

**CASE REPORT**

Extra Adrenal Paraganglinoma : A Case Report

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Abstract

Extra adrenal pheochromocytomas (EAPs) commonly arise in the organ of Zuckerkandl. Their presentation symptoms are similar to pheochromocytomas. Biochemical investigation demonstrating elevated blood and urine catecholamines and their metabolites along with imaging studies such as ultrasonography (USG), CT, MRI and ^{131}I labeled MIBG (meta-iodobenzylguanidine) scan help in achieving the diagnosis. We present a 35 yr old adult female, who presented with severe headache, abdominal pain and episodes of vomiting, examination revealed a firm to hard retroperitoneal mass right to umbilicus, she was hypertensive and had raised urinary VMA. Clinically an extra adrenal paraganglinoma was suspected. USG abdomen and CT guided FNAC confirmed the diagnosis. Tumor was removed with right mid laparotomy leading to cure of disease.

Key Words

Extra Adrenal Paraganglinoma, Benign Pheochromocytomas, Retroperitoneum

Introduction

Extra adrenal pheochromocytomas (EAPs) commonly arise in the organ of Zuckerkandl (*Fig 1*). They are tumors of APUD (Amine and precursor uptake and decarboxylation) system. Retroperitoneal tumors are almost always functional (1). Their presentation symptoms are similar to pheochromocytomas and thus can mislead many surgeons and thus the case is worth reporting. We hereby present a case of extra adrenal paraganglinoma.

Case Report

We hereby report a 35 yr old adult female, who presented with severe headache, abdominal pain and episodes of vomiting. Examination revealed a firm to hard retroperitoneal mass of 4cm. x 3cm. right to umbilicus. She was hypertensive, blood pressure ranged between 160-180 systolic and 100-120 diastolic. USG abdomen

revealed a homogenous mass of 4cm x 3 cm on right side of Inferior vena cava near the lower pole of kidney which was confirmed by CT scan. Her 24 hour urinary VMA was 17mg/litre, CT guided FNAC revealed EAP. As per the presentation and imaging and FNAC an extra adrenal paraganglinoma was suspected. Patient was put on alpha blockers, her BP settled down and she was subjected to right mid Para median laparotomy. Intraoperative her BP was maintained with nitroglycerine infusion. A tumor 4cm x 3cm was near the right side of Inferior vena cava abutting the lower pole of right kidney with few feeding vessels of the tumor. After careful dissection and ligation of vascular feeders the tumor was removed in toto (*Fig 2*). Histopathology revealed a paraganglinoma (*Fig 3*). Patient had uneventful recovery, post operatively her BP settled and she was pain free.

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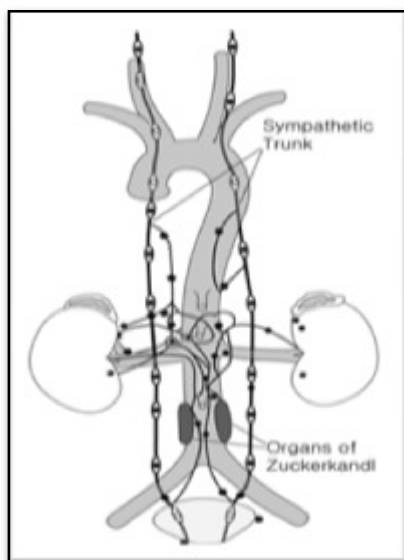


Fig.1 Specialized neural crest cells distributed along Aorta with organ of Zuckerkandle

She was discharged with advise to follow-up regularly in surgical OPD.

Discussion

Extra adrenal Paraganglinomas (EAPs) may arise in any portion of paraganglionic system though most commonly occur below the diaphragm, frequently in the organ of Zuckerkandle (1). The retroperitoneal paraganglinomas arise from specialized neural crest cells distributed along aorta in association with sympathetic chain. (Fig 1). Retroperitoneal tumors are almost always functional (2). When this tissue aggregates in adrenal medulla it gives rise to adrenal paraganglinoma known as pheochromocytoma, additionally they have been found in mesentery (3) and even intra renal where they may be mistaken as renal cyst (4). EAPs have a slight male preponderance (5) and most commonly these patients present with headache, palpitation, sweating, vomiting and episodes of paroxysmal hypertension. Our patient 35 year old adult female also presented severe headache abdominal pain and episodes of vomiting. She was hypertensive and on examination she had retroperitoneal firm to hard mass on the right side of umbilicus. On



Fig 2. Cut Section of Tumor

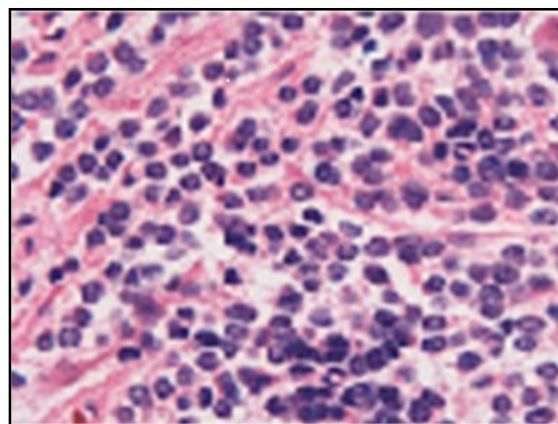


Fig. 3 Microscopic Picture of Tumour

ultrasound a homogenous mass was seen on the right side of inferior venacava. Although patient had severe pain abdomen but there were no signs of haemoperitonium as seen by others(6) where rupture of intraperitoneal paraganglinoma presented with haemoperitonium .

Biochemical investigation demonstrating elevated blood and urine catecholamines and there metabolites help in achieving the diagnosis (1) .Our patient also had high urinary VMA levels and an EAP was suspected. Imaging studies such as ultrasound, CT, MRI and ^{131}I labeled (MIBG) scan, demonstrate the tumor and its site of presence and the extent of distribution this also helps



in preoperative diagnosis (7). CT guided FNAC clinched the diagnosis. Preoperative control of hypertension with alpha blockers, control of arrhythmias with beta blockers and attentive anesthetist with careful intraoperative monitoring is required. Surgical Excision laparotomy / Laparoscopic removal (8) achieves successful outcome and cures the disease. Our patient underwent right mid paramedian laparotomy and tumor was removed. Patient improved and her symptoms disappeared. EAPs are known to reoccur and metastasise more often than their adrenal counterparts making life long follow up essential (9).

EAPs commonly arise in organ of Zuckerkandl. They are tumors of APUD system. Retroperitoneal tumors are almost always functional. Their presentation symptoms are similar to pheochromocytomas, at times they are asymptomatic or they present due to the compression of adjacent structures. The diagnosis is confirmed by demonstrating elevated blood and urine levels of catecholamines and their metabolites. They are accurately imaged by CT, MRI, and ^{131}I -MIBG syntigraphy. Preoperative assessment of the tumor and control of hypertension with pharmacological drugs and attentive anesthetist with careful intraoperative monitoring the surgical or laparoscopic removal of the tumor leads to successful outcome and cure of the disease.

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