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Primary Neuroendocrine Tumor of the Liver in a Cirrhotic Patient: A Case Report

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Abstract **Case Report**

Primary hepatic neuroendocrine tumors (PHNETS) are rare entities, often misdiagnosed as hepatocellular carcinoma (HCC), especially in patients underlying cirrhosis. We report a case of A 63-year-old male follow-up for liver cirrhosis, who presented with incidentally detected 2 livers masses on abdominal computed tomography. Imaging studies suggested HCC. However, histopathological analyses confirmed the diagnosis of a well differentiated grade 1 neuroendocrine tumor. This case highlights the importance of considering PHNET in the differential diagnosis of hepatic tumor in cirrhotic patient.

Keywords: Primary hepatic neuroendocrine tumors (PHNETs), Liver cirrhosis, Hepatocellular carcinoma (HCC), Neuroendocrine tumors (NETs), Computed Tomography/CT).

INTRODUCTION

Neuroendocrine tumors (NETs) are uncommon growths that develop from cells of the neuroendocrine system. They primarily occur in the gastrointestinal and respiratory tracts [1].

NETs represent approximately 1% to 2% of all gastrointestinal tumors. However, primary hepatic neuroendocrine tumors (PHNETs), first described by Edmondson in 1958 [2], are exceptionally rare and grow slowly, which often makes them difficult to identify until the disease has advanced to a later stage [3-5]. Diagnosing PHNETs in cirrhotic patient is particularly challenging due to their resamblance to hepatocellular carcinoma on imaging.

Given the rarity of PHNETs, there are no established algorithms for the diagnosis and management of these lesions, which presents a real challenge. Here we report a case of cirrhotic patient presenting with a hepatic mass, ultimately diagnosed as PHNET.

CASE REPORT

A 63-year-old male follow-up for liver cirrhosis of unknown etiology. clinically, the patient was asymptomatic. Laboratory investigations revealed pancytopenia with GB: 900/µL, Hb: 7,9 g/dl, thrombocytopenia at 32000/µL. TP: 27%, Alpha-fetoprotein (AFP) was normal. Serum liver function tests were normal. Imaging studies, including abdominal ultrasound and contrast-enhanced computed tomography (CT), revealed A dysmorphic liver with 2 poorly circumscribed heterogeneous hypodense lesions with peripheral reaggregation at arterial time with lavage at portal time measuring 49x39mm and 58x40mm and a dialated portal trunk measuring 20 mm with marginal endoluminal thrombosis in the retro pancreatic area and at the level of the 2 left and right portal branches with splenomegaly at 24 cm.

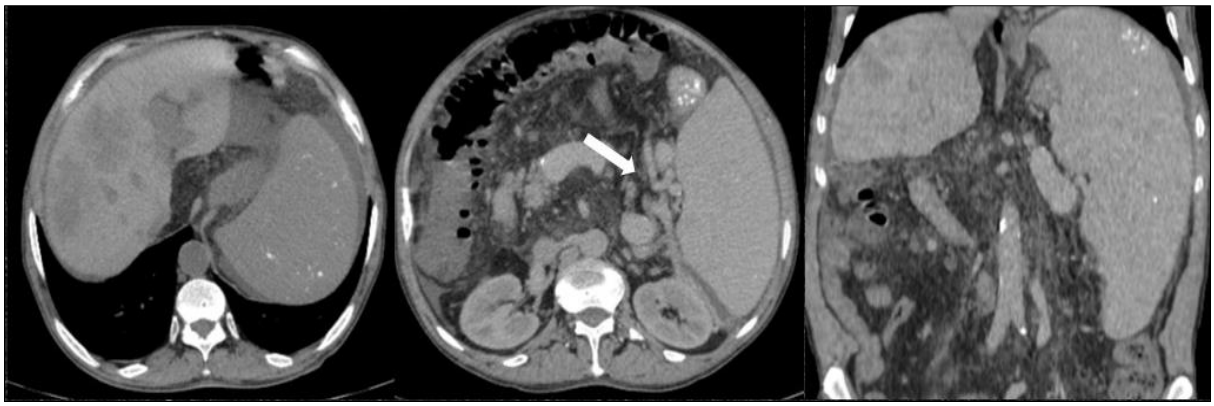

Figure 1: Portal phase CT showing features of portal hypertension, including a dysmorphic liver with irregular nodular contours, significant splenomegaly, and prominent perisplenic collateral venous circulationand associated ascites

