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Retroperitoneal NonFunctioning Paraganglioma with Solitary Sternal Metastasis – A Rare Presentation

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ABSTRACT

Paragangliomas are rare and arise from neural crest cells. They are often difficult to diagnose because of varied clinical presentation.

Reporting a case of 47 year male presenting with lower abdominal pain, diagnosed to have a retroperitoneal tumour situated at the aortic bifurcation. Serum catecholamine levels were within normal limits. Complete resection of the tumour was done in June 23. The HPE and IHC analysis was suggestive of intermediate risk paraganglioma. Patient was on regular follow up.

In December 23, he presented with complaints of pain over the sternum . On evaluation , he was found to have a solitary lesion in the sternum , which was DOTA avid. 2 cycles of Lu therapy was tried, but in view of progressive nature of the disease and considering the fact that it was the solitary site of metastasis, he was taken up for sternal resection with reconstruction in Jan 23. Postop HPE was suggestive of a paraganglioma.

Keywords: Paraganglioma , Neuroendocrine tumour

INTRODUCTION

Paragangliomas are neuroendocrine tumours arising from neural crest cells [1]. Tumours derived from adrenal medulla are more common than extra abdominal tumors. 10% of paragangliomas arise outside the adrenal glands [2]. The most common site of extra-adrenal tumors is the para aortic region. [3].

They may be functional due to excess catecholamine release. Approximately 10–15% of such tumours are non-functional, and another 10% have hormone activity but do not present clinically. They are often locally invasive and associated with increased risk of local recurrence [4].

Genetic mutations happen in 25% cases , with mutations in the SDH gene being the most common. Patients with a mutation in the B subunit of SDH, are more likely to have metastatic disease - lung , liver , bone , spleen . Here we report a rare case of retroperitoneal paraganglioma , which presented with a solitary sternal metastasis

withtin 6 months of the operation of the primary site of disease.

PRESENTATION OF CASE

A 47 year old male came with the complaints of vague lower abdominal pain since 1 year, dull aching type , intermittent in nature. He is a known hypertensive, on regular medications and his blood pressure was 140/90 mmHg at the time of admission, with no history of headaches/ palpitations/ excessive sweating . There was no family history of illness. The physical examination was unremarkable with no palpable abdominal mass.

CT abdomen showed a 4x3x3 cms solitary well-defined mass at L5-S1 level abutting the right common iliac blood vessel **(Fig. 1)** which was likely to be a paraganglioma. PET CT whole body was done which was suggestive of a 4.6 x 4.2 cms just below the aortic bifurcation , abutting the right common iliac / internal iliac artery and prevertebral fascia

(Fig. 2), no distant metastasis was noted. Serum chromogranin A level was within normal limits.

After obtaining anaesthetist fitness , patient was taken up for laparotomy in June 2023. The lesion was completely resected through a midline laparotomy incision.

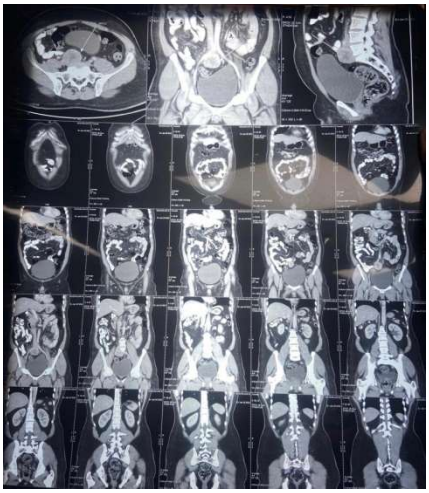
On grossing, mass measured 10x5x4 cms ,was encapsulated with a variegated orange/brown cut surface**.** Histological examination showed a encapsulated tumour composed of compact nests of varying sizes. Zellballen pattern of paraganglioma was apparent. No capsular invasion / lymphovascular invasion was present. GAPP score of 4/10 noted. Diagnosis of intermediate risk retroperitoneal paraganglioma was made, with a Ki67 index of 4%.

Genetic testing was positive for SDH B mutation. Postoperative period was uneventful and patient was discharged on POD 7. Patient was on regular followup.

