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Case Report

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Anesthetic Management of a Large Retroperitoneal Paraganglioma: A Rare Case Report

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ABSTRACT

Paragangliomas are rare tumors derived from the neural crest cells. Most of the paragangliomas occur as sporadic tumors. The commonest incidence occurs in the second and third decade of life with a slight male preponderance. Clinically patients with a retroperitoneal paraganglioma often present with back pain or a palpable mass. There is a 5% incidence of turning into malignancy and these tumors are associated with a high risk of morbidity and mortality from cardiovascular complications. Management for paragangliomas typically involves complete surgical excision. A multi-disciplinary approach is suggested for a better outcome of the procedure.

Keywords:

Retroperitoneal paraganglioma, Anesthesia concerns, Sporadic tumor, Neural crest.

Introduction

Phaeochromocytoma is an endocrine tumor arising from catecholamine producing chromaffin cells in the adrenal medulla by 2004 WHO classification. Closely related tumors found in the extra-adrenal sympathetic and parasympathetic paraganglia were classified as extra-adrenal paragangliomas [1]. Paraganglioma are rare tumors derived from the neural crest cells. Extra adrenal paragangliomas may develop anywhere from chromaffin tissue along the autonomic nervous system usually found with many of the major blood vessels over the head, neck, thorax, abdomen, and pelvis [2]. In this context, we present the anesthetic management of such rare tumors.

Case Report

29 year aged male, Engineer by profession working in a private concern was referred to a general surgical department in our institute with a history of abdominal fullness and discomfort for the past 2 years on and off. No significant past medical or surgical history of any systemic illness and neither a smoker nor an alcoholic. On examination, he was fairly nourished and found to be anemic. His weight was 52 kg, height was 160 cm with a BMI of 20.3 Vitals findings showed a heart rate of 110 beats per minute and blood pressure was 124 over 90 mm/Hg. Airway and spine examination was normal. Abdominal examination revealed a firm mass of size 15 × 20 cm occupying the whole of the abdomen (Figure 1). No tenderness was present over the abdomen. Another systemic examination was normal.

Routine blood investigations revealed anemia with hemoglobin of 7.1 g/dl with all other blood parameters within normal limits. Urine metanephrines was 166.2 µg/24 hours. Chest X-ray, electrocardiography, Renal, and liver functions were within normal limits. Ultrasound abdomen revealed a large

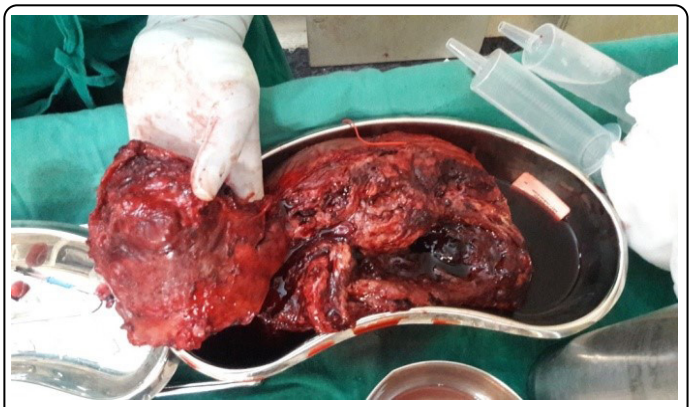

Figure 1: Specimen of paraganglioma.

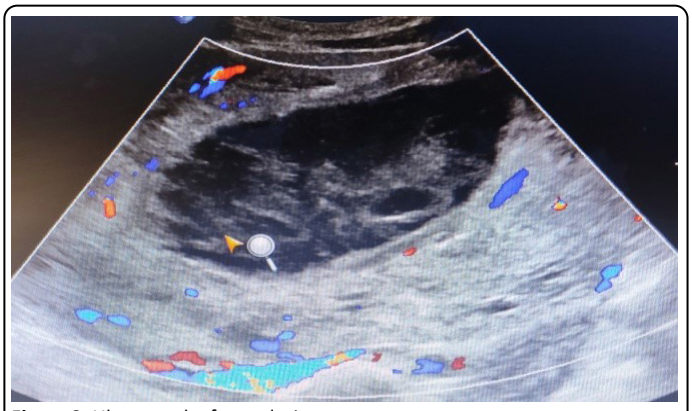

Figure 2: Ultrasound of mass lesion.

extensively vascular heterogeneous mass lesion in the centre of the abdomen (Figure 2). Computed tomography with contrast, revealed a large well defined solid mass involving mesentery and retroperitoneum occupying the whole of the abdomen, displacing right iliac vessels posteriorly and laterally with no evidence of encasements along with right ureter displacement. Anteriorly mass extended till abdominal wall without any infiltration and posteriorly it was compressing both psoas

muscle. Bowel loops were displaced laterally and all features were suggestive towards the diagnosis of the gastrointestinal stromal tumor.