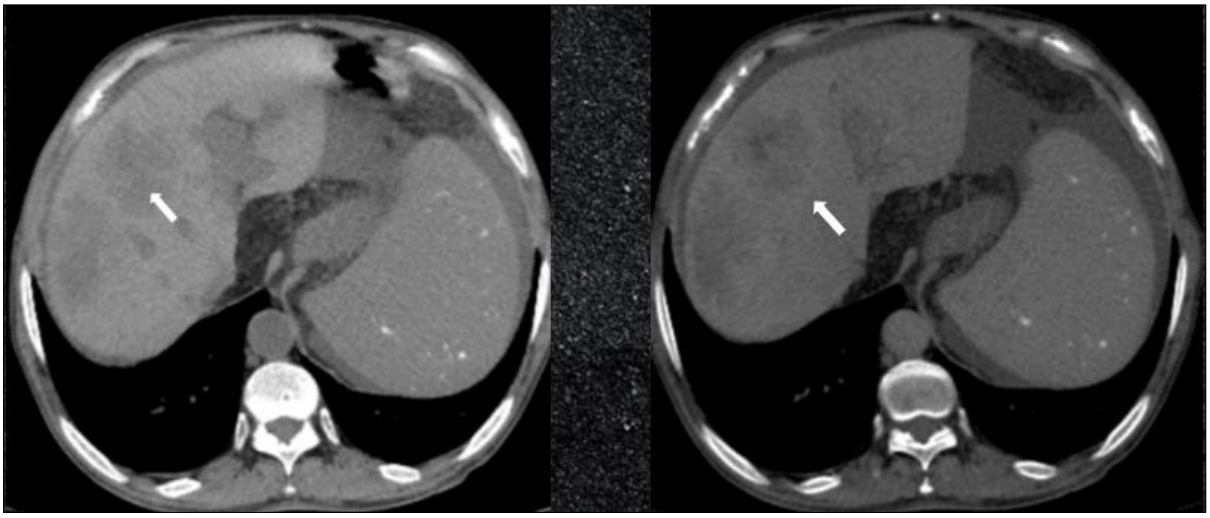

Figure 2: Axial abdominal CT scan showing two irregular, heterogeneous, hypodense lesions in segment WIII of the liver, with peripheral arterial phase enhancement and washout in the portal venous phase

A liver biopsy was performed due to the absence of AFP elevation, and histopathological analysis revealed well-differentiated neuroendocrine tumor cells with positive immunohistochemistry for synaptophysin and chromogranin A, confirming the diagnosis of a primary hepatic neuroendocrine tumor. The ki-67 index was 15%, confirming a grade GII NET.

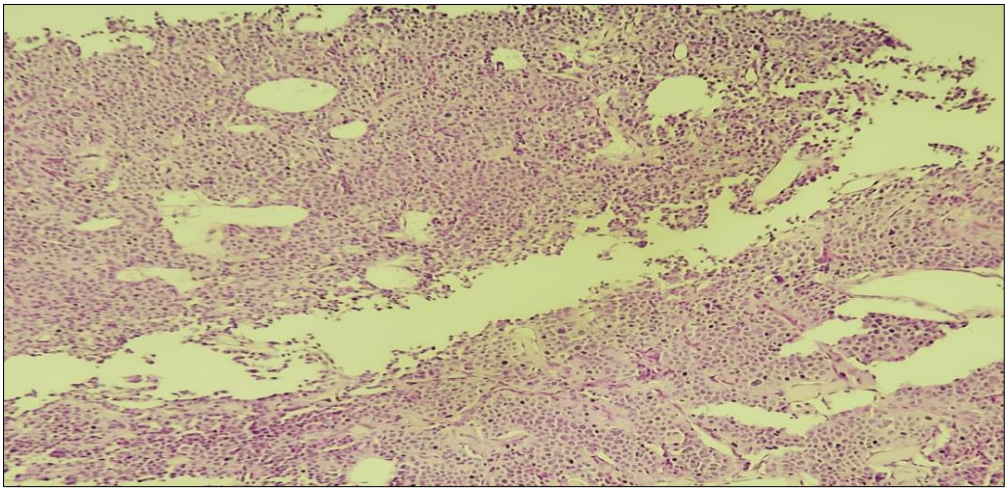

Figure 3: Neuroendocrine tumor showing trabecular and organoid architecture. No necrosis was observed. (Eosin-hemalun x20)

