

# Extra-adrenal Non-functioning Paraganglioma – A Diagnostic and Surgical Challenge

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## Research Article

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# Abstract

Paragangliomas are rare neoplasms arising from the neuroendocrine cells. Extra-adrenal paragangliomas arise from the neuroendocrine cells spread in the retroperitoneum from the skull base to the retroperitoneum and sacrum. Non-functioning paragangliomas are a diagnostic challenge as they are clinically silent and attain large size. The clinical implication of these non-functioning paragangliomas is the greater metastatic potential of extra adrenal paraganglioma (20–42%) when compared to adrenal paraganglioma.

We report a very rare case of a 60-year lady who presented with abdominal heaviness and retroperitoneal lump. Imaging was suggestive of a retroperitoneal tumor. Serum chromogranin A level was elevated with normal catecholamine levels. The tumor was resected along with metastasectomy of isolated liver metastasis. The patient has no recurrence or metastasis at one year of follow-up.

To conclude paraganglioma should be considered as a differential diagnosis in all retroperitoneal tumors. Serum chromogranin A levels and plasma metanephrines should be routinely done for all retroperitoneal cases before planning for the biopsy of the lesion. Resection of the lesions should always be performed as per the oncological principles as pre-operative differentiation of benign and malignant lesions is difficult.

## Introduction:

Paragangliomas are rare neoplasms arising from the neuroendocrine cells. The adrenal gland is the most common site for paraganglioma as it is the largest collection of neuroendocrine cells in the body. Extra-adrenal paragangliomas arise from the neuroendocrine cells spread in the retroperitoneum from the skull base to the retroperitoneum and sacrum. They constitute about 10% of the paragangliomas [1]. The most common site for extra-adrenal paraganglioma is the organ of Zuckerkandl present between the origin of inferior mesenteric artery and aortic bifurcation and was named after Zuckerkandl who has first described the distribution of paraganglia in a human fetus [2]. Extra-adrenal paragangliomas are difficult to diagnose owing to their location. Clinically, vast possibilities of differential diagnosis are possible. Non-functioning paragangliomas further add to the diagnostic difficulty. Due to a higher proportion of malignant lesions of extra-adrenal paragangliomas accurate pre-operative diagnosis is essential. The below case report is one such scenario of non-functioning extra-adrenal paraganglioma.

## Case Report:

A 60-year lady presented to the surgical department with complaints of heaviness in the upper abdomen for three years and a lump in the right flank which was gradually increasing in size from one year. There were no urinary symptoms or symptoms of headache, palpitations, anxiety or diaphoresis. There was no significant past, personal or family history. On examination her vital parameters were normal. There was an ill-defined retroperitoneal lump in the right lumbar region with right renal angle fullness. On contrast-

enhanced computed tomography (CECT) of the abdomen, there was a large well circumscribed moderately enhancing retroperitoneal mass in the right lumbar region, measuring approximately 14 x 8.5 x 7.8 cm, abutting the right renal pelvis and compressing and displacing right proximal ureter with loss of fat planes with the underlying psoas muscle [Figure 1]. Serum chromogranin A level was raised. Plasma and urine metanephrines were within normal limits. CT chest was normal. Patient was taken up for laparotomy with a working diagnosis of non-functioning paraganglioma.

On laparotomy, there was a huge retroperitoneal mass displacing the right kidney [Figure 2]. The tumor was invading the adjacent psoas muscle which and was resected with adequate margins. There was a single 1 x 1 cms metastatic lesion in segment five of the liver, which was missed on CT and was resected. Patient had an uneventful post-operative recovery. The histopathological report was suggestive of malignant paraganglioma with metastasis. Ga<sup>68</sup> labeled tetraazacyclododecane tetraacetic acid (DOTA) coupled peptide positron emission tomography (PET) scan in the post-operative period was not suggestive of any residual metastasis. The patient has no recurrence or metastasis at one year of follow-up and prognosis was good.

## Discussion:

The median age at diagnosis of paraganglioma is 37–43 years with similar incidence in both males and females [2, 3]. A significant proportion of paragangliomas occur as a part of genetic syndromes such as Von Hippel Lindau syndrome, Multiple Endocrine Neoplasia type 2, Neurofibromatosis, Carney's triad, etc. About 10–15% of paragangliomas are non-functional and about 10% of patients with functional tumors do not have any overt symptoms [4]. Non-functioning paragangliomas are a diagnostic challenge as they are clinically silent and attain large size [4]. The clinical implication of these non-functioning paragangliomas is the greater metastatic potential of extra adrenal paraganglioma (20–42%) when compared to adrenal paraganglioma [3].

