International Journal of Medical Research Professionals

Case Report

A rare case of renal hilum extra adrenal pheochromocytoma

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Article History
Received: 29 July 2015
Revised: 4 Aug 2015
Accepted: 6 Aug 2015

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ABSTRACT

Paragangliomas are tumors that arise from neural crest tissue and can be found throughout the body. The World Health Organization advocates using the term "paraganglioma" for extra-adrenal tumours. An 18 year old girl with history of exploratory laparotomy for ileal perforation presented with several attacks of nasal bleeding, recurrent headache and dizziness since last one year. Patient was on antihypertensive medication for nine months. Patient underwent ultrasound abdomen and incidentally found to have left suprarenal mass. Contrast CT abdomen revealed a well-defined intensely enhancing 6cm×5cm×3.5cm mass in relation to anterior aspect of left kidney hilum and postero-inferior aspect of pancreas, adrenal gland can be visualised separately. There was no nodal or distant metastasis. Provisional diagnosis of extra adrenal pheochromocytoma was made. Total laparoscopic excision of tumour was performed after opening of gerota's fascia with preservation of left kidney. Histopathological examination of specimen revealed diagnosis of extra adrenal pheochromocytoma without vascular or capsular invasion. Organ of Zuckerkandl is a common site and constitutes at least 30% of childhood and 15% of adult pheochromocytomas. The elevated blood and urine levels of catechol amines and their metabolites confirms the diagnosis and they are imaged by CT, MRI, and 131I-MIBG scintigraphy accurately. Lifelong follow-up is essential as these tumours tends to recur and metastasize more than adrenal pheochromocytoma.

KEYWORDS: Pheochromocytoma, Organ of Zuckerkandl, Paragangliomas.

INTRODUCTION

Paragangliomas are tumors that arise from neural crest tissue and can be found throughout the body. The World Health Organization advocates using the term "paraganglioma" for extra-adrenal tumours, while the term "pheochromocytoma" is restricted to tumors with similar gross, microscopic, clinical, and molecular features originating from the adrenalglands[1]. About 10% of paragangliomas occur in extrarenal location; about half of these arise from the organs of Zuckerkandl and most of the remainder from the retroperitoneum. Rare sites of occurrence of paragangliomas are the bladder, urethra, prostate and seminal vesicles, kidneys and renal hilum[2]. When possible, total excision of paragangliomas is the first treatment modality and is curative in benign cases. We herein report the clinical, imaging, and pathologic features of a case of renal hilar paraganglioma.

CASE REPORT

An 18 year old girl with history of exploratory laparotomy for ileal perforation presented with several attacks of nasal bleeding, recurrent headache and dizziness since last one year. Patient was on antihypertensive medication for nine months. Patient underwent ultrasound abdomen and incidentally found to have left suprarenal mass. Her general physical examination was with in normal limits except tachycardia (pulse -95).

Contrast computed tomography of abdomen revealed a well-defined intensely enhancing 6cm×5cm×3.5cm mass in relation to anterior aspect of left kidney hilum and postero-inferior aspect of pancreas, adrenal gland can be visualised separately. There was no nodal or distant metastasis. CT angiogram revealed arterial supplies from left renal artery and small branches from the aorta with multiple draining veins. 24 hours urinary catecholamine 2022 mcg (normal 14-110mcg) and VMA level 1534mcg (normal 2-7 mcg) were significantly raised. Provisional diagnosis of extra adrenal pheochromocytoma was made. Patient was posted for laparoscopic excision of mass after appropriate premedication with alpha blocker with consent of left nephrectomy as tumour was in close vicinity of renal vessels. Total laparoscopic excision of tumour was performed in this case after opening of gerota's fascia with preservation of left kidney.