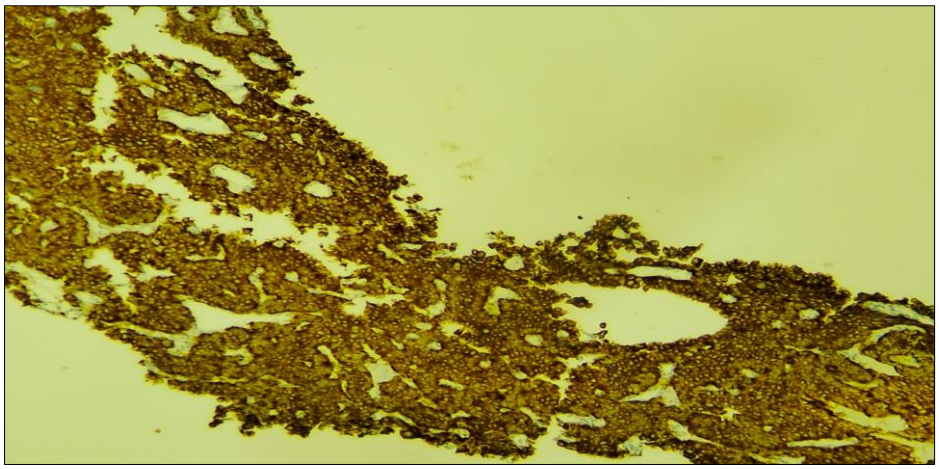

Figure 4: The diffuse expression of synaptophysin supports its neuroendocrine differentiation

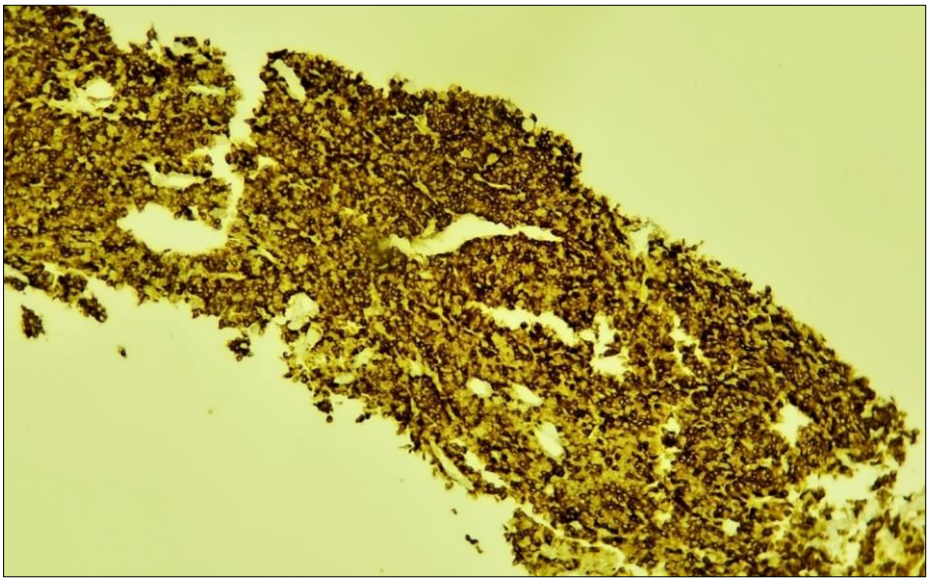

Figure 5: Diffuse chromogranin expression

Given the patient's cirrhosis and the tumors well differentiated nature, a multidisciplinary approach was taken. Surgical resection was deemed high risk due to liver dysfunction (child-pugh B7). He was started on somatostation analog therapy. At 12-month follo up, the tumor remained stable, and the patient had no significant symptoms.