In December 2023 (6 months postoperatively) patient came with the complaints of pain over the sternum . CECT thorax revealed a 5x3x2.4 cms lesion involving the body of sternum **(Fig. 3).** Tumor Board concurrence was obtained for Lu therapy , in view of solitary site of metastasis and DOTA avidity . Patient underwent 2 cycles of Lu Therapy , following which pain over the sternum increased. Ga 68 DOTANOC PET CT whole body was done which revealed an increase in the size of the lesion involoving the sternum (6.5x3.8x2.6 cms) **(Fig. 4).**

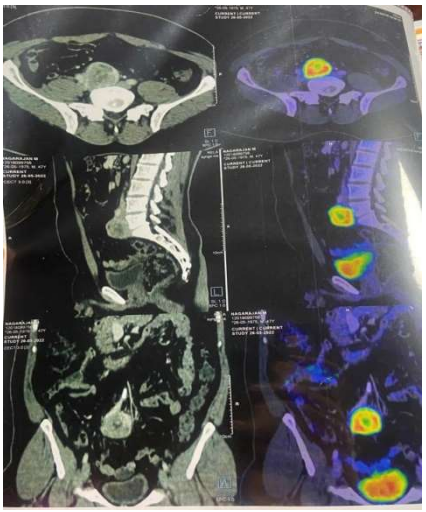
After obtaining anaesthetist fitness, patient underwent sternal resection with reconstruction using sandwich technique (mesh and cement) **(Fig. 5).** Histopathology was suggestive of a metastatic paraganglioma.

Postoperative period was uneventful. Patient is at present symptomatically better and on regular follow up.



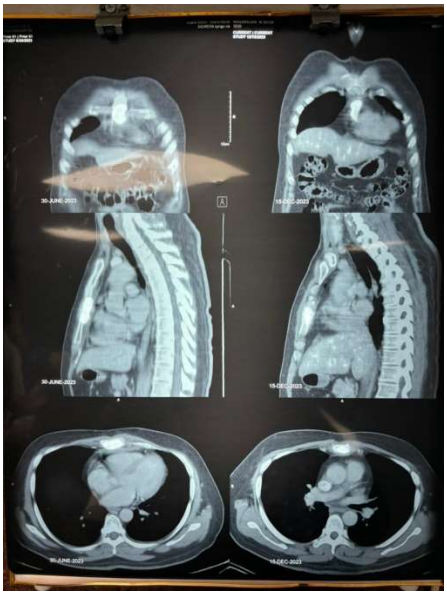
CT abdomen : Retroperitoneal paraganglioma below the aortic bifurcation

(Fig. 1)

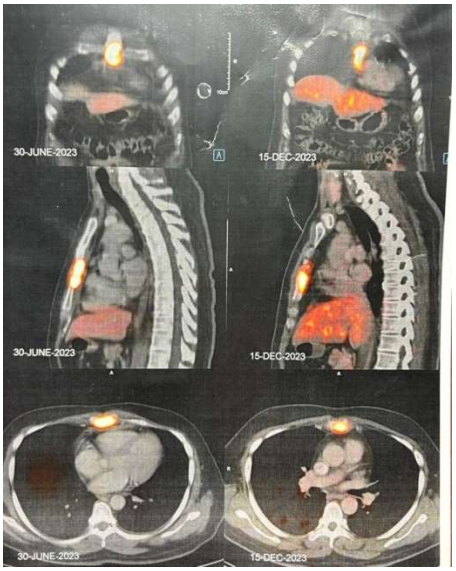


PET CT : DOTA avid Retroperitoneal paraganglioma below the aortic bifurcation

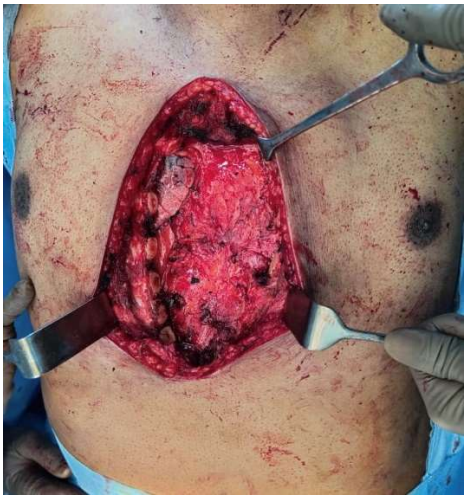
(Fig. 2)



