# Primary Hepatic Neuroendocrine Tumour: A Rare Entity

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## ABSTRACT

Hepatic neuroendocrine tumours (NETs) are of infrequent occurrence and account for less than 0.3% of all NETs. The authors hereby describe the case of a young female, who presented with abdominal pain, fever, and weight loss. CT scan abdomen showed a large tumour in the liver, with areas of necrosis and calcification. The serum tumour markers were normal. A liver lesion biopsy along with a dotatate scan helped establish the diagnosis. Since the lesion involved the nearby vessels, the patient was advised management as per palliative care.

**Key Words:** *Neuroendocrine tumour, Liver, Palliative care.*

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## INTRODUCTION

Neuroendocrine tumours (NETs) arise from the neuroendocrine cells of neural crest origin. Common sites of origin include the appendix, small intestine, rectum and bronchi.[1] Primary hepatic neuroendocrine tumour (PHNET) is a rare entity, representing about 0.3% of all NETs.[2] These are characterised by the presence of neurosecretory granules and stain positive for specific markers including neuron-specific enolase (NSE), synaptophysin, and chromogranin. Histological confirmation of a PHNET is required to make a primary diagnosis and to rule out other primary hepatic tumours or metastatic NETs.

## CASE REPORT

A 24-year female with no prior comorbids, presented with the complaint of right upper quadrant pain, low-grade fever, and undocumented unintentional weight loss for 1 month after a Caesarean section. Patient denied any history of jaundice, nausea or vomiting. Her baseline laboratory workup including complete blood count, and renal and liver function tests were normal with viral markers, e.g. Hepatitis B surface antigen and Hepatitis C antibody being non-reactive. For the evaluation of her symptoms, she was advised abdominal ultrasound, which revealed a large cystic-cum-solid lesion with vascularity. The serum tumour markers, including carcinoembryonic antigen (CEA) and Alpha-fetoprotein (AFP), were reported to be in the normal range.

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For further characterisation of the liver lesion, she underwent computed tomography of the chest, abdomen, and pelvis with contrast (CT CAP with contrast) which showed a large well-defined heterogeneous lesion involving the right lobe of the liver with predominant involvement of segment V, VII, and VIII having internal necrotic component and calcifications (Figure 1). It measured 14.3 x 15.1 x 23.0 cm (AP×TR×CC) in dimensions. Extensive peritumoural vascularity was also evident. It was perceived to be crossing the midline and displacing the bowel loops and mesenteric vessels towards the left side. Inferiorly, the lesion was abutting the right kidney with indistinct fat planes and also compressing the head and body of the pancreas with indistinct fat planes. Posteriorly, it was compressing the inferior vena cava (IVC) and was also seen adjacent to the right renal artery and bilateral renal veins with no evidence of thrombus formation.

The case was discussed in the tumour board meeting and a liver lesion biopsy was planned. The liver lesion tru-cut biopsy showed a neoplastic lesion comprising of a monotonous population of large-sized cells with abundant, amphophilic cytoplasm (Figure 2A). Immunohistochemical (IHC) markers were done and showed synaptophysin and chromogranin positivity, while Hep Par- 1 and CK19 were negative (Figure 2B & C). Ki-67 was seen positive in approximately <2% of tumour cells (Figure 2C). The morphological and IHC features were suggestive of well-differentiated NET (large cell type) of WHO Grade I.

On dotatate scan, a DOTA-avid bulky mass with central photopenia and specks of calcification was seen infiltrating the right lobe of liver, obliterating the pancreas and adjacent bowel. There were no DOTA-avid lesions in the spleen, lungs, adrenal gland, or lymph nodes, suggestive of no distant metastasis.

Considering the extensive vascular involvement of the lesion, the patient was labelled to have an unresectable disease and was advised for palliative care.