DISCUSSION

NETs arise from neuroectodermal cells that migrate from the neural crest to different parts of the body during embryogenesis [6]. They originate from enterochromaffin cells that have the ability to secrete functional hormones [7].

However, these cells do not migrate to the liver on a regular basis, which explains the rarity of PHNET [8, 9]. On the other hand, the liver is usually a site of metastases of digestive digestive neuroendocrine tumours. PHNETs differ from other NETs because they do not secrete biologically active polypeptides or amines, which leads to the absence of carcinoid syndrome.

PHNET is a rare disease, and there is limited guidance available for its diagnosis and management. In 2011, Quartey [10], reviewed 124 cases of PHNET, with a mean age of 51.9 years and a slight female predominance. This demographic trend was confirmed by another review in 2009 (11). We report a case of PHNET in cirrhotic patient with atypical characteristics, including being male, elderly. These clinical features would likely suggest Hepatocellular Carcinoma HCC as a probable diagnosis. The coexistence of a hepatic NET with cirrhosis is an uncommon condition that pose diagnostic and therapeutic challenges. This case highlights the diagnostic challenges associated with PHNET.

These tumors grow slowly and generally become clinically noticeable only at later stages. Clinical signs include weight loss, fatigue, abdominal distension, pain, and a palpable mass in the right upper quadrant [12, 13]. Hepatic lesions are often detected during physical examinations in the absence of symptoms. In the case

reported in this article, the lesions were identified during the patient's follow-up.

Serum tumor markers like alpha-fetoprotein (AFP) and carcinoembryonic antigen (CEA) are not useful for the diagnosis of PHNETs [15].

On imaging, PHNET is frequently mistaken for HCC or cholangiocarcinoma (CCC). PHNET lesions are atypical, appearing as masses that can be nodular or cystic, and may be either well-defined or poorly defined.

Imaging methods such as ultrasound, CT scans, and MRI are essential for diagnosing liver masses. PHNETs generally have a rich blood supply but do not present distinct radiological features that differentiate them from other liver tumors, often leading to misdiagnosis as hepatocellular carcinoma [14, 15]. CT imaging in PHNETs, generally present as low-density masses that show prominent enhancement during the arterial phase and decreased enhancement in the portal or delayed phases. MRI of PHNETs usually reveals ring-like enhancement in the arterial phase, with sustained enhancement in the portal or delayed phases, and a high signal intensity on the DWI sequence [16]. The most effective radiological exam for detecting neuroendocrine tumors is Octreoscan, assuming the tumor expresses somatostatin receptor subtype 2, with a sensitivity ranging from 75% to 95%.

Histopathological examination is one of the most accurate methods for diagnosing PHNETs. A biopsy can be obtained either through fine needle aspiration or surgically after mass resection [17]. Neuron-specific enolase, chromogranin A and synaptophysin are generally accepted as highly sensitive immunohistochemical markers for the diagnosis of NETs. The tumor in our case was immunoreactive for synaptophysin and chromogranin A.

Due to the rarity of PHNETs, there are no established treatment guidelines at present. However, surgical resection, particularly anatomical liver resection, is generally regarded as the preferred treatment option. This approach offers the potential for a complete cure, with a reported 5-year survival rate of 78% [18]. When primary lesions are considered unresectable, alternative treatments, including arterial embolization, chemotherapy, radiotherapy, somatostatin analogs, conservative management, and liver transplantation, may be considered once all other options have been explored.

Conclusion

This case emphasizes the importance of considering PHNET in the differential diagnosis of liver masses in cirrhotic patients. Primary neuroendocrine tumors of the liver are rare, and their diagnosis in a cirrhotic patient presents unique challenges. They are typically asymptomatic, making them difficult to differentiate from other liver tumors, such as HCC and cholangiocarcinoma, based on medical imaging findings. The diagnosis of PHNET is determined by pathological characteristics and treatment strategies must be individualized, taking into account the patient’s liver function and tumor characteristics.

