A Rare Case of Primary Hepatic Neuroendocrine Tumor

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INTRODUCTION

Neuroendocrine tumors (NETs) are a heterogeneous group of neoplasms, most commonly originating in the gastrointestinal tract or lungs.

Hepatic involvement typically reflects metastatic disease; hence, primary hepatic NETs are exceptionally rare and pose a significant diagnostic challenge. With less than 200 cases reported in the English literature, they account for approximately **0.3 – 4% of all NET cases**, highlighting their scarcity.

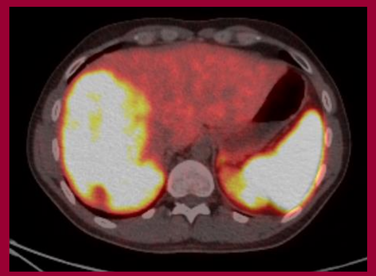

Fig. 1 PET-Ga before PRRT & Somatostatin analogues

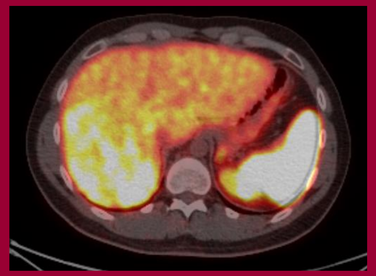

Fig. 2 PET-Ga after PRRT & Somatostatin analogues

CASE PRESENTATION

A 32-year-old man was admitted to our Department due to a solitary hepatic mass, found coincidentally on ultrasound. Because of initial suspicion of hepatocellular carcinoma, the patient underwent biopsy following MRI (Fig. 3, 4), which revealed features consistent with well – differentiated neuroendocrine tumor grade 2, corroborated by immunohistochemical staining.

Comprehensive workup failed to identify an extrahepatic primary source, supporting the diagnosis of a primary hepatic NET. He received peptide receptor radionuclide therapy followed by somatostatin analogues (Fig.1). Subsequent 68Ga-PET indicated a regression of the lesion, with a reduction in size and a diminished degree of radiopharmaceutical intake (Fig.2).

A right hepatectomy was carried out and histopathological evaluation confirmed the initial diagnosis (Fig.5, 6). The patient was discharged following an uncomplicated recovery.

DISCUSSION

Primary Hepatic Neuroendocrine Tumor (PHNET) is an exceedingly uncommon malignant tumor, comprising **under 1% of primary liver malignancies**. Originating most likely from neuroendocrine cells within the intrahepatic bile duct epithelium, PHNET is fundamentally a **diagnosis of exclusion**, requiring a thorough investigation to rule out a metastatic source from the more common primary sites. It typically affects middle-aged patients (mean age 47–50 years) and shows no strong sex predilection, though some studies report a slight female predominance. Due to its nonfunctional nature and slow growth, it often presents asymptomatically or at an advanced stage with non-specific symptoms like abdominal pain.

The clinical challenge in managing PHNET lies primarily in its preoperative diagnosis. The tumor's imaging characteristics (CT/MR) are highly variable and **frequently mimic Hepatocellular Carcinoma** (HCC) due to similar hypervascularity and enhancement patterns, complicating differentiation. While standard imaging lacks specificity, specialized techniques like Octreotide radionuclide imaging (**Ga68-DOTATATE PET-CT**) are crucial, leveraging the tumor's expression of somatostatin receptors to achieve high sensitivity (up to 95%). **Definitive diagnosis relies on post-surgical histopathology and immunohistochemistry** (Synaptophysin, Chromogranin A, NSE).

Surgical resection (hepatectomy) is the treatment of choice, showing the highest long-term survival rates. The choice of surgical technique depends on the tumor's size and location, and achieving an R0 resection with negative margins, along with appropriate lymphadenectomy, is considered optimal. For unresectable disease, palliative strategies such as TACE, systemic chemotherapy, and somatostatin analogs are utilized, but their long-term efficacy is limited and largely extrapolated from data on other NETs.

CONCLUSION

The reported **5-year survival rate** after surgical resection ranges from **74% to 78%** , while the **recurrence rate is ∼ 18%.** Postoperative recurrence can occur many years later, necessitating long-term follow-up.

This case highlights the importance of considering neuroendocrine tumor in the differential diagnosis of liver masses, particularly in the absence of underlying liver disease. Due to the difficulty in achieving an accurate preoperative diagnosis, surgical resection remains the cornerstone of both diagnosis and treatment, offering favorable long-term survival rates for resectable disease.

Given the rarity of primary hepatic NETs, further studies are needed to elucidate their clinical behavior, optimal treatment strategies, and long-term outcomes.

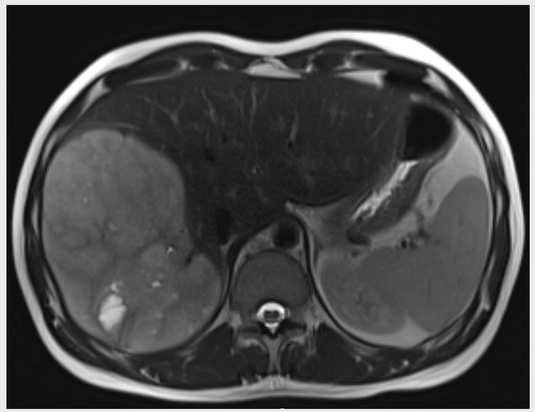

Fig. 3 Pre-operative MRI (T2)

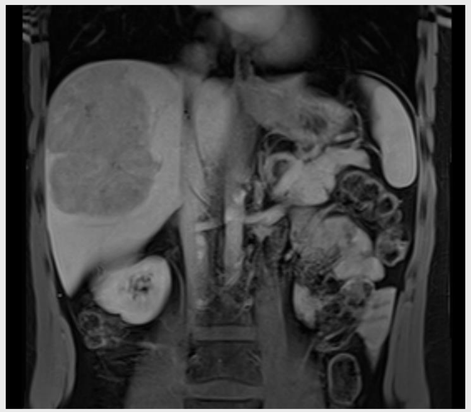

Fig. 4 Pre-operative MRI (T1)

Fig. 5 Right Hepatectomy

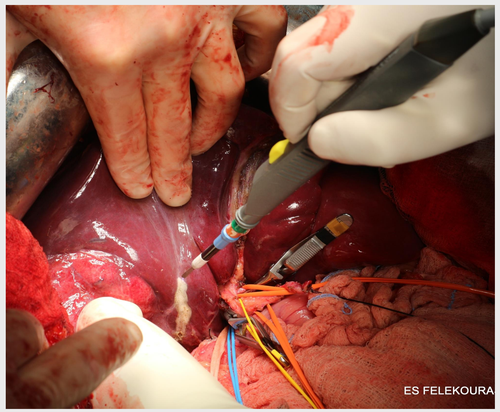

Fig. 6 Intra-operative resection and Pringle maneuver

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