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Case Report

Primary hepatic neuroendocrine tumor: A rare entity ☆

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ABSTRACT

Liver is the most common site for neuroendocrine metastasis. However, primary neuroendocrine tumor is a rare focal hepatic lesion with a better prognosis than hepatocellular carcinoma and other malignant hepatic lesions. We present a case of primary hepatic neuroendocrine tumor in a 38-year-old female patient with a radiological diagnosis of atypical focal hepatic lesion, and a confirmed diagnosis on histopathology. Few radiology features like predominant cystic component with absence of focal lesion at any other site, aids the imaging diagnosis of primary hepatic neuroendocrine tumor.

Case report

A 38-year-old female patient came to the emergency room with a history of abdominal pain, loss of weight and appetite. Vitals were normal at the time of the visit. An ultrasound abdomen was suggested, and it showed a heterogeneous lesion in the left lobe (Fig. 1); not seen separate from the gallbladder with cystic areas within and vascularity on color Doppler.

A triple phase contrast-enhanced computed tomography (CT) of the abdomen and pelvis was performed for further evaluation and characterization of the hepatic mass. It showed a predominant heterogeneous enhancement of the exophytic mass in the segment IVB inseparable from the medial wall of the gallbladder. Nonenhancing cystic/necrotic components were seen within the mass (Figs. 2–4).

No regional adenopathy or intrahepatic biliary radicle dilatation was seen. An associated bland/nonenhancing

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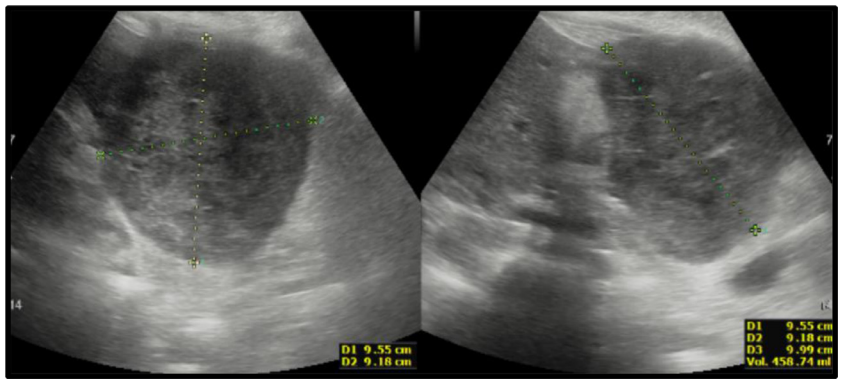

Fig. 1 – Ultrasound abdomen: heterogeneous mass in the left lobe.

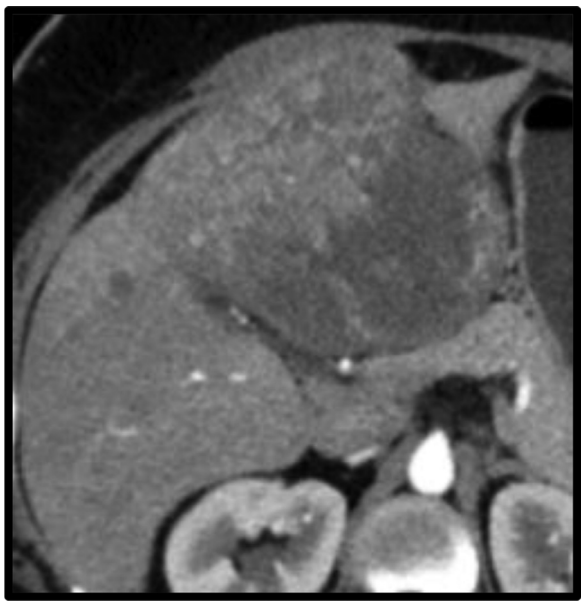

Fig. 2 – Arterial axial phase CECT: heterogeneously enhancing mass in the left lobe with nonenhancing regions.

thrombus was noted in the left portal vein and right anterior branch of the portal vein. No other focal enhancing lesions were seen elsewhere in the abdomen and pelvis sections.

Based on the CT features, neoplastic possibility of a primary gall bladder malignancy or neuroendocrine tumor was suggested. Additionally, nuclear medicine/ fluro deoxy glucose-Positron emission tomography scan showed predominant peripheral fluro deoxy glucose uptake with nonavid cystic/ necrotic areas (Fig. 5).

