Case Report

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Extra Adrenal Paraganglioma: A Case Report with Review of Literature

Yalagachin Gurushantappa[1], Mashal Sanjay[2], Huchchannavar Suresh[3], B Jyothi[4], N Nisha[5]

[1]Professor and Head [2]Senior Resident [3]Assistant Professor [5]Junior Resident, Department of Surgery, [4]Associate Professor, Department of Anaesthesia, Karnataka Institute of Medical Sciences, Hubli, Karnataka 580022 India.

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Abstract

Paragangliomas (extra-adrenal pheochromocytomas) are rare tumours that arise from extra adrenal chromaffin cells. Paragangliomas originate from para-ganglia at number of anatomical sites, including the head, neck, thorax and abdomen. Paragangliomas are characterized by secretions of excessive catecholamines. However, between 40 and 50% of paragangliomas are non-functional and/or potentially functional. Functional and potentially functional (difficult to diagnose preoperatively) paragangliomas during intra-operative handling of the tumour may cause a sudden release of catecholamines, leading to disastrous consequences [1]. We present a case of a middle aged male presenting with abdominal mass and pain was found to be a potentially functional paraganglioma intraoperatively. With this report we aim to raise awareness that a paraganglioma should be considered in differential diagnosis for intra-abdominal tumours.

Keywords: Hypertensive crisis; Pheochromocytoma; Paraganglioma.

Introduction

Pheochromocytoma /paraganglioma (PCC/ PGL) as rare neuroendocrine tumours that arise from sympathetic and parasympathetic paraganglia. Paraganglioma comprises an extra-adrenal subset of PCC and is often characterized by secretion of catecholamines, although sometimes are biochemically inactive. This makes diagnosis challenging [2]. About 25% are hereditary or have a family relation and are associated to the multiple endocrine neoplasia type 2 (MEN 2), Hippel-Lindau syndrome, neurofibromatosis, Carney triad or mutations in the oncogenes suppressors of the Succinate dehydrogenase (SDHB, SDHC, SDHD) [3]. Clinical presentation includes symptoms related to catecholamine hypersecretion and/or tumour mass effect. Surgical resection with appropriate perioperative management of catecholamine-related symptoms remains the treatment of choice [2].

Case Report

A 46-year-old male presented with history of pain abdomen of one and a half months duration, occasional palpitations and sweating and was recently diagnosed to have diabetes mellitus. However, patient denied history of any headache. On examination pulse rate was 84 per minute, blood pressure was 130/80 mm of Hg, with a vague mass of 6 x 5 cm in the left hypochondrium. CT abdomen and pelvis suggested a well defined heterogeneously enhancing soft tissue density mass lesion measuring 10 x 8 x 7 cm in the left upper quadrant, a retroperitoneal tumour possibly GIST (Fig. 1).

On exploration, a retroperitoneal tumour of size 15 x 10 cm found to the left of lumbar vertebra below the splenic flexure between descending colon and root of the mesentery near the left renal hilum (Fig. 2). Left suprarenal gland appeared

Corresponding Author: Mashal Sanjay, Senior Resident, Department of Surgery, Karnataka Institute of Medical Sciences, Hubli, Karnataka 580022, India.

E-mail: dr.sanjaymashal@gmail.com

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normal. Patient pre-op blood pressure was 160/100 mm of Hg, Increased to 260/150 mm of Hg on handling the tumour. Hypertensive crisis was managed effectively by anaesthesiologist with nitro-glycerine, Metoprolol, and Propofol. However, patient was hypotensive immediately following resection of the tumour for which he was initiated on adrenergic support. Patient was continued on adrenergic support for two days following surgery. Patient was asymptomatic and normotensive and his blood sugars were in the normal range following surgery without any medications. Subsequent post-op recovery was uneventful. Patient discharged on postoperative day seven with advice to follow-up regularly.

Specimen consisted of a single globular mass measuring 12 cm in diameter (Fig. 3). External surface is congested, nodular, well encapsulated, soft in consistency with cystic areas. Cut surface shows multilobulated tan yellow areas with areas of haemorrhage seen. Multiple sections studied from mass shows tumour cells arranged in solid and trabecular pattern. Cells surrounded by capillaries showing Zellballen appearance. These cells had rounded nucleus with coarse clumped chromatin with granular amphophilic cytoplasm (Fig. 4). The features are suggestive of Paraganglioma.

