A benign extra adrenal pheochromocytoma

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Introduction

Pheochromocytomas are functional catecholamine secreting tumours of chromaffin cells found in the adrenal medulla. It is a rare tumour [1,2]. Traditionally it was considered that 10% of Pheochromocytomas were extra-adrenal, but recent literature reviews show that extra-adrenal pheochrocytomas constitute 15% of adult and 30% of paediatric pheochromocytoma. Extra adrenal Pheochromocytomas (EAP) are observed in the second and third decade of life with slight male predominance. They are often multi-centric and more likely to be malignant [1,2].

The most common location of EAP is the superior para-aortic region between the diaphragm and lower pole of the kidney [2]. Less commonly reported sites are bladder, thorax, neck and pelvis [1]. Here we report a case of EAP which was successfully treated with surgical intervention.

Case presentation

A 47-year old house wife presented with headaches and dizziness which were paroxysmal for last one year duration. Her medical history was notable for hypertension which was kept under control with medications. She was on Prazocin, Amlodipine and Losartan. On examination, the patient had no abnormal physical findings. Complete blood count, blood sugar, blood urea, chest x ray and ECG reports were within normal limit. 24 hour urinary Vanillylmandelic acid level was done twice which found within normal range. Computed Tomography (CT) scan revealed a solid lesion measuring 3.3 x 2.6 x 2.4 cm, located in the left para-aortic region at first and second lumbar vertebrae level. The lesion was demonstrated between the left main renal artery and renal vein. It was closely related to the renal vessels but had no vascular invasion or encasement (Figure 1). In the pre-operative preparation, Propranolol was added to the drug list and the patient was well hydrated.

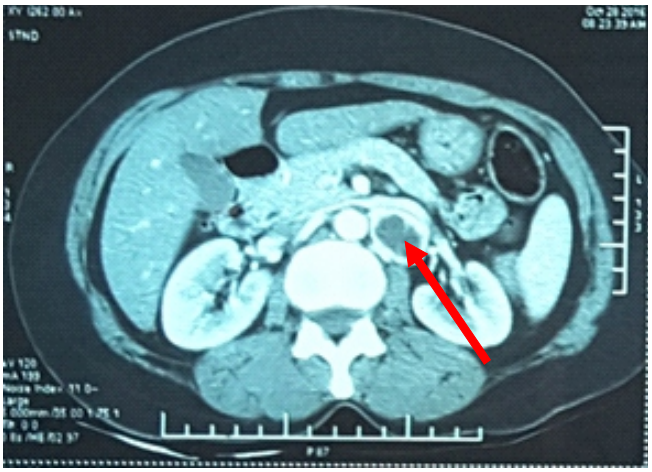

Figure 1: CT scan of abdomen. Red arrow indicates the tumour

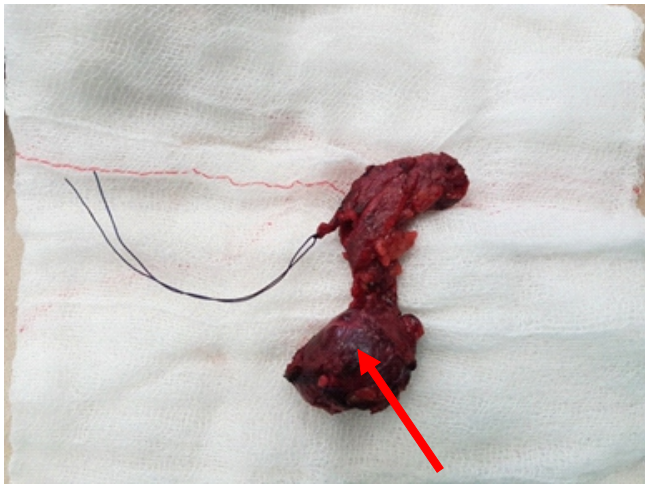

Figure 2: Arrow indicates the tumour

The tumour was accessed through a left subcostal incision. It was approached retroperitoneally and the adhesions to the left renal artery and vein were released. Continuation with left adrenal gland was noted. Therefore tumour was excised with left adrenal gland after ligating all feeding vessels (Figure 2). Blood pressure was maintained within normal limit throughout the procedure. Post operative recovery was uneventful. Histopathology confirmed the benign nature of the EAP with an unremarkable adrenal gland. The adrenal gland was separated from EAP by fibro fatty tissue. At the 1st postoperative visit in 1 month, the patient was normotensive