In view of absence of regional adenopathy and relatively younger age of the patient, a USG Guided biopsy of the mass was performed to confirm the diagnosis of primary hepatic neuroendocrine tumor.

Tumour markers like Serum Alpha feto protein, Serum Carcinoembryonic antigen, and CA19-9 levels were within normal limits.

Histopathology showed an infiltrating tumor in the form of cohesive sheets of small to medium sized blue round cells with perivascular pseudorosettes and areas of tumor necrosis (Fig. 7).

Immunohistochemistry was positive for synaptophysin, chromogranin, CD117 and CK with high Ki 67, proliferative Index of >90% consistent with high grade neuroendocrine tumor. The tumor cells were negative for CD-45 (Fig. 8). Subsequently, the patient underwent 3 cycles of adjuvant chemotherapy including Etoposide and Cisplatin.

A follow-up 3-month nuclear medicine scan showed good metabolic response with small residual hypodense mass in the segment IVB (Fig. 6). The residual mass was operated with tumor-free margins, and pathology showed predominant tumor infarcted tissue.

Discussion

Neuroendocrine tumor metastasis to the liver is very common, with some studies quoting ˜80% of the patients diagnosed to have liver metastasis at the time of diagnosis [1].

Primary neuroendocrine tumor of the liver: are very rare and account for ˜0.3% of the primary neuroendocrine tumor [2]. These are predominantly seen in the age group of 40-50 years, with no sex predilection [3]. These are slow growing and most of the times present late. Primary neuroendocrine tumor of the liver has a good prognosis, and a satisfactory 5-year survival rate of 74%-78% and 5-year recurrence rate of 18% after a hepatectomy [11,12]. Diagnosing primary neuroendocrine tumor of the liver is challenging, as the symptoms are unremarkable and the radiological findings are not very specific. Histopathology diagnosis addtionally requires Immunohistochemistry.

These tumors are thought to originate from the ectopic pancreatic or adrenal cells in the liver, neuroendocrine tissue in the intrahepatic biliary epithelium, or chronic inflammation in the biliary tract causing intestinal metaplasia [4].