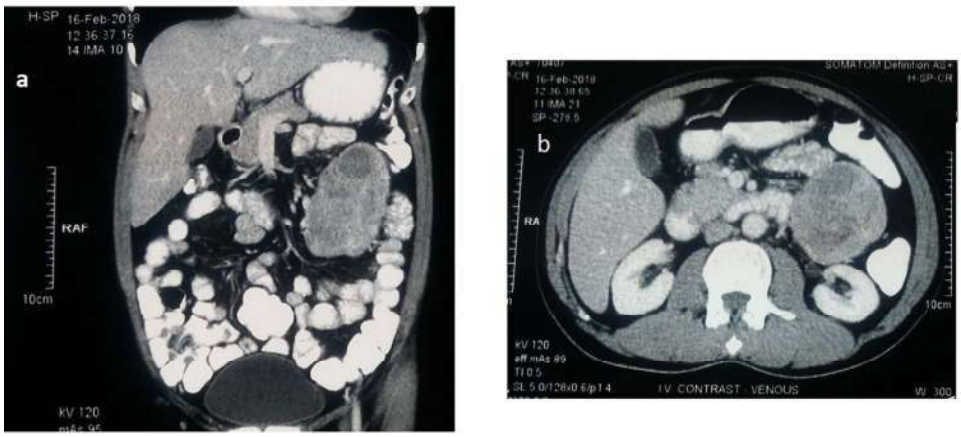

Fig. 1: Image showing a well-defined heterogeneously enhancing soft tissue density mass lesion measuring 10 x 8 x 7 cm in the left upper quadrant. (a- coronal view, b- sagittal view)

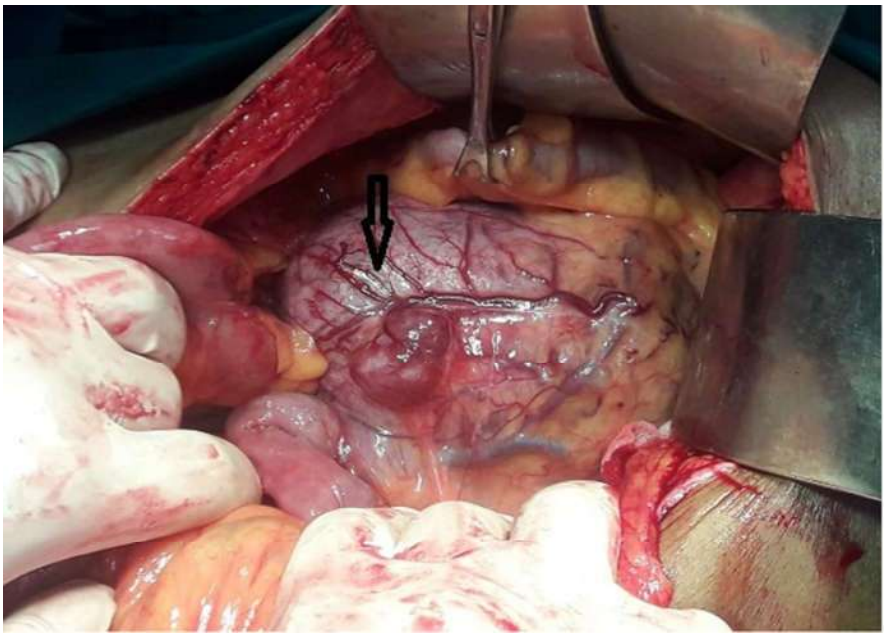

Fig. 2: Intraoperative image of paraganglioma found to the left of lumber vertebra below the splenic flexure between descending colon and root of the mesentery near the left renal hilum.