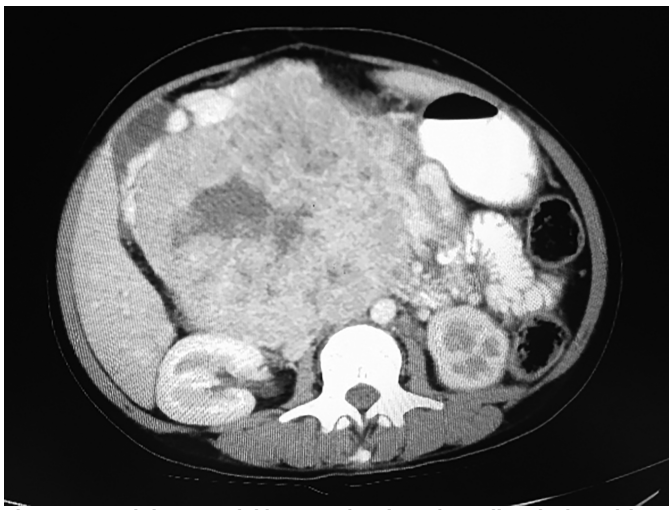

**Figure 1: CT abdomen axial images showing a large liver lesion with an internal necrotic component.**

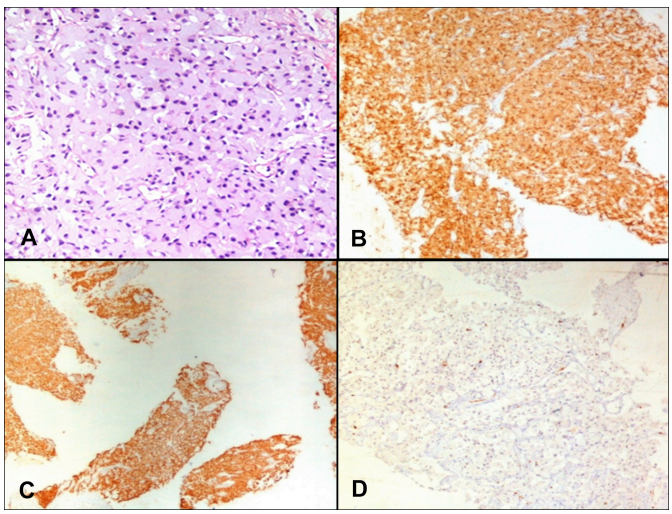


**Figure 2: (A) Monotonous population of well-differentiated, large sized neuroendocrine cells with amphophilic cytoplasm. (B and C) The synaptophysin and chromogranin immunohistochemical stains highlight the neuroendocrine cells. (D) Ki-67 is seen to be positive in approximately <2% of tumour cells.**

## DISCUSSION

NETs of primary hepatic origin are of rare occurrence; consequently, only a handful of studies have been done so far regarding their clinical manifestations, staging, treatment options, and outcomes.[3] The liver is a common site of metastasis for NETs; hence, it is important to distinguish between primary and metastatic liver disease. These tumours are rare in those under 40 years of age[4] and can either be functional or non functional.[5]

In general, NETs are classified by their grade and differentiation, which are essential prognostic markers. Grade refers to the proliferative activity of the tumour, measured by mitotic rate and Ki-67 index. Differentiation denotes the extent to which tumour cell morphology resembles endocrine cells of origin.[6] NETs are slow-growing tumours except for poorly differentiated ones which are highly aggressive tumours.

There is no specific treatment strategy for PHNETs. Currently, surgical resection is considered the most appropriate treatment modality for PHNET.[7] For patients with restricted but unresectable liver tumours, transplantation has appeared to be the best therapeutic alternative.[8] On the other hand, trans-arterial chemoembolisation (TACE) can be considered for advanced PHNETs that are poor candidates for resection.[8] Further studies are required to establish a definitive treatment strategy.

NETs can manifest as primary hepatic tumours; one must distinguish these from metastatic liver disease. Being a rare entity along with the scarcity of data, less is known regarding its staging, treatment options, and clinical outcomes. Since no guidelines or consensus statement exist for it,[9] surgical resection appears to be the most appropriate treatment option available.

In conclusion, PHNET is a very rare tumour entity. Its accurate diagnosis and management require a multidisciplinary team approach.

**PATIENT'S CONSENT:**

Informed consent was obtained from the patient for this publication.

**COMPETING INTEREST:**

There is no competing interest declared by the authors.

**AUTHORS' CONTRIBUTION:**

NR: Managed the patient and wrote the initial manuscript.
ZM: Wrote the initial and final manuscript.
KB: Managed the patient.
AA: Wrote the final draft.
NHL: Conceptualised and oversaw the management.
All authors approved the final version of the manuscript to be published.

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