Non-functioning paragangliomas present with vague symptoms such as backache, abdominal heaviness, early satiety and urinary complaints. Some of these tumors may even present with atypical symptoms such as paralytic ileus [5]. Imaging studies are the mainstay for the diagnosis of these silent tumors. CECT and magnetic resonance imaging (MRI) are commonly used. Paragangliomas appear as enhancing lesions with areas of necrosis, hemorrhage and calcification. CT angiography may be required to identify vascular invasion. Metaiodobenzylguanidine (MIBG) and DOTA-PET scans can be used for imaging of doubtful cases and to assess the metastasis of paragangliomas [6]. Serum chromogranin A levels can be useful to rule in neuroendocrine tumors when a retroperitoneal tumor is suspected on imaging. Plasma and urinary catecholamines are used to diagnose functional paragangliomas.

All paragangliomas need surgical excision as per the oncological principles as they harbor malignant potential. Preoperatively presence of metastasis is the only sign to differentiate malignant and benign tumors. These tumors are an operative challenge to the surgeon as they are highly vascular and are present in difficult to access locations. Preoperative embolization will reduce the size of the tumor and

also decreases the intra-operative hemorrhage. The abdominal cavity should be examined for metastasis before the resection of the tumor. The most common sites for metastasis include the lymph nodes and the liver. Metastatic disease should be treated palliatively with octreotide, peptide receptor radionuclide therapy and radiation [7].

## **Conclusion:**

To conclude, the above case report is a rare case of extra adrenal non-functioning retroperitoneal paraganglioma. Paraganglioma should be considered as a differential diagnosis in all retroperitoneal tumors. Serum chromogranin A levels and plasma metanephrines should be routinely done for all retroperitoneal cases before planning for the biopsy of the lesion. Resection of the lesions should always be performed as per the oncological principles as pre-operative differentiation of benign and malignant lesions is difficult.

## **Declarations**

## **Funding:**

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## **Conflict of interests:**

Authors declare that they have no conflicts of interest.

## **Ethics Approval:**

Ethics approval is not required for publishing an anonymous case report in our institute.

## **Consent for participation and participation:**

Written informed consent was obtained from the patient to publish his personal and clinical details along with identifying images.

## **Availability of data and material:**

Available on personal request from authors.

## **Code availability:**

Not applicable.

## Author's Contributions:

RY evaluated the case and wrote the manuscript. NG operated the case and reviewed the manuscript. MDM reviewed the manuscript and helped in data search. CKD reviewed the manuscript and guided the study.