Echocardiography showed features of sinus tachycardia with left ventricular hypertrophy with an ejection fraction of 39%. Cardiac magnetic resonance imaging with contrast showed moderate systolic dysfunction due to left ventricular moderate hypokinesia, thickening of the interventricular septum, left ventricular myocardial risk index was in upper normal limits with diffuse thin subendocardial enhancement in the left ventricle suggesting amyloidosis.

Pre-operatively blood transfusion was done to improve hemoglobin to 10 g/dl along with haematinics. USG guided trucut biopsy was taken from the mass under conscious sedation. There was no hemorrhage or any hemodynamic instability following the biopsy and the histopathology report was suggestive of paraganglioma/phaeochromocytoma. Endocrinology opinion was sought and advised for urine metanephrines, serum prolactin, and ANA profile. All of these test reports were within normal limits. The cardiologist diagnosed as catecholamine-induced cardiomyopathy provisionally and started on alpha (Tablet Prazosin 2.5 mg OD) and beta-blockers (Tablet Inderal 10 mg BD). Specialist opinion including pre-anesthetic evaluation and fitness for surgery was obtained from all concerned specialties before planning for wide excision of the mass with prophylactic bilateral ureteric stenting. He was categorized as American Society of Anesthesiologist ASA Grade II and proceeded to surgery with adequate reservation of blood and blood products, starvation of solids 6 hours and clear fluids up to 2 hours before surgery and obtained consent for epidural anesthesia, invasive lines, general anesthesia including post-operative ventilation considering the major surgery. Normal saline was started at a rate of 50ml/Hr as maintenance from midnight.

The plan of anesthesia executed was combined epidural and general anesthesia with controlled ventilation and invasive lines. Emergency cardiac drugs were kept ready. Datex ohmeda anesthesia Boyle's machine was used; ASA standard monitoring was done and baseline readings noted. Under local anesthetic infiltration, Epidural anesthesia was instituted at the level of T8-T9 with 18G Tuohy needle and invasive lines were secured under USG guidance in the right internal jugular vein for CVP monitoring and fluid administration. Right radial artery cannulated for continuous arterial pressure monitoring and blood sample analysis when needed.

The patient was premedicated with Injection Glycopyrrolate 0.1 mg and Injection Midazolam 2 mg intravenously (IV) in the pre-operative room half an hour before shifting the patient inside the OT. Analgesia was supplemented with Injection Fentanyl 100 µg IV and stress response was attenuated using preservative-free 2% lignocaine 40 mg IV 90 seconds before intubation. Anesthesia was induced with Injection Etomidate 10mg and the patient was intubated using 4 MAC blade 8.5 mm portex cuffed endotracheal tube with Injection Vecuronium 5mg IV. Laryngoscopic Grade 1 was noted and the endotracheal tube was fixed at 22 cm at the angle of the mouth, bilateral air entry checked and confirmed by waveform capnography. Anaesthesia maintained with controlled ventilation using N_2O:O_2 (50:50) and Sevoflurane achieving a MAC of 1 to 1.3. Epidural anesthesia initiated with 0.2% Ropivacaine with Fentanyl 2µg/ml at a titrated dose. BIS monitoring is used to assess the depth of anesthesia.

Bilateral ureteric stenting was done before laparotomy. The abdomen was opened in layers and a solid encapsulated tumor of size 20 × 15 cm noted, occupying the mesentry of small bowel and retroperitoneum displacing the large bowel and small bowel to the left. No metastasis or ascites noted. On mobilization of the tumor, it was found to be adherent to right common iliac artery and internal iliac artery. The vascular surgeon was sought for dissection and the tumor was excised partially leaving behind the adhered part and primary repair of the common iliac artery was done with prolene sutures. Nitroglycerin infusion was titrated to blood pressure response at the rate of 0.5 to 1.5 µg/kg/min. Retroperitoneum closed in layer and good hemostasis was attained before abdominal closure.

During the surgery, the patient had hypotension during tumor dissection and following excision, requiring Dopamine about 8 to 10 µg/kg/min and Noradrenaline support up to 0.08 µg/kg/min and a single dose of Hydrocortisone also supplemented. Intraoperative blood loss was around 1500 ml and required 2 packed cells, 2 fresh frozen plasma transfusions. 10% of Calcium gluconate 10cc was given. No evidence of hypoxia, acidosis or hypothermia was noted intraoperatively. Arterial blood gases were satisfactory after elective ventilation for two hours in Surgical ICU. The patient had a good clinical recovery, normothermic, weaned off vasopressors, no acidosis or alkalosis and was extubated in the surgical ICU on the same day of surgery (Figure 3).

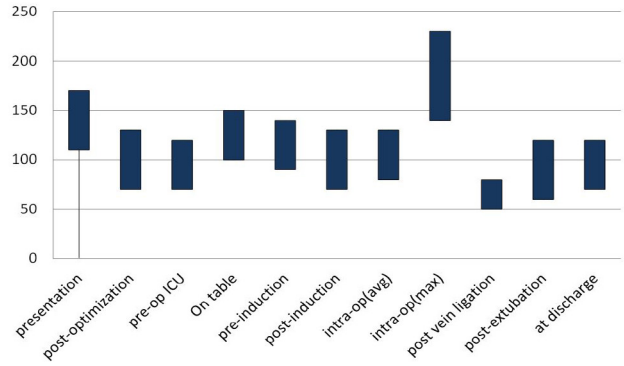

Figure 3: Systolic blood pressure variations.

