ORIGINAL RESEARCH PAPER
Volume - 14 | Issue - 01 | January - 2025 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

A RARE CASE OF EXTRA-ADRENAL RETROPERITONEAL NON-FUNCTIONAL PARAGANGLIOMA: DIAGNOSTIC CHALLENGES AND CLINICAL INSIGHTS

Radiology

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ABSTRACT

Paragangliomas (PGLs) are rare neuroendocrine tumors arising from extra-adrenal chromaffin cells, with an incidence of approximately 3 cases per million annually. These tumors can be sympathetic (functional) or parasympathetic (non-functional), with retroperitoneal paragangliomas being particularly rare and diagnostically challenging due to nonspecific symptoms. We report a 24-year-old male presenting with acute abdominal pain unresponsive to analgesics. Imaging identified two metabolically active retroperitoneal masses with central necrosis and vascular involvement, diagnosed as paragangliomas through histopathology. PET-CT revealed hypermetabolic activity, suggesting metastatic potential. Given the inoperable nature of the tumors, the patient received 177Lu-DOTATATE peptide receptor radionuclide therapy (PRRT), a novel targeted treatment for neuroendocrine tumors. This case highlights the diagnostic and therapeutic challenges of non-functional retroperitoneal paragangliomas. Advanced imaging techniques and histopathological confirmation were crucial in diagnosis. While surgical resection remains the gold standard for localized tumors, PRRT is emerging as a viable option for metastatic or unresectable cases, as supported by similar reports in the literature.

KEYWORDS

Paraganglioma, Extra-adrenal chromaffin cells, Hyper metabolic activity, Neuroendocrine tumors, PRRT.

INTRODUCTION

Paragangliomas (PGLs) are rare neuroendocrine tumors that arise from extra-adrenal chromaffin cells. Paragangliomas can be categorized as either sympathetic or parasympathetic. Sympathetic paragangliomas, such as pheochromocytomas, are typically functional and symptomatic and parasympathetic paragangliomas are generally non-functional and tend to manifest through mass effect [1]. The estimated incidence is approximately 3 cases per million individuals, with a marked increase observed in recent years [2,3]. Paragangliomas originating from the adrenal medulla are referred to as pheochromocytomas, whereas those occurring outside the adrenal gland are broadly categorized as extra-adrenal paragangliomas [4]. The intra-abdominal region, particularly the para-aortic and peri-nephric areas, represents the most frequent site for extra-adrenal paragangliomas [5].

Here, in this case report we presenting a case of Extra-Adrenal Retroperitoneal Non-Functional Paraganglioma.

CASE STUDY

A 24-year-old male presented with a two-day history of ill-defined abdominal pain, unresponsive to painkillers. Initial evaluations, including ultrasound, revealed a large heterogeneous mass in the posterior aspect of the portal vein with extensive vascular involvement. His vitals on presentation were stable, with a pulse rate of 78 bpm, BP 110/76 mmHg, SpO2 98%, and temperature 98.2°F. The patient had no prior medical or significant family history.

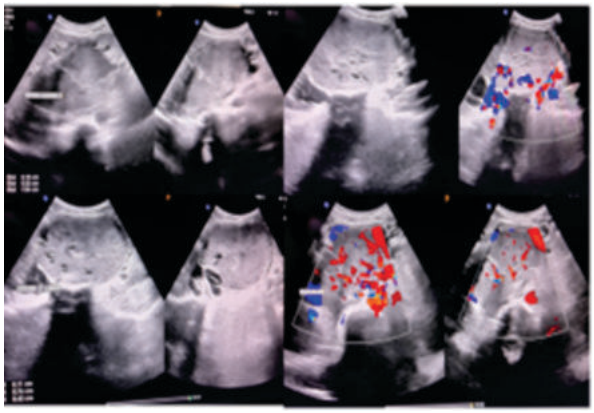

Figure 1: USG Abdomen- Liver, gall bladder, spleen (9.1 cm), kidneys, and prostate are normal. Heterogeneous masses (97x71x103 mm near portal vein, 83x79x104 mm at aorta bifurcation) with necrosis, vascularity, and structural displacement noted. Tortuous epigastric vessels suggest neoplastic etiology.