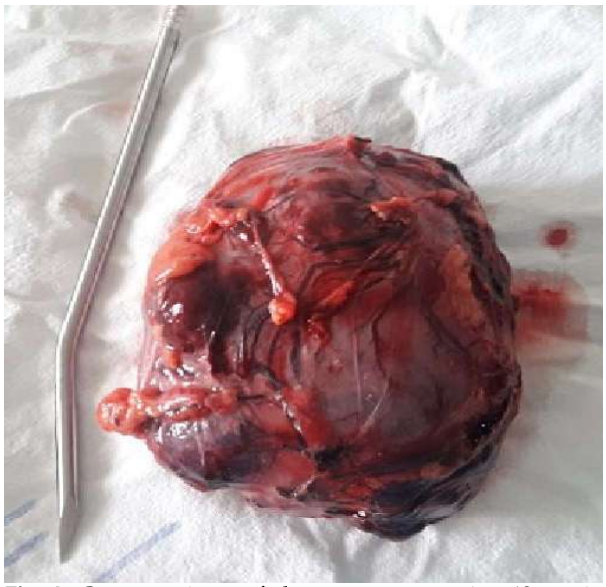

Fig. 3: Gross specimen of the tumour measuring 12 cm in diameter

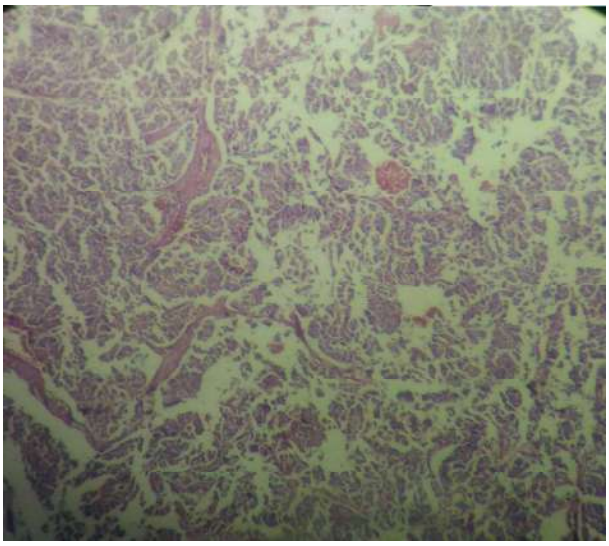

Fig. 4: Characteristic growth pattern of a pheochromocytoma with tumour cells arranged in solid and trabecular pattern surrounded by capillaries showing Zellballen appearance

Discussion

Pheochromocytomas are tumours arising from chromaffin cells of the adrenal medulla. They are called paraganglioma if chromaffin-cell tumours originate from extra-adrenal sites along the sympathetic and/or the parasympathetic chain [4]. Extra adrenal paraganglioma can occur in four types of locations—branchiomeric, intravagal, aorticosympathetic and visceral autonomic. The branchiomeric and intravagal tumours are found in head and neck region and are rarely functional. The aorticosympathetic tumours are found along the length of aorta, between the renal arteries, around the iliac bifurcation and include the organ of Zuckerkandl. The visceroautonomic paraganglioma occurs in association with blood vessels or visceral organs like the bladder. The aorticosympathetic and visceroautonomic tumours are mostly functional. Extra-adrenal sympathetic paraganglioma most commonly arise from chromaffin tissue around the inferior mesenteric artery and aortic bifurcation and less commonly from chromaffin tissue at other sites, whereas the extra-adrenal parasympathetic paraganglioma are most commonly found in the head and neck region [6]. Clinical incidences of pheochromocytomas have been estimated to range from 0.4 to 9.5 per million per year, and approximately 1.5 per million per year in terms of paragangliomas [5]. Long-term follow-up has shown that PCCs/PGLs exhibit a 15–20% 10-year probability of recurrence and up to 20% malignancy rate [8].

Patients may present with symptoms secondary to secretion of excess norepinephrine and epinephrine. Up to 25% of cases will be entirely asymptomatic. The classic triad of palpitations, headache, and diaphoresis is present in less than a quarter of patients. A majority of patients will have hypertension, but it can be intermittent in 50% [7].

The best screening tests for initial assessment is measurement of free plasma and urinary fractionated metanephrines. Imaging modalities used for the detection of primary or metastatic PCCs/PGLs include computed tomography (CT) and magnetic resonance imaging (MRI) [8]. Functional imaging techniques, including 123 I-meta-iodobenzylguanidine (MIBG) scan and somatostatin receptor scintigraphy, in combination with CT or MRI scans may be used to improve the sensitivity and specificity of diagnosis [1].

Surgery is the treatment of choice for paraganglioma. Before surgery, appropriate medical preparation with α-blocking and β-blocking agents is very crucial to avoid intraoperative hypertensive crisis. Use of β-blockers prior to α-blocker can lead to unopposed α-adrenergic vasoconstriction that predisposes to hypertensive crises. Preoperative volume expansion with saline infusion is often used to prevent postoperative hypotension secondary to chronic volume contraction [6]. The laparoscopic excision should be considered for small PGL tumours with a favourable surgical location [3]. In the case of malignant paragangliomas postoperative radionuclide treatment should be initiated to eradicate the residual tumours or micro

metastases [4]. In patients with metastatic disease, palliative chemotherapy with cyclophosphamide, dacarbazine and vincristine is recommended [6]. Approximately one third of patients have persistent or recurrent paragangliomas, and long-term follow-up is important [9].

Conclusion

The biggest problem of PGL is to suspect it in the first place. Functional PGL are evaluated preoperatively by biochemical and imaging modalities. The principle challenge arises in making the diagnosis of non-functional and potentially functional PGLs. Clinically and biochemically silent paragangliomas may cause hypertensive crisis and result in serious consequences when they are manipulated intraoperatively and certainly this case represents a good example. When there is a clinical suspicion the diagnosis could be made by the determination of high catecholamines and metanephrines in plasma and urine and locating the tumour through imaging studies. Alert anaesthesia team to anticipate and manage hypertensive crisis and post-operative care is paramount in good outcome.

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