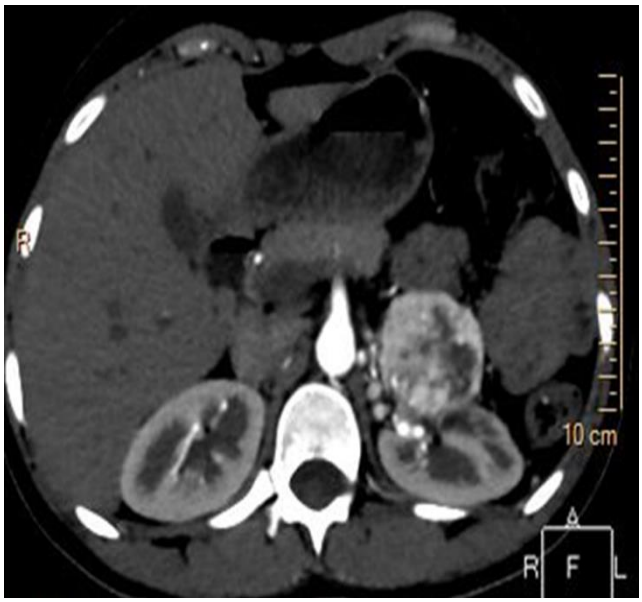

Fig 1: CECT ABDOMEN showing intensely enhancing 6cm×5cm×3.5cm mass in relation to anterior aspect of left kidney hilum and postero inferior aspect of pancreas, adrenal gland can be visualised separately.

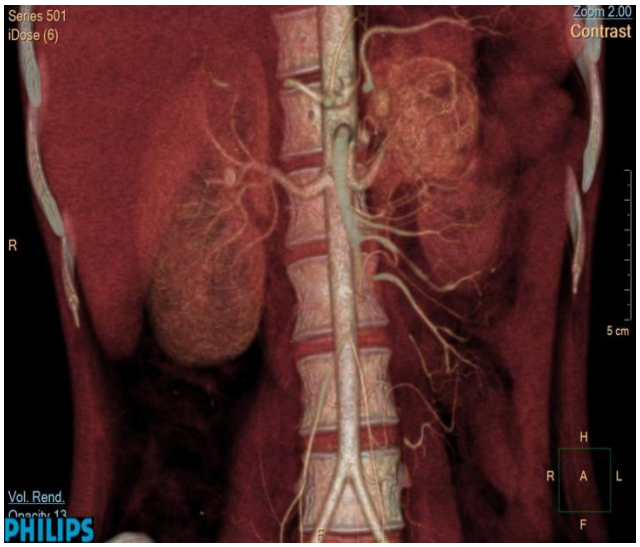

Fig 2: CT ANGIOGRAPHY depicts arterial supplies from left renal artery and small branches from the aorta with multiple draining veins.

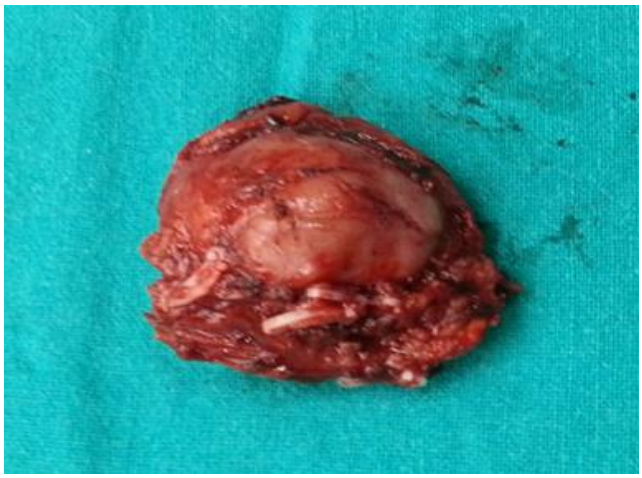

Fig 3: Specimen

Histopathological examination of specimen revealed diagnosis of extra adrenal pheochromocytoma without vascular or capsular invasion. Postoperative course of patient was uneventful and at the time of discharge her blood pressure was normal without antihypertensive medications.