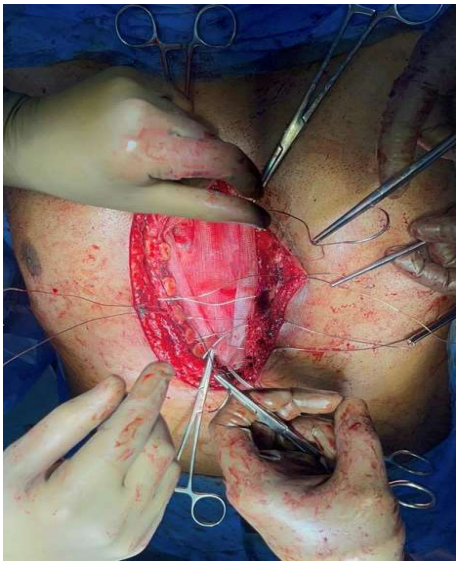
CECT thorax : solitary lesion over the body of sternum **(Fig. 3)**



PET CT : DOTA avid lesion over the body of sternum **(Fig. 4)**

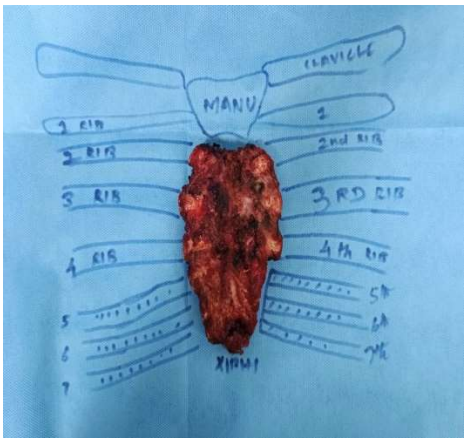


Intraoperative image **(Fig. 5a)**



Reconstruction using sandwich technique **(Fig. 5b)**

Reconstruction using sandwich technique **(Fig. 7b)**



Specimen image **(Fig. 5c)**

DISCUSSION

Paragangliomas are derived from neural crest cells. They can arise from adrenal medulla

(phaeochromocytoma) -80%. Extra-adrenal paragangliomas arise from chromaffin tissue along the ANS - found in the head, neck, thorax, abdomen and pelvis. The most common site of paragangliomas is in the abdomen is at the aortic bifurcation.

May be related to hereditary syndromes such as VHL gene mutations, MEN2, NF1, Carney triad , SDH gene mutations. [10,11].

Patients with functional paragangliomas can have hypertension, flushing, tachycardia, palpitations, anxiety due to catecholamine excess [4,5,12]. Those with non-functioning retroperitoneal paragangliomas may be diagnosed incidentally or have vague abdominal pain that may be associated with nausea, vomiting, abdominal distension and weight loss [4,13]. The varied range of presentations often makes the clinical diagnosis of these tumors

difficult.

Ultrasound is the first-line investigation however CT and MRI have higher sensitivity. MIBG scintigraphy/ DOTANOC PET may be used for better localisation of extraadrenal disease or metastatic sites. Ultimately although imaging techniques are helpful, the diagnosis of paragangliomas can only be confirmed with careful HPE and IHC examination. [17,18].

Due to risk of malignancy, surgery is the preferred management. Resection is challenging as these are highly vascular tumours and are frequently located near vital blood vessels. If a tumor is unresectable , chemo/ radiation / embolization can be tried initially [19]. Radiotherapy may also be used for inoperable tumours or for palliation. Inoperable paragangliomas can be treated with octreotide [20].

Malignancy cannot be diagnosed histologically and is diagnosed based on presence of metastasis. Approximately 20–40% of extra-adrenal sympathetic paragangliomas metastasise, compared to only 2–10% of phaeochromocytomas.

Solitary metastasis to the sternum is unheard of, and not reported in literature.

Metastatic lesions have a poor prognosis, with a reported 5-year survival rate of 36%.

CONCLUSION

Paragangliomas of the retroperitoneum are rare with malignant potential that are difficult to both diagnose and treat.

Advances in genetic testing help in the increased understanding of paragangliomas, however at present there is no way to definitively predict metastatic risk.

Since recurrence and metastasis are common, lifelong follow-up is required.

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