Monitored in High dependency unit for 24 hours with epidural infusion for pain relief and was transferred to the surgical ward on 1st postoperative day with epidural 0.2% Ropivacaine and Fentanyl 2 µg/ml at a rate of 7 ml/Hr. He was started on oral diet gradually, mobilized and the epidural catheter was removed on 3rd POD. Post-operative specimen confirmed the diagnosis exhibiting strong diffuse cytoplasmic staining for chromographin and less than 2% express nuclear positivity for ki67. The patient was discharged on the 5th post-operative day.

Discussion

Catecholamine producting tumors of extra-adrenal origin are known as paragangliomas. They arise in any portion of the

paraganglionic system though they are most commonly seen below the diaphragm in the organ of Zuckerkandl.

They represent atleast 15% of adults and 30% of childhood phaechromocytomas [3]. The incidence most commonly occurs in the second and third decade of life with a slight male preponderance. This is in contrast to adrenal phaechromocytoma which typically are diagnosed in the fourth and fifth decades with a slight propensity for women [3]. Episodic tachycardia, sweating, headache, and signs of paraoxysmal hypertension are classic of phaeochromocytoma or paraganglioma [4,5]. Clinically patients with a retroperitoneal paraganglioma often present with back pain or a palpable mass [6].

There are two types of paragangliomas as sympathetic paraganglioma (arising from the sympathetic paraganglia located along the paravertebral and paraaortic axis) and parasympathetic paraganglioma (arising from the paraganglia located in the head and neck proximal to the vascular structures that gave rise to the carotid body, aortic pulmonary septum, intravaginal and jugulo tympanic tumors, as well as those arising from the wall of some organs such as the urinary bladder). Nearly 5 to 10% occurring in the extra-adrenal sites are from the upper cervical region to the pelvis related to the autonomic nervous system [7].

85 to 90% of the paragangliomas are found within the abdomen. There is a 5% incidence of turning into malignancy and these tumors are associated with a high risk of morbidity and mortality from cardiovascular complications [8]. When presenting within the abdominal cavity, they may arise as a primary retroperitoneal neoplasm.

Visceral paragangliomas is an uncommon tumor but there is a presentation in the urinary bladder, liver, gall bladder, larynx, and interatrial septum of the heart. The clinical presentation depends on the secretion of catecholamine, malignancy or mass effect; paragangliomas may also be discovered incidentally and are termed as “Incidentalomas” [9]. They synthesize, store and secrete catecholamines because of which they may present with headache, sweating, palpitations, and symptoms of functional hypertension [10]. On the other hand, they may remain silent and nonfunctional presenting with vague aching pain abdomen due to the episodic release of catecholamines.

Most of the paragangliomas occur as sporadic tumors. However certain hereditary syndromes such as Von Hippel-Lindau syndrome, multiple endocrine neoplasia type 2, neurofibromatosis type 1, and familial paraganglioma syndrome have been associated with the development of paragangliomas [11].

In the presence of histological diagnosis and symptoms of catecholamine excess (nonfunctional), these may be mistaken for Gastrointestinal Stromal Tumors (GIST). Approximately 10-15% of such tumors are nonfunctional. They are often locally invasive and associated with a high incidence of local recurrence. Nonfunctional paragangliomas pose a significant diagnostic challenge [2]. In abdominal computed tomography there are no unique imaging characteristics specific for paragangliomas. Consequently, these tumors may be mistaken for other primary epithelial or mesenchymal abdominal tumors [12].

Management for paragangliomas typically involves complete surgical excision. Surgical debulking is considered as palliative therapy for malignant paragangliomas. However complete excision might be difficult due to adherent to major blood vessels. The anesthetic management of the surgical excision of the tumor requires the knowledge of physiological and pathological effects of catecholamine secreting tumors under the influence of anesthesia and surgery as they have a significant effect on the outcome of the surgery [13]. Excessive catecholamine production and secretion in the circulation poses serious life-threatening cardiovascular complications like hypertension, myocardial infarction, pulmonary edema, arrhythmias, and cardiomyopathy [14]. The major anesthetic concerns include adequate preoperative optimization of the blood pressure, safe conduct of anesthesia, smooth intubation and extubation attenuating the stress response, maintaining hemodynamic stability, supplementation of steroids, monitoring glycaemic status and providing balanced multimodal analgesia [15].

Conclusion

A retroperitoneal paraganglioma is a rare type of tumor that often is misdiagnosed as gastrointestinal tumor. Hence adequate pre-operative workup and specialist opinion is needed before planning for an elective surgery. A multi-disciplinary approach is suggested for a better outcome of the procedure. Paragangliomas are usually nonfunctional.

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