DISCUSSION

Historically paraganglioma that arise from adrenal medulla is defined "pheochromocytoma", and extra-adrenal paragangliomas that are associated with clinical evidence of epinephrine secretion are defined as "extra-adrenal pheochromocytoma". However, there are no histological criteria to distinguish between functioning or non-functioning tumours[3]. Paragangliomas are secretory in more than 50% of cases, the majority of which present with palpitations[4,5]. Other typical symptoms include episodic hypertension, headache and diaphoresis. Paroxysmal hypertension is seen only in 50% of these patients[4]. Unspecific presenting symptoms such as fever and lumbar pain have been reported with cases of retroperitoneal paragangliomas[5,6]. Paraganglioma may develop in the context of type IIA and IIB MEN syndrome, neurofibromatosis, von Hippel-Lindau syndrome or Carney syndrome, but most frequently it arises spontaneously[7].

Our patient presented with history of episodic nasal bleeding, headache along with dizziness. Patient was hypertensive and tachycardic. The diagnosis of pheochromocytoma, as well as paraganlioma involves demonstrating elevated catechol amines and their metabolites in the blood and urine. Measurement of plasma free metanephrines is the single best for test for excluding or confirming pheochromocytomas. The high sensitivities of plasma free metanephrine or urinary fractionated metanephrines means that a negative test virtually excludes the presence of a functional pheochromocytoma[8]. Our patient had elevated urinary metanephrines and VMA levels.

Radiological assessments are must before surgery in evaluation for localisation, extent and multifocality of disease, as well as for the presence of metastatic disease. CT scan of abdomen and pelvis is usually the first study performed. 131I-MIBG is excellently suited as an initial localization study for EAPs. For most patients, a negative result excludes a pheochromocytomas and focus of uptake usually confirms a pheochromocytoma[9]. In the initial evaluation of pheochromocytomas, it was validated that PET-CT complements other modalities. However, the value of PET alone in determining metastatic disease is still vague due to the rarity of the tumour[6,10].

The traditional treatment of pheochromocytomas and EAPs consists of open exploration and resection. With advances in laparoscopic technique and highly refined preoperative imaging, laparoscopy is increasingly being

used in their management. Additionally, in recent years, robotic adrenalectomy has also been reported[11]. Previously nephrectomy was usually done in order to excise the tumour. In our case we were able to excise the tumour completely by laparoscopic method with preservation of kidney. Histopathology of tumour showed no vascular invasion and patients symptoms subsided after surgery.

The chance of malignancy in patients with EAPs has been reported to range from 20% to 50%, greater than the frequently quoted 10% risk for pheochromocytomas[12]. There are no pathognomonic findings that histologically distinguish EAPs as malignant versus benign, and parameters, such as mitotic rate and nuclear atypia, have not been shown to correlate with malignant potential. Histopathology of our case showed no capsular or vascular invasion. Therefore, the diagnosis of a malignant EAP is commonly made on the basis of recurrence and the development of metastasis to lymph nodes or to other organs[13]. Surveillance is advised every 6-12 months in the first 5 years post resection and then annually until 10 years post resection. It is recommended to follow up with physical exam, history and measurement of blood pressure, as well as metanephrine levels. In case of new clinical findings, imaging should be done. According to NCCN 2013 guidelines genetic counselling and testing are recommended in patients under the age of 45, or those with multifocal, bilateral or recurrent lesions, are more likely to have a heritable mutation[14].

CONCLUSION

Organ of Zuckerkandl is a common site and constitutes at least 30% of childhood and 15% of adult pheochromocytomas. Patients having EAPs usually present with symptoms similar to pheochromocytomas, asymptomatically or due to compression of adjacent structures. The elevated blood and urine levels of catechol amines and their metabolites confirms the diagnosis and they are imaged by CT, MRI, and 131I-MIBG scintigraphy accurately. PET scans hold promise for detecting metastatic disease and useful in follow-up to detect recurrences. Recent reports suggest that a laparoscopic or robotic assisted approach, along with intraoperative ultrasound, can safely remove these tumours with minimum invasion. Lifelong follow-up is essential as these tumours tends to recur and metastasize more than adrenal pheochromocytoma.

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How to cite the article: Kaswan RS, Tyagi VK, Patel B, Gurjar KS, Pahwa M, Chadha S. A rare case of renal hilum extra adrenal pheochromocytoma. Int J Med Res Prof. 2015,1(2);14-16.