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Case Report

Case report of extra-adrenal pheochromocytoma

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Abstract

Pheochromocytoma is a neuroendocrine tumour that usually develops ahead of the chromaffin cells of the adrenal medulla. Embryonic rests of chromaffin tissue arising from sympathetic plexus within bladder wall are believed to be the source of these tumors. Here we reported a case report of 14 year old male patient having possibilities of Pheochromocytoma.

Keywords: pheochromocytoma, neuroendocrine tumour, chromaffin cells.

1. Introduction

The first reported case of pheochromocytoma is attributed to Frankel in 1886[1]. Pheochromocytoma is a neuroendocrine tumour that usually develops ahead of the chromaffin cells of the adrenal medulla. Embryonic rests of chromaffin tissue arising from sympathetic plexus within bladder wall are believed to be the source of these tumors. Extra-adrenal pheochromocytomas usually are located within the abdomen in association with the celiac, superior mesenteric, inferior mesenteric ganglia and Organ of Zuckerkandl (chromaffin body derived from neural crest located at the bifurcation of the aorta or at the origin of the inferior mesenteric artery)[2].

2. Case Report

A 14 year old male patient weighing 36kg presented with complaints of breathlessness and palpitations to medicine OPD. He was admitted for further evaluation. Patient's Blood pressure was 160/110 and Pulse was 114bpm. The patient had similar on and off complaints of breathlessness in past which aggravated on moderate work. Cardiovascular examination was normal. Abdominal examination did not reveal any mass or organomegaly. On routine U.S.G, a mass of 4x6cms was found anterior to inferior vena cava and aorta and was radiologically diagnosed to be paraganglioma arising from organ of Zuckerkandl. The case was referred to Department of Surgery for further evaluation.

C.T scan was done which revealed 32x46x62mm sized soft tissue mass lesion anterior to inferior vena cava and aorta at L3-L4 vertebral level. The lesion was supplied by a branch arising from aorta and right common iliac artery. Metanephrine levels were also analysed and were found to be raised (702.72ug/24 hrs). Surgery was planned and risks were explained to the patient. Preoperative preparation of the patient and control of hypertension was done in consultation with anaesthetist with nifedipine, aten and then with doxacard. Patient was taken up for surgery. Through midline incision abdomen was opened, kocherisation of the duodenum done. Caecum was mobilized. A 4×3 cm tumor was found over IVC and aorta at L3-L4 level. Tumour was excised with slow and steady dissection after ligating all feeding vessels. Blood pressure was maintained within normal limits throughout the procedure. Post-operative period was uneventful with blood pressure maintained within normal limits. Histopathological examination confirmed the diagnosis.

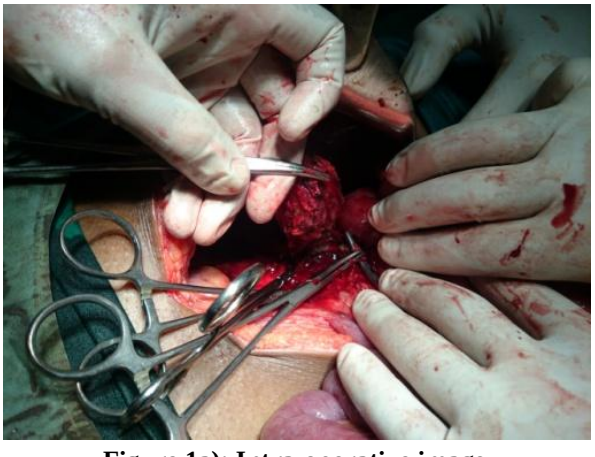

Figure 1a): Intra-operative image

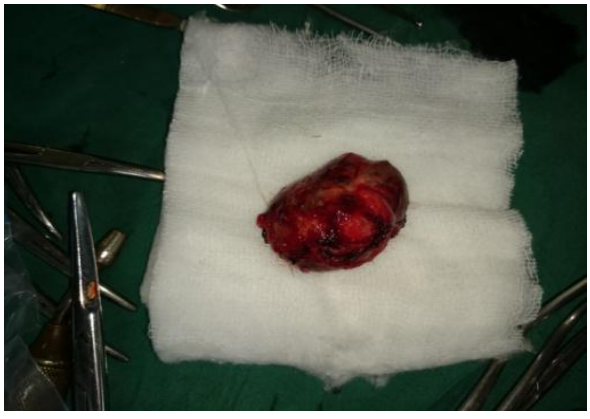

Figure 1 b): Post-operative specimen

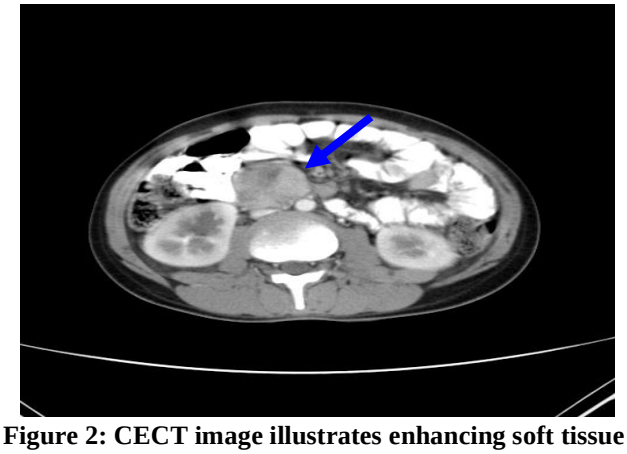

Figure 2: CECT image illustrates enhancing soft tissue mass lesion anterior to right kidney

3. Discussion

The diagnosis of pheochromocytomas, as well as EAPs, involves demonstrating elevated catecholamines and their metabolites in the blood and urine. Lenders *et al* reported on 1003 patients, 214 of which were later confirmed to have pheochromocytomas[1]. With the new advances in imaging technology, USG, Computed Tomography (CT), Magnetic Resonance Imaging (MRI), may be useful in localizing the tumor.

While $I^{(131)}$-methylliodobenzylguanidine and PET-CT helps to evaluate its function[3]. The traditional teaching of the "10% rule," which noted that 10% of all pheochromocytomas are at extra-adrenal sites, may actually be an underestimation. A review of the literature suggests that EAPs actually constitute 15% of adult and 30% of pediatric pheochromocytomas[4].

Relatively few studies have addressed the issue of appropriate evaluation and surgical treatment of adrenal masses in patients with a history of malignancy. Reports of adrenal incidentaloma often specifically exclude patients with synchronous or metachronous extra-adrenal malignancies, whereas most reports of adrenalectomy for metastatic cancer exclude those with primary tumors[5]. EAPs recur and metastasize more often than their adrenal counterparts, making lifelong follow-up essential [4].

References

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