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Case Report

Paraganglioma of the left common iliac artery: case report and review of the literature

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ABSTRACT

Paraganglioma is a neoplasm derived from the neural crest of the neuroendocrine system. Head and neck account for its more frequent localizations. Parasymphatic paraganglioma have been encountered at cauda equina and iliac vessels. A 37–year old female patient with abdominal pain diagnosed as adnexal mass was surgically operated on at the SMS medical college and research group in Jaipur, Rajasthan, India. During surgery, a retroperitoneal mass was found located lateral to the common left iliac artery. It was completely resected. Microscopic study established characteristics of paraganglionoma. Patient outcome was good. Scanning for other tumour site was negative.

Keywords: Pheochromocytoma, Extraadrenal, Paraganglioma, Neuroendocrine tumours, Organs, Patients

INTRODUCTION

Pheochromocytoma accounts for 0.1% of hypertensive patients. Most of them (90%) arise from the adrenal medulla, whereas 10% are extra-adrenal. Extra-adrenal pheochromocytomas are called paragangliomas and may arise anywhere but are mostly located in the retroperitoneum arising from the sympathetic chain or from the organ of Zuckerkandl.[1] Patients with such tumors typically present with symptoms resulting from excess production of catecholamines.[2]

CASE REPORT

A 37 year old female was admitted to the surgical emergency ward with complaints of abdominal pain radiating to the back for past one week, she had giddiness associated with sudden postural changes, sudden paroxysm of sweating and palpitation. No headache or vomiting. He had no co morbidities, no surgeries in the past. On examination, she was not anemic, no pedal oedema, pulse rate was normal and severe hypertension (180/100 mm Hg in the left arm sitting), cardio respiratory system clinically normal, no masses in the abdomen, no swelling in the neck. Patient had fluctuating hypertension (110-190), put on anti-hypertensive. Patient had two episodes of sudden giddiness, sweating and hot flushing of face during hospital stay.

Investigations

Complete hemogram, liver and renal function tests were within normal limits, normal electrocardiograph and normal chest radiograph. Her abdominal x-ray and ultrasound examinations were with no abnormality detected. Her chest computed tomography (CT) was normal. CT abdomen and pelvis showed a heterogeneous intensely enhancing mass lesion with mild internal necrosis and significant enhancing solid component seen in the left lower abdomen /pelvis likely left adnexal in origin abutting the left fallopian tube and left ovary which. Ovarian markers were within normal limits.

Put on alpha blocker-prazocin 1 mg and analapril 5 mg. Patient was taken up for laparotomy for excision of tumour. Transperitoneal approach was followed.

Intraoperatively there was high BP (180/100). Direct vascular connections from left common iliac artery and venous tributaries to IVC were ligated. Tumour was removed en- bloc. Post operatively maintained the blood pressure with tapering of alpha blockers. Patient had an uneventful post-operative recovery. The histopathology confirmed the diagnosis of paraganglioma.

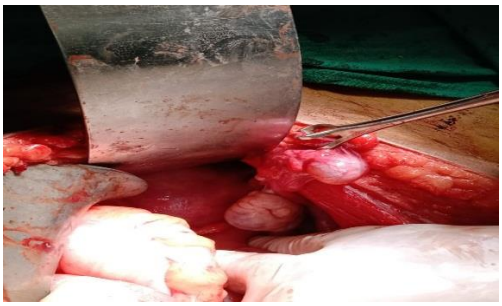

Figure 1: Intraoperative picture showing left common iliac paraganglionoma.

Figure 2: Postoperative healthy wound.

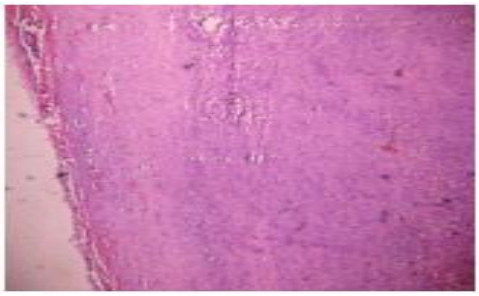

Figure 3: Histopatholgy showing well encapsulated tumor.

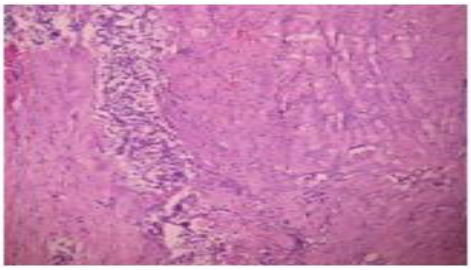

Figure 4: Histopathology – paraganglioma.