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and free of episodic symptoms of headache and dizziness.

Discussion and conclusion

Patients with pheochromocytoma or EAP presents with headache, excessive and inappropriate sweating and palpitation. Other symptoms include weakness, fatigue, nausea, abdominal pain and dizziness [1,2]. The most common sign is hypertension. Clinical characteristic of hypertension may vary and may show a sustained or paroxysmal pattern [4]. The clinical manifestations are due to uncontrolled secretion of catecholamines, mainly norepinephrine. Excessive secretion may leads to life threatening hypertension and cardiac arrhythmias. Therefore missing the diagnosis almost invariably results in cardiovascular complications and death [4].

EAP can also be non functional. They may develop symptoms and signs related to the organs compressed by the tumour, e.g. unilateral ureteric obstruction, small bowel obstruction and ileofemoral deep venous thrombosis [1].

Biochemical evaluation is the first step in diagnosis of pheochromocytoma and EAP by demonstrating elevated level of catecholamines and their metabolites in the blood and urine. The sensitivity of Vanillylmandelic acid (VMA) is 64% and specificity is 95%. Normal values were found in 25% of VMA assay [5]. Plasma free metanephrine is the test of choice for confirming the diagnosis. It has sensitivity of 99% and specificity of 89% [1]. However this test is not currently available at our institution.

Imaging is important for evaluating the location, extent and multifocality of disease [1]. The most widely used imaging technique is the CT scan [1, 5]. Sensitivity of CT is 98.9% for intra-adrenal pheochromocytoma and 90.9% for extra-adrenal pheochromocytoma [5]. MRI is recommended for patient with biochemically proven disease when CT is negative. In addition it can be used in pregnancy, children and patient with renal insufficiency or contrast allergy [1]. If preliminary imaging failed to identify the tumours, an 131I labelled Metaiodobenzylguanidine (I-MIBG) study should be ordered. It is extremely useful in EAP and a negative result excludes a pheochromocytoma. It also helps to evaluate multifocality and metastatic disease [1, 3]

Symptomatic treatment should be started with the aim of controlling blood pressure and heart rate before definitive treatment. Initially blood pressure can be controlled with alpha blockers or vasodilators with additional beta blockers to control heart rate. The most effective alpha blocker is Phenoxybenzamine, an irreversible long acting alpha adrenoceptor blockers that oppose vasoconstriction caused by catecholamines.

Surgical removal of the tumour is the definitive management. Preoperatively patient must be properly prepared with combined alpha and beta blockers to combat the effect of noradrenaline. Even with the best preoperative preparation, wide changes in blood pressure, irregular rhythm and bleeding can occurs. A hypertensive crisis occurs during handling of the tumour and ligation of adrenal vein may precipitate a hypotensive crisis. Therefore the patient needs invasive arterial and cardiac monitoring. Intravenous infusion of alpha blockers and beta blockers are given during surgery according to intra operative blood pressure and pulse rate. A sudden fall in blood pressure is managed with an infusion of large volume of fluid and intravenous epinephrine [1, 3]. Post operative normalization of blood pressure was observed in 60% and an improvement in 26% of patients. However blood pressure did not change significantly in 14% of patients [5].

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Learning Points:

- This case alerts us that when patient presented with typical symptoms and signs pheochromocytoma must be excluded.
- VMA can be normal in small proportion of patient but imaging will reveal the tumour.