Ultrasound demonstrated a mass near the portal vein and another similar lesion near the aortic bifurcation, suggestive of neoplastic etiology. The mass measured approximately 10.6 x 8.0 x 8.9 cm, with intense heterogeneous enhancement in the arterial phase and areas of necrosis. Multiple vascular channels surrounding the mass in the periportal and peripancreatic regions suggested collateral formation (Figure 1).

CECT Abdominal Angiography showed Numerous dilated vascular channels around the periportal and pelvic regions were noted, with collateral channels extending into the right paravertebral region and joining the IVC (Figure 2).

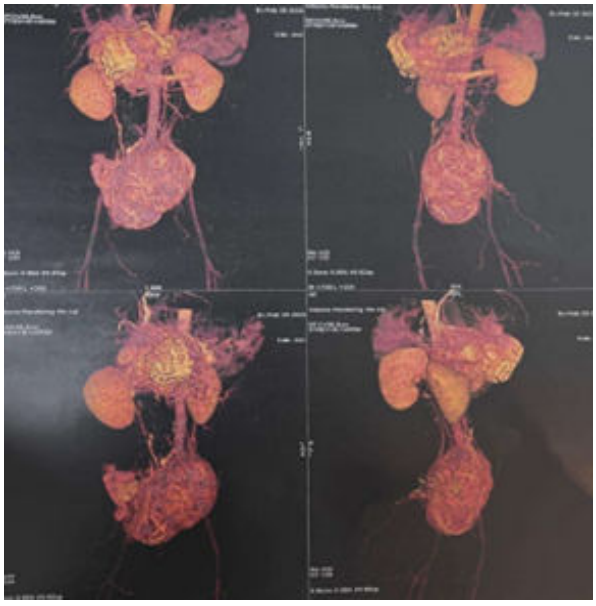

Figure 2: CT ANGIOGRAPHY 3-D - Multiple dilated contrast filled vascular channels are seen surrounding the mass lesion.

18FDG PET-CT Scan showed Two metabolically active masses with central necrosis were seen, with hypermetabolic lytic-sclerotic

skeletal metastases indicating metastasis. The differential diagnosis initially included leiomyosarcomas versus retroperitoneal sarcomas.

A USG-guided biopsy confirmed the diagnosis of paraganglioma, with histology consistent with neuroendocrine tumor characteristics. The patient was referred to the Nuclear Medicine department for 177Lu-DOTATATE therapy, a targeted radionuclide therapy for neuroendocrine tumors. The 177Lu-DOTATATE therapy was started and continues follow-up will be done.

DISCUSSION

Paragangliomas (PGLs), particularly extra-adrenal retroperitoneal types, present a diagnostic challenge due to their rarity and nonspecific symptoms. This case of a 24-year-old male presenting with acute abdominal pain emphasizes the diagnostic and therapeutic complexities associated with these tumors. The diagnostic pathway for PGLs relies heavily on imaging modalities and histopathology. The CT findings of a large, hyper vascular mass with areas of necrosis were consistent with known imaging features of retroperitoneal paragangliomas. PET-CT further confirmed metabolic activity consistent with PGL, highlighting the importance of functional imaging in detecting neuroendocrine activity and metastases. Similar cases in the literature illustrate the diagnostic challenges and diverse presentations of paragangliomas. For instance, Brahmbhatt et al. described a retroperitoneal paraganglioma presenting with chest pain and abdominal pain, underscoring the variability in clinical presentation [4]. Chattoraj et al. reported a non-functional retroperitoneal paraganglioma initially misdiagnosed as a GIST, which aligns with the diagnostic challenges faced in the present case [6]. Surgical excision remains the gold standard for localized PGLs. However, in metastatic or inoperable cases, peptide receptor radionuclide therapy (PRRT) with agents like 177Lu-DOTATATE offers a promising alternative [7].

CONCLUSION

Non-functional retroperitoneal paragangliomas are rare group of tumors which are difficult to diagnose, owing to their asymptomatic behaviour. This case highlights the importance of a multidisciplinary approach in managing paragangliomas, from diagnosis to treatment. The integration of advanced imaging, histopathological confirmation, and targeted radionuclide therapies offers hope for improved outcomes in these complex cases. Future research focusing on the genetic and molecular aspects of paragangliomas could provide further insights into tailored management strategies.

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