DISCUSSION

Phaeochromocytoma is a functioning tumor arising in the catecholamine producing chromaffin cells derived from neural crest ectodermal in origin). The first case described by Frankel in 1886 was an 18 year old presented with hypertension, sudden collapse and death and postmortem demonstrated bilateral adrenal tumors.[3] This is explained by the paroxysmal release of excess catecholamines into systemic circulation. This functional extremism can also present with massive myocardial infarction and cerebrovascular events. Intraabdominal tumors constitute 98% of the phaeochromocytomas and the rest are found in the neck and mediastinum. Of the intraabdominal tumors more than 85% are adrenal tumors. The remaining is distributed along the sympathetic ganglia chain in the retroperitoneum. These extra-adrenal tumors are called paragangliomas. Pheochromocytoma has an incidence of 2 to 8 cases per million person's annually.[4] Of this pheochromocytoma arising in extra adrenal locations is less than 10% only. It is mostly discovered during autopsy/ incidental workup. 75% died suddenly from either myocardial infarction or a cerebrovascular catastrophe. One third of the sudden deaths occurred during or immediately after unrelated minor operations.

Hypertension is the commonest clinical sign though it contributes less than 0.5% of the newly diagnosed hypertensive's.[5] These tumors are bilateral in 10% cases and in view of wide range of possible locations are located with appropriate imaging. Due to the low lipid content of these tumors compared to the other adrenal tumors they may be easily distinguished from them in imaging.[6] Contrast enhanced computed tomography (CECT) or magnetic resonance imaging (MRI) can be used since most of the tumors are more than 3 cm in size. In doubtful cases metaiodo benzyl guanidine (MIBG) scintigraphy can also be useful.[7] Presence of malignant phaeochromocytomas is a well-established entity and no reliable histological feature is confirmatory but only presence of metastasis confirms the tumor to be malignant. Adrenal phaeochromocytomas are less likely to be malignant than paragangliomas (10% versus 35%).[8] In our case patient has benign paraganglioma confirmed by absence of metastasis in other parts of the body. After confirming the diagnosis and the location of the tumor it is imperative to undertake complete surgical resection of the tumor though special preoperative preparation has to be done adequately. These patients are in a state of contracted intravascular volume and hence adequately hydrated and put on alpha adrenergic blockers like phenoxybenzamine or prazocin and beta blockers if needed to control tachycardia or cardiac arrhythmias.[9] In our patient reversible alpha adrenergic blocker prazocin is used with enalapril. Laparascopicadrenalectomy has become the gold standard except in the presence of large tumor or malignant disease.[10] We preferred to resort to open laparotomy as exploratory laparotomy. Patients are kept on postoperative follow up for prolonged period to diagnose early of

recurrence which is reported in more than 15% cases even after 10 years.[11]

CONCLUSION

Even though paraganglioma is rare, it should be considered in the differential diagnosis of hypertension associated with incidentaloma and/or with unexpected drug-induced blood pressure changes.

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Ethical approval: Not required

REFERENCES

1. de Graaf JS, Nieweg OE, Oosterkamp AE, Zwierstra RP. Results of 25 years of treatment of Phaeochromocytoma. Nederlands Tijdschrift voor Geneeskunde. 1997;141:148-51.
2. Tohya T, Yoshimura T, Honda Y, Miyoshi J, Okamura H. Unsuspected extra-adrenal pheochromocytoma simulating ovarian tumor. Eur J Obstet Gynecol and Reprod Biol. 1999;82:217-8.
3. Messerli FH, Michalewicz L. Clinical and experimental pheochromocytoma. JAMA. 1997;278(1):78-9.
4. Lenders JW, Eisenhofer G, Mannelli M, Pacak K. Phaeochromocytoma. Lancet. 2005;366(9486):665-75.
5. Lenders JW, Pacak K, Walther MM, Linehan WM, Mannelli M, Friberg P, Keiser HR, Goldstein DS, Eisenhofer G. Biochemical diagnosis of pheochromocytoma: which test is best? JAMA. 2002;287(11):1427-34.
6. Motta-ramirez, Remer EM, Herts BR. Comparison of CT findings in symptomatic and incidentally discovered pheochromocytomas. AJR Am J Roentgenol. 2005;185(3):684-8.
7. Ilias I, Divgi C, Pacak K. Current role of MIBG in the diagnosis of pheochromocytoma and medullary thyroid cancer. SeminNucl Med. 2011;41(5):364-8.
8. Scholz T, Eisenhofer G, Pacak K. Clinical review: Current treatment of malignant pheochromocytoma. J Clin Endocrinol Metab. 2007;92(4):1217-25.
9. Pacak K. preoperative management of the pheochromocytoma patient. J Clin Endocrinol Metab. 2007;92(11):4069-79.
10. Pacak K, Eisenhofer G, Ahlman H. Pheochromocytoma: recommendations for clinical practice from the first international symposium. October 2005. Nat Clin Pract Endocrinol Metab. 2007;3(2):92-102.
11. Khorram-Manesh, Ahlman H, Nilsson O. long term outcome of a large series of patients surgically treated for pheochromocytoma. J Inten Med. 2005;258(1):55-6..

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