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# A Brief Study on Retroperitoneal Paraganglioma

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***Authors' contributions***

*This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.*

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***Case Report***

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## ABSTRACT

Paragangliomas are tumors of the autonomic nervous system, located extra-adrenally, presenting inside the base of the head, chest and abdomen. They can appear as a primary retroperitoneal neoplasm when viewed inside the abdominal cavity and might resemble as vascular malformations or any other conditions same as retroperitoneal organs particularly such as pancreas, kidney. These are relatively uncommon endocrine tumors that originate from specific tissue of the paraganglion. They synthesise, store and secrete catecholamines, which can contribute to headache, sweating, palpitation and hypertension. These may be mistaken for GISTs with no histological diagnosis and symptoms of catecholamine overload (non functional). We are reporting a case of 42 yrs male with clinical features of slow backache radiating to both lower legs and with the presence of tingling sensation. Corresponding contrast enhanced CT scan shows a para – aortic mass.

*Keywords: Paraganglioma; catecholamines; retroperitoneal organs.*

## 1. INTRODUCTION

Sometimes referred to as extra adrenal pheochromocytomas, paragangliomas are infrequent neuroendocrine neoplasm of the paraganglia, a diffuse neuro-endocrine system that is spread from the base of the skull to the pelvic floor, and these tumors are seen in people

of all age groups [1,2]. Few tumors are functioning paragangliomas that synthesize, store and secrete catecholamines [3]. Higher urine/serum catecholamine levels and classical clinical features such as episodic headache, sweating and palpitation are often present [4]. Around ten to fifteen percent of tumors are not functional. Mostly they are locally invasive and are linked with a high local recurrence rate [5]. A major diagnostic issue is seen while diagnosing a non-functional paraganglioma. There are no special imaging features noted exclusively for paraganglioma in abdominal CT [6,7]. Consequently these tumors can be confused for other abdominal tumors that can be primary epithelial or mesenchymal tumors [8].

## 2. CASE PRESENTATION

A 42 yr old male came with the complaints of aching back and pain in the abdomen. This pain was radiating downwards to both lower legs along with tingling sensation. There was no history of vomiting or changed intestinal patterns with unremarkable physical test. Palpable mass was absent. CECT abdomen and chest done on 18/06/2020 and 19/06/2020

CECT abdomen:- Revealed heterogeneously enhancing lobulated lesion in para aortic region, measuring 7x5.2x3.4 cm. Multiple small necrotic areas are noted within. No calcification is present but displacement of the neighboring vessels is present. There was no infiltration into the adjacent structures [9,10].

CT guided FNAC and core biopsy was done. CECT chest was unremarkable. On Histopathology of core biopsy and cytology- it was reported as Poorly Differentiated Malignant Tumor and Immunohistochemistry was advised. Paraffin blocks were sending to SRL Mumbai for IHC, where it was reported as Pheochromocytoma/Extra-adrenal
Paraganglioma. These tumor cells expressed synaptophysin and chromogranin A, S-100 protein highlighting sustentacular cells and were immune-negative for Calretinin. The Mib-1 labeling index was approximately 5 % in areas of highest proliferative activity. Decision was taken to excise the mass, and on 30/07/2020, exploratory laparotomy and excision of left para-aortic mass was done [11,12].

On gross examination - Mass was of size 6.5x4x3 cm, cut section was brownish in colour and soft in consistency. Areas of haemorrhage and necrosis were seen. On Microscopic examination it showed nests and trabeculae of oval to polygonal cells having granular eosinophilic cytoplasm and round to oval nuclei along with giant multinucleated cells were seen. So it was reported as Paraganglioma as it was confirmed by IHC and core biopsy.

## 3. DISCUSSION

Paragangliomas are unusual tumours originating from neural crest cells that arises from sympathetic and parasympathetic paraganglia. Extra-adrenal paragangliomas in the autonomic nervous system originate from chromaffin tissue and may be located in the head, neck, thorax, abdomen and pelvis. Retroperitoneal paragangliomas are tumors that are uncommon, out of which the nonfunctioning ones usually remain in isolation. They are distinguished by their asymptomatic nature with normal levels of catecholamine in urine and blood. For retroperitoneal paragangliomas the median diagnostic age is 37-43 years with similar incidence among males and females. However non-specific features such as lower back pain, abdominal heaviness, urinary symptoms or changes in general health will also take place in patients but may have in the middle a cystic or necrotic appearance, or it may appear calcified. Finally, a definitive diagnosis can be reached only via histology. Paragangliomas with chief cells and subtentacular cells grouped in clusters are called as Zellballen pattern and are histologically diagnosed with their highly vascular appearance. Chief cells are also immunohistochemically positive for neuroendocrine markers (synaptophysin, NSE, chromogranin), whereas sustentacular cells are S100 protein positive. However, since these tumours are potentially malignant, complete excision is the cornerstone of curative care. The tumor is cancerous may only be confirmed by the presence of distant metastasis. Lymph nodes, bone, liver, and lung are sites of such metastasis. Follow up is carried out with meta iodobenzyl-guaninidine scintigraphy showing the presence of metastasis or any foci of recurrence detection. Since complete mass excision has been achieved in our case, CECT abdomen of the patient is followed up for every 3 months in the first year.

## 4. CONCLUSION

This case highlights the need for the inclusion of extra additional adrenal paraganglioma in the differential diagnosis and treatment of retroperitoneal tumors considering its uncommon

occurrence. Role of histology remains the mainstay of the diagnostic approach towards non-functional retroperitoneal gangliomas. The single treatment of choice is surgical excision. As recurrence and metastasis are normal, with ultrasound and CT scans, lifelong follow up is required.

## DISCLAIMER

The products used for this research are commonly and predominantly use products in our area of research and country. There is absolutely no conflict of interest between the authors and producers of the products because we do not intend to use these products as an avenue for any litigation but for the advancement of knowledge. Also, the research was not funded by the producing company rather it was funded by personal efforts of the authors.

## CONSENT

As per international standard or university standard, patient's written consent has been collected and preserved by the author(s).

## ETHICAL APPROVAL

Ethical clearance taken from institutional ethics committee

## COMPETING INTERESTS

Authors have declared that no competing interests exist.

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