References

1. Linh LT, Minh NM, Minh HT, et al.: Primary hepatic neuroendocrine tumor. Endocrinol Diabetes Metab Case Rep. 2021, 2021:10.1530/EDM-20-0220
2. *Edmondson HA. Tumor of the liver and intrahepatic bile duct. Atlas of tumor pathology. 1958;7:105–109.*
3. [1] Zhao ZM, Wang J, Ugwuowo UC, et al. Primary hepatic neuroendocrine carcinoma: report of two cases and literature review. BMC Clin Pathol 2018;18:3.
4. Modlin IM, Sandor A. An analysis of 8305 cases of carcinoid tumors. Cancer 1997;79:813–29.
5. *Tuan Linh L, Minh Duc N, Tu Minh H, Ngoc Cuong N, Thu Ha V, Luan DT, et al. Primary hepatic neuroendocrine tumor. Endocrinol Diabetes Metab Case Rep (2021) 2021:20-0220. doi: 10.1530/EDM-20-0220*
6. K. Mima, T. Bepppu, A. Murata, *et al.*Primary neuroendocrine tumor in the liver treated by hepatectomy: report of a case Surg. Today, 41 (2011), pp. 165 5-1660
7. L. M Fenoglio, S. Severini, D. Ferrigno, *et al.* Primary hepatic carcinoid: a case report and literature reviewWorld J. Gastroenterol., 15 (19) (May 21, 2009), pp. 2418-2422
8. K.Mima, T. Bepppu, A. Murata et al. Primary neuroendocrine tumor in the liver treated by hepatectomy: Report of a case. Surg today (2011) 41 :165 5 -1660.
9. I.M.Lambrescu IM, S. Martin, L. Cima et al. Primary hepatic neuroendocrine tumor after 4 years tumor - free follow -up. J Gastrointestin Liver Dis. 2015 Jun ;24(2) :241 - 4
10. Quartey B. Primary hepatic neuroendocrine tumor: what do we know now?. World J Oncol. 2011;2(5):209–16.
11. Lin CW, Lai CH, Hsu CC, Hsu CT, Hsieh PM, Hung KC, et al. Primary hepatic carcinoid tumor: a case report and review of the literature. Cases J. 2009;2(1):90.
12. J.E. Song, B.S. Kim, C.H. Lee, Primary hepatic neuroendocrine tumor: a case report and literature review, World J. Clin. Cases 4 (8) (2016) 243–247.
13. C. Shi, Q. Zhao, B. Dai, F. Xie, J. Yang, Primary hepatic neuroendocrine neoplasm: long-time surgical outcome and prognosis, Medicine (Baltimore) 97 (31) (2018), e11764.
14. Primary hepatic neuroendocrine tumour with multiple liver metastases: a case report with literature review. Yu WM, Li R, Sun BL, Du JK,

Tuo HF. Int J Surg Case Rep. 2021;89 doi: 10.1016/j.ijscr.2021.106590.
15. The outcome of primary hepatic neuroendocrine tumors: a single-center experience. Shah D, Mandot A, Cerejo C, Amarapurkar D, Pal A. J Clin Exp Hepatol. 2019;9:710–715. doi: 10.1016/j.jceh.2019.08.002.
16. Li RK, Zhao J, Rao SX, Chen CZ, Zeng MS, Qiang JW. Primary hepatic neuroendocrine carcinoma: MR imaging findings including preliminary observation on diffusion-weighted imaging. Abdom Imaging (2013) 38(6):1269–76. doi: 10.1007/s00261-013-0029-1
17. Primary hepatic neuroendocrine tumor with multiple liver metastases: a case report with review of the literature. Yang K, Cheng YS, Yang JJ, Jiang X, Guo JX. World J Gastroenterol. 2015;21:3132–3138. doi: 10.3748/wjg.v21.i10.3132
18. Li YF, Zhang QQ, Wang WL. Clinicopathological characteristics and survival outcomes of primary hepatic neuroendocrine tumor: A surveillance, epidemiology, and end results (SEER) population-based study. Med Sci Monit (2020) 26:e923375. doi: 10.12659/MSM.923375.