

Mysterious Mass in the Retroperitoneum Space: Case Report (Retroperitoneal Extra Adrenal Paraganglioma)

Dr. Rakshith, Dr. Jagadeesha BVC, Dr. Mahesh Kariyappa

Abstract: Extra- adrenal retroperitoneal paragangliomas are extremely rare neuroendocrine neoplasms with an incidence of 2-8 per million. They emanate from embryonic neural crest cells and are composed mainly of chromaffin cells located in the para- aortic sympathetic chain. It is a kind of pheochromocytoma which occurs on the outside of the adrenal gland. They synthesize, store and secrete catecholamines due to which they may present with symptoms of hypertension like headache, sweating and palpitation and non functional paraganglioma sometimes they may present with vague symptoms like pain abdomen and lump abdomen. Primary methods of pre-operative diagnosis include imaging techniques which also help in surgical planning and pre-operative preparation of the patient. We present a case of non- functional extra- adrenal retroperitoneal paraganglioma occurring in a 40-year-old male patient presenting with mass per abdomen. On Ultrasonography, suspicion was towards a retroperitoneal mass of probable retroperitoneal cyst. CT is the investigation of choice. Surgical resection is main modality of treatment. Confirmation of the diagnosis is done histopathological examination and immunohistochemistry markers.

Keywords: retroperitoneal tumor: CD- Cluster of differentiation: paraganglioma

1. Introduction

Paragangliomas (also known as extra-adrenal pheochromocytomas) are rare tumors that arise from extra-adrenal chromaffin cells¹. Retroperitoneal paraganglioma represents between 21.5 and 87% of all paragangliomas². However, between 40 and 50% of paragangliomas are non-functional and potentially functional³. Since retroperitoneal paragangliomas are rare, the behavior and treatment outcomes of this type of tumor remain unclear.

2. Case report

A 40 years old male patient presented to surgical OPD with the chief complaint of mass in the left upper abdomen since 3month and pain abdomen since 3 month.

Initially when the patient noticed the mass in the left upper abdomen in which was size 5*6cm. There was no increase or decrease in the of the mass. No history disappearance of since in the period of 3 month. Patient complaint of pain abdomen since 3 month colicky in nature, mild to moderate in intensity. No history radiation or diurnal variation of the pain.

There was no history of vomiting or altered bowel habits or previous history of similar attacks. The patient was not a known hypertensive and was not on any medication.

On examination the was moderately build and nourished and well oriented with time place and person.

His was vital was stable at the time of examination and general condition of the patient was normal.

Per abdomen examination revealed a mass size approximately 5*6cm in the left lumbar region extending into left hypochondriac region which did not move with respiration but side to side movement was present, globular in shape, rounded margins, smooth surface, firm in

consistency, non ballotable and non pulsatile. Bowel sounds and rectal examination were normal.

Ultrasonography was diagnosed as the retroperitoneal mass probably Retroperitoneal cystic lesion /long standing calcified aortic aneurysm.

CECT abdomen showed cystic mass in the retroperitoneum of size 82mm*56mm which is closed to abdominal aortic wall (retroperitoneal paraganglioma)

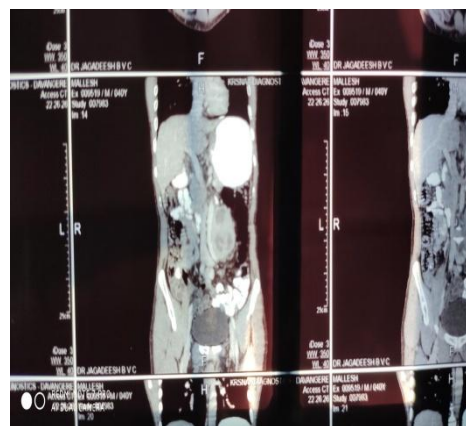


Figure 1: Sagittal view of mass

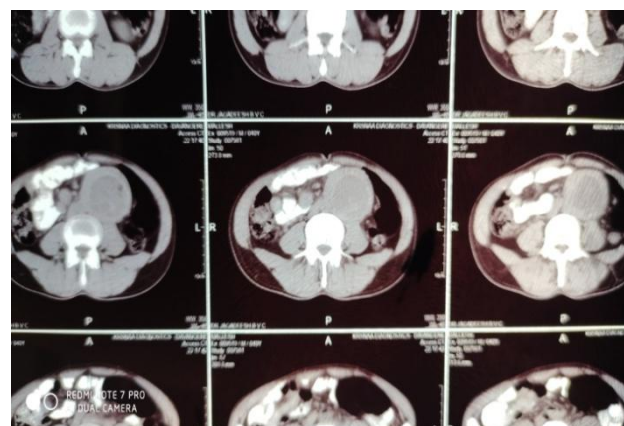


Figure 2: Cross section view of paraganglioma

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The hematological and biochemical profiles were normal. Laparotomy was undertaken for exploration was done by putting lazy S incision on left side of the abdomen. Per-operatively, a retroperitoneal mass was found which was highly vascular. The mass was found located anterior to the sympathetic and lymphatic chain in the para-aortic region

which was just adherent to the Aorta. While attempting to separate the mass. Further dissection revealed that the mass was arising from the sympathetic chain. The mass was completely resected and was sent for histopathology.