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## Figures

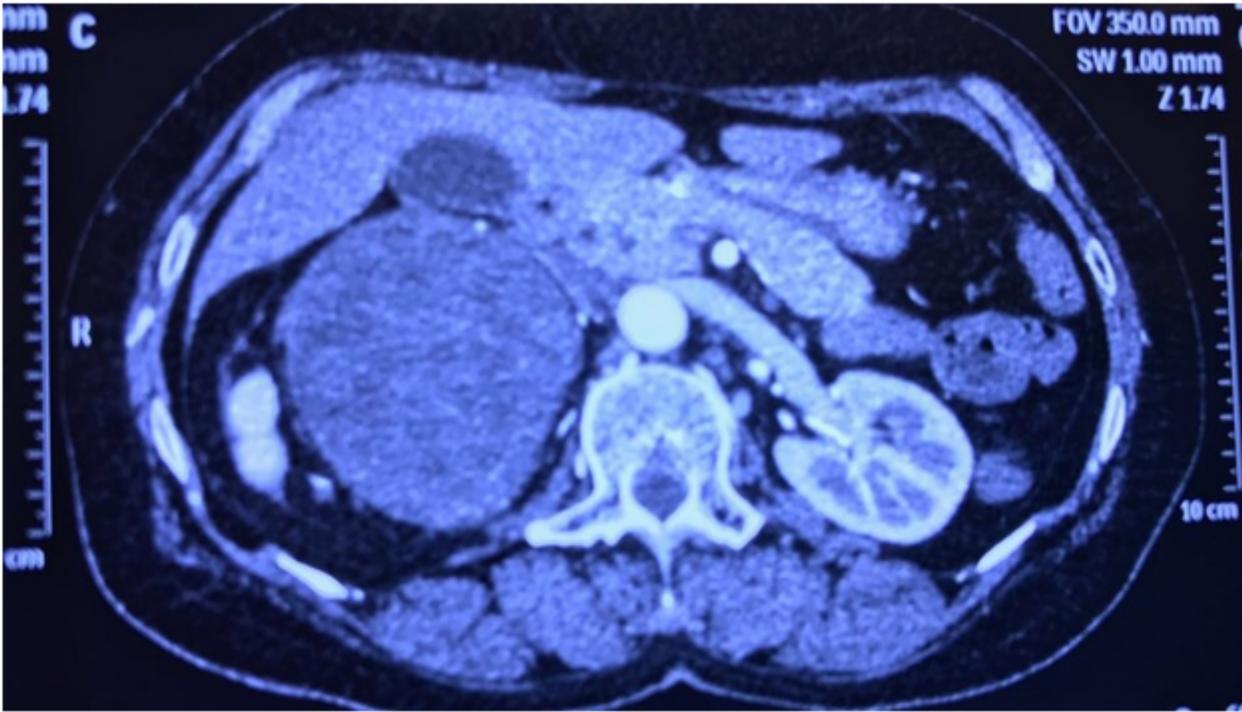


Figure 1

CECT image showing the retroperitoneal tumor

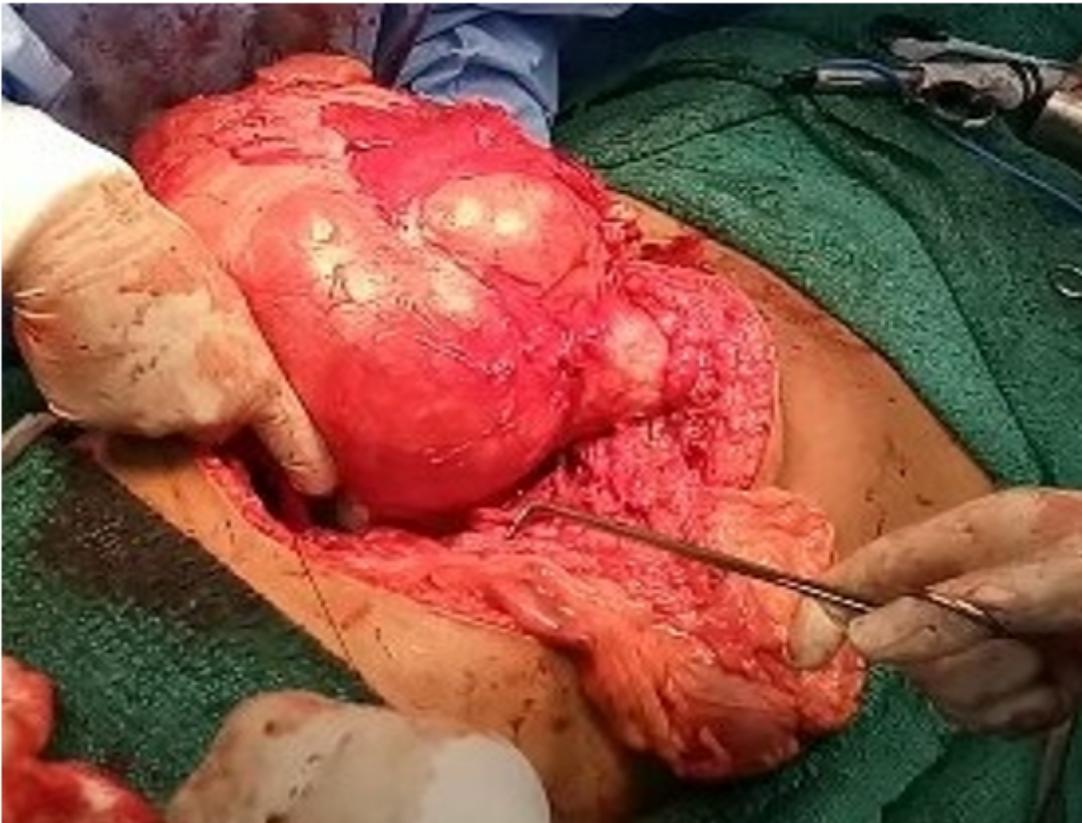


Figure 2

Intra-operative image of the tumor during resection

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