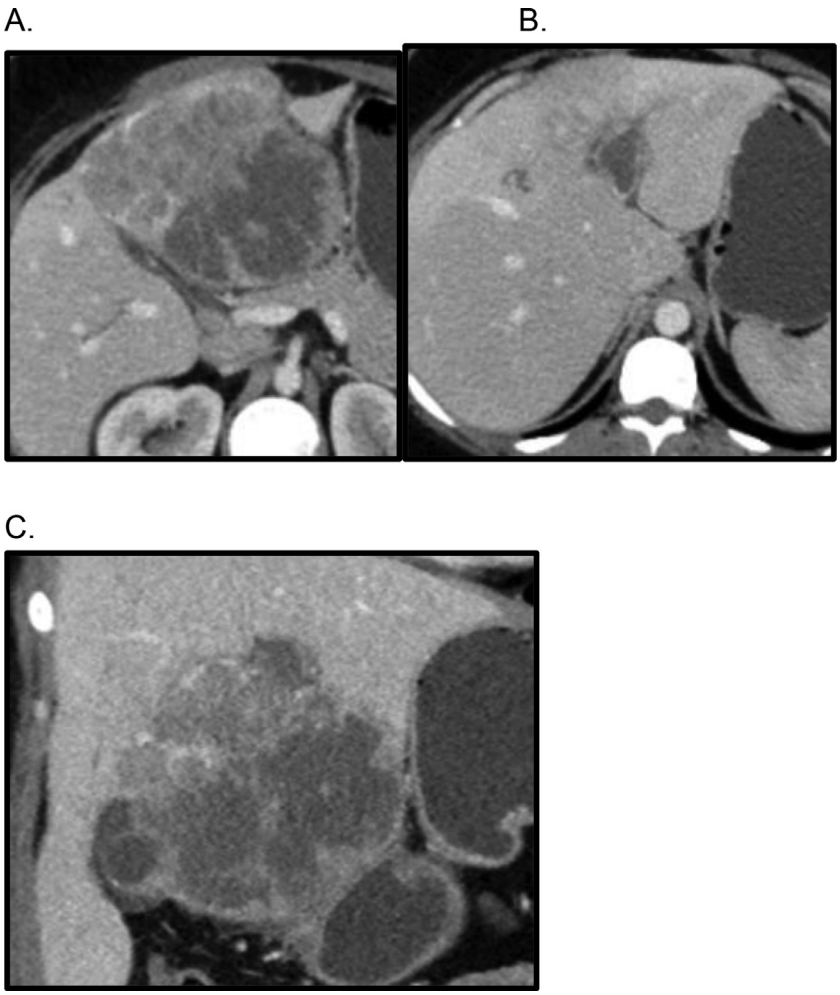

Fig. 3 – (A-C) CECT: portal phase shows enhancing solid areas with nonenhancing hypodense regions. Coronal image shows no fat plane between the mass and wall of the gall bladder.

Most commonly, the Neuroendocrine tumors are seen in the gastrointestinal tract, pancreas and bronchopulmonary tract [5]. Till date, about 150 cases have only been reported in literature [6].

Radiological appearances make it difficult to differentiate it from other liver lesions like HCC, cholangiocarcinoma and metastatic disease(when multiple). Most of these lesions have cystic areas/necrotic areas [7]. One of the diagnostic criteria for a primary hepatic neuroendocrine neoplasm is the absence of lesions at other sites commonly affected by this type of tumor, such as the small intestine, the pancreas, and the lungs.

Based on histological criteria, WHO grades the Neuroendocrine tumor into 3 Grades [8].

Grade 1 tumors are singular, solid nodules with enhancement on the arterial phase and washout on the venous phase on CT and magnetic resonance imaging scans. Grade 2 tumors can have a singular or multiple distribution pattern, necrosis, and nodule or marginal ring-like enhancements. Grade 3 tumors have multiple lesions, internal necrosis, and evidence of hemorrhage.

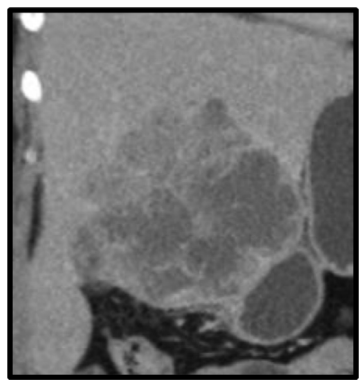

Fig. 4 – Delayed CECT coronal image shows washout with peripheral enhancement and hypodense to cystic areas.

Various therapeutic approaches may be attempted for Primary hepatic neuroendocrine Tumors (PHNETs), such as hepatic lobectomy, systemic chemotherapy, transhepatic arterial chemoembolization, radiofrequency ablation and liver transplantation [9,10].

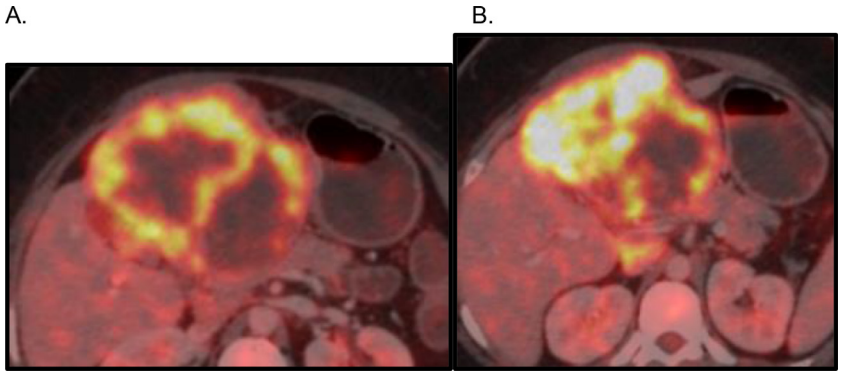

Fig. 5 – (A and B) PET CT images with hypermetabolic mass in the left lobe with FDG uptake in the solid component.