Figure 3: Operative finding and images of paraganglioma

Histopathological report

Multiple section showed a well encapsulated lesion with dense fibrous tissue along with cells arranged in nest (Zellballen) pattern, cell have mildly pleomorphic round

oval nuclei, coarse chromatin and indistinct eosinophilic granular cytoplasm. With impression the feature suggestive of paraganglioma

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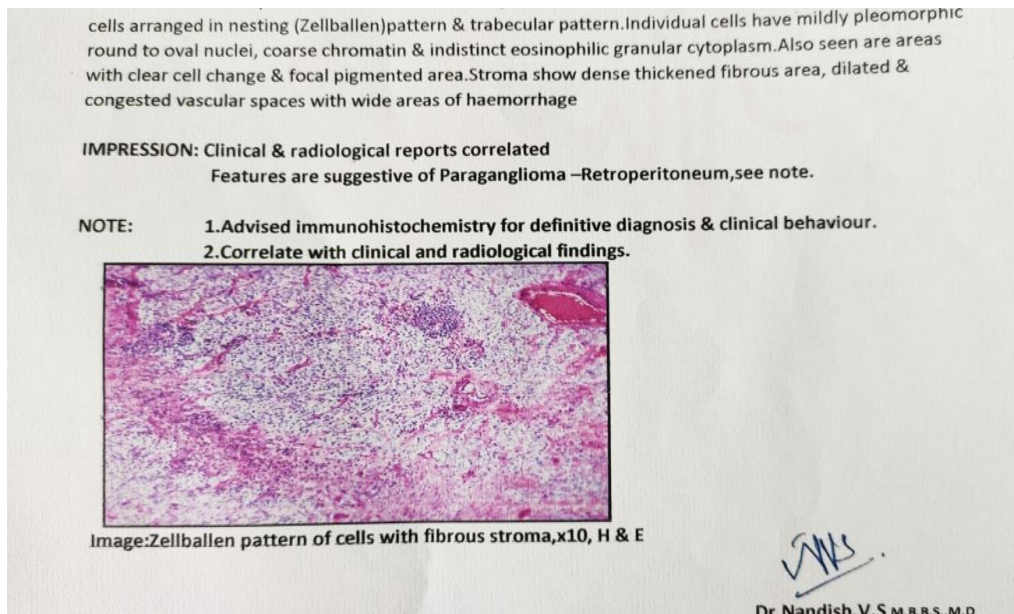


Figure 4: Histopathological finding of paraganglioma

3. Discussion

Paragangliomas are rare neuroendocrine tumours emanating from specialized cells referred to as chief cells of paraganglia which are spread over the entire body ranging from the skull to the pelvic floor. Regardless of their location, these are referred to as “paragangliomas”⁴.

The term “extra adrenal paraganglioma” is appropriate for paragangliomas situated outside adrenal medulla. Most of these paragangliomas are known to occur in specific locations like the carotid body, jugular foramen, middle ear, aortico-pulmonary region, posterior mediastinum and abdominal para aortic region including the Zuckerkandl’s body or aortic body. Some of the paragangliomas are known to create, store and secrete catecholamines and are hence termed “functional paragangliomas”. While these are diagnosed easily, non-functional paragangliomas may create major difficulties in pre-operative diagnosis [6].

A number of factors contribute to the difficulties encountered in the management of extra-adrenal paragangliomas. They include the rarity of extra-adrenal paragangliomas which account for just 2-8 per million incidence, the insufficiency of information available regarding their natural history and outcome and the non-availability of a definitive pre-operative diagnosis especially in the case of a non-functional tumour [7].

Extra-adrenal paragangliomas can be uni or multicentric with a tendency for local invasion and therefore a thorough evaluation needs to be undertaken to rule out multicentricity. Periodic follow-ups will have to be scheduled to rule out recurrence. 24- 50 % of extra-adrenal paragangliomas are malignant in contrast to malignant pheochromocytomas which have an incidence of 10%. The tumour in the present case was benign, unicentric and lacked local invasion [8].

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