRI), or on the operating table. Clinical suspicion is the most important factor for identifying paragangliomas. Handling of the tumour results in "hypertensive crisis" which is defined as systolic BP more than 180mmHg and/or diastolic BP more than 120mmHg that requires immediate lowering of BP to prevent end organ damage.[1] This is believed to be because of sudden excessive release of catecholamines from the tumour.[2] Our patient's only past history was hypertension, without palpitations, sweating or any other symptoms related to paraganglioma, neither were there intubation nor incision responses; hence, a vasoactive neoplasm was not suspected till much after in the surgery.

Preoperative investigations to rule out any evidence of paraganglioma are crucial to carry out the surgery safely. Laboratory tests include twenty-four hour urinary metanephrine and vanillylmandelic acid levels.

Also, clonidine suppression test will rule out other causes for rise in catecholamines. Catecholamine secretion from a paraganglioma is independent of neurogenic control, hence will not be suppressed by clonidine. Plasma free metanephrine levels provide the best test for excluding or confirming paraganglioma.[3] Also, scintigraphic localization with ^{123}I- or ^{131}I-metaiodobenzylguanidine (MIBG) is highly specific for paraganglioma and is indicated even in asymptomatic cases.[7] Preoperatively diagnosed patients should be started on oral alpha-blockers (phenoxybenzamine) started at 40mg/day and gradually increased to 80-120mg/day, two weeks prior to surgery. Beta-blockers are started after alpha-blockade if tachycardia or other cardiac arrhythmias develop.[1,3] Surgery in "prepared" patients is associated with near 0% perioperative mortality.[1,2] In our patient, no additional investigations were performed as CT suggested a retroperitoneal mass.

Whenever there is a sudden hypertensive crisis during surgery in a case with suspicion of paraganglioma, it depends on the individual case whether to go ahead with the surgery and remove the tumour, or terminate it to operate again after stabilization of the patient. [2,7] We decided to continue as we were able to control the haemodynamics with antihypertensives and the tumour was easily resectable. Hypertension and tachycardia in our patient was controlled by i.v. NTG, esmolol and by deepening the plane of anaesthesia. The patient developed ST-T depression and fall in SpO_2. Beta-blockers should never be started before alpha-blockers, because in the absence of beta-2-mediated vasodilation, profound unopposed alpha-mediated vasoconstriction can cause hypertensive crisis or pulmonary oedema.[3] Hence, on giving phenoxybenzamine and furosemide i.v., there was improvement in ST-T depression and SpO_2. In the study by Hariskov and Schumann, sixty-two case reports were retrospectively studied for intraoperative management of patients with incidental catecholamine producing tumours, all the patients who died had received only one antihypertensive drug, and none of them was treated with an alpha-blocker intraoperatively.[1] Hypotension in our case followed immediately after ligation of the tumour vessels, and required i.v. noradrenaline. Patients with paraganglioma are chronically vasoconstricted because of high levels of circulating catecholamines, and have secondary hypovolemia [1,3], hence the haemoglobin/haematocrit of our patient was high initially on the ABG, which reduced after giving fluid. Noradrenaline or adrenaline should be given only when fluid boluses are ineffective; however an increased circulating blood volume may not prevent hypotension.[4]

Although the majority of paragangliomas are benign, approximately 10%-15% are malignant.[2,7] There are no standard histologic criteria for diagnosing malignancy, hence malignancy is suspected only when the tumour is present at sites where paraganglion cells are not normally found.[7] Phaeochromocytomas/paragangliomas are associated with genetic syndromes such as neurofibromatosis 1, von Hippel-Lindau syndrome, multiple endocrine neoplasia type 2.[2,8] Thus preoperative genetic counselling can help in diagnosing phaeochromocytomas/paragangliomas.

4. Conclusion

Paragangliomas are very difficult to diagnose preoperatively, because patients may have no symptoms or atypical symptoms. Thus, our case report emphasizes that any patient with a retroperitoneal tumour should be suspected and appropriately investigated for a paraganglioma, and if diagnosed, optimized before surgery to prevent life-threatening perioperative events.

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