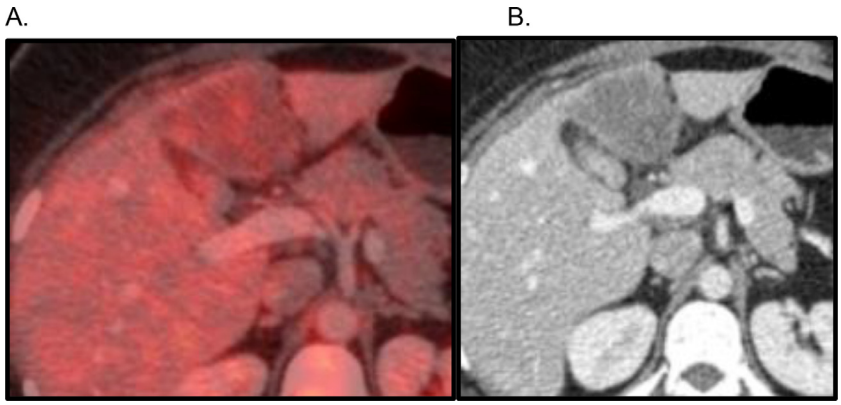

Fig. 6 – (A and B) Postchemotherapy follow-up PET images showing complete metabolic response and reduced size of the mass.

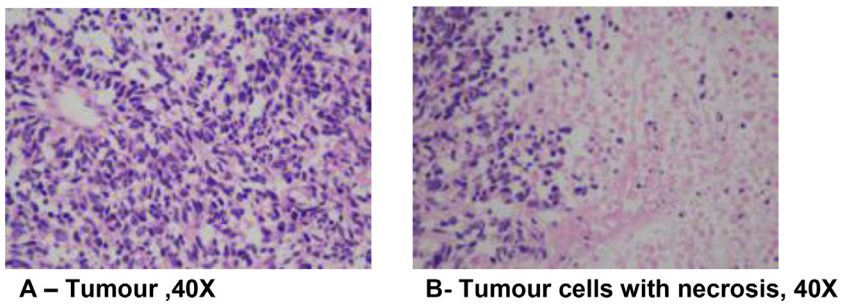

Fig. 7 – (A and B) Histopathology slides with 40x magnification showing tumor cells with necrosis.

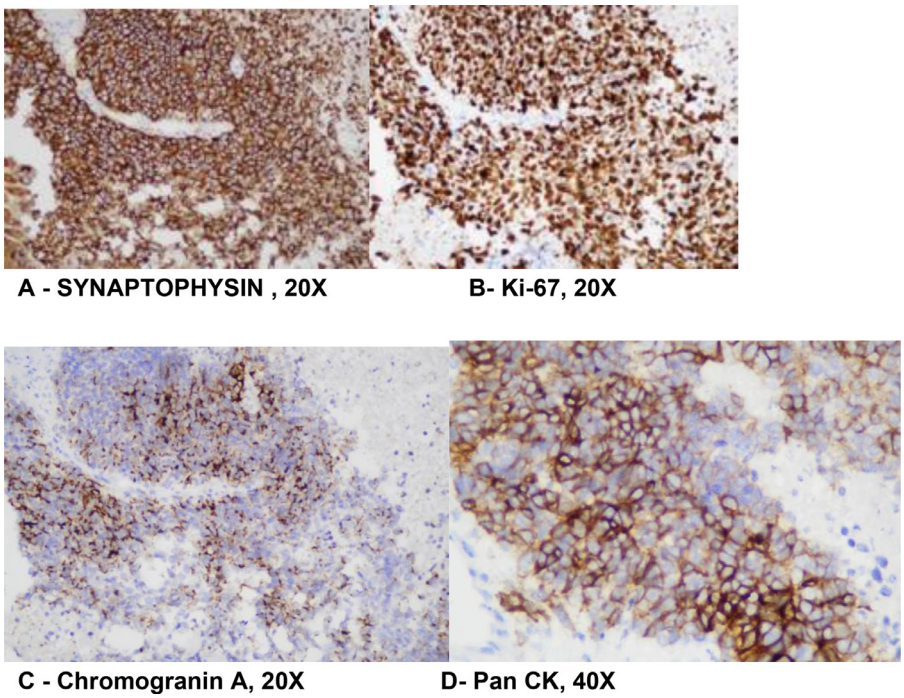

Fig. 8 – (A-D) Immunohistochemistry images positive for Synaptophysin, Ki-67 index, Chromogranin A and Pan:CK

Conclusion

Primary neuroendocrine tumors of the liver are rare, have better prognosis and different therapeutic management as compared to the rest of the malignant hepatic tumors; though these have nonspecific imaging appearance, they need to be considered as a differential in hepatic tumors; with predominant cystic/necrotic component, and an absence of significant lymphadenopathy and any other enhancing mass in the abdomen (Figs. 2-8).

Patient consent

Adequate consent has been obtained, however there is no disclosure of patient details in any form.

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