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Right hepatectomy for combined primary neuroendocrine and hepatocellular carcinoma. A case report

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ABSTRACT

INTRODUCTION: Cases of primary neuroendocrine tumors in the liver combined with hepatocellular carcinoma are scarce. Such cases could present either as combined-type tumor or collision type.
PRESENTATION OF CASE: A 51-year-old man presented with a mass in the right hemiliver. Serum level of alpha-fetoprotein was slightly elevated (2.3 ng/ml), with normal CA19-9 and CA125. The patient underwent right hepatectomy. The resected specimen showed a well-defined and heterogeneous gray-white to brown friable tumor, 20 cm in diameter. Microscopically, the tumor consisted predominantly of monotonous small- to medium-sized neoplastic cells arranged in trabeculea separated by sinusoidal spaces. Immunohistochemically, the tumor cells were strongly positive for synaptophysin and focally positive for chromogranin-A. Interestingly, the tumor cells showed patchy positive coarse granular staining of HerPar-1 involving about 1% of the tumor cells. Glypican-3 staining was negative. These immunohistochemical findings supported the diagnosis of combined high grade neuroendocrine carcinoma and hepatocellular carcinoma.
DISCUSSION: Cases of primary neuroendocrine tumors in the liver combined 82 with hepatocellular carcinoma are scarce. The uniqueness of this case lies in the fact that the neuroendocrine carcinoma component comprised more than 99% of the tumor area, and the minor hepatocellular carcinoma component was detected only by the immunohistochemical staining for HepPar-1.
CONCLUSION: To the best of our knowledge, this is the first case of combined neuroendocrine carcinoma and hepatocellular carcinoma in Egypt.

1. Introduction

Primary hepatic neuroendocrine carcinoma (NEC) is rare and its origin is not clearly understood. An admixture of hepatocellular carcinoma (HCC) and neuroendocrine carcinoma is particularly rare.[1] There are 2 types of primary mixed NEC and HCC in the liver. The combined-type tumor in which both NEC and HCC components intermingle with each other and cannot be clearly separated in the transitional area within a single tumor nodule,[2] and the collision-type in which the tumor have two histologically distinct tumors simultaneously involving the same organ with no transition between them.[3] We herein describe a case of combined primary high grade NEC and HCC of the liver in which the NEC component comprised more than 99% of the tumor area, and only small nests of HCC were detected only by immunohistochemical staining for HepPar-1.

2. Case report

A 51-year-old man presented to our outpatient clinic with a 9-month history of dull aching abdominal pain and a medical history of hepatitis C. Periodic hepatic ultrasound follow-up showed a mass in the right hemiliver. A CT scan of the abdomen revealed a 20 cm × 15 cm mass occupying most of the hemiliver with cystic degeneration and calcification and multiple dilated collaterals related to the mass (Fig. 1). The mass was compressing the right portal vein, yet no invasion detected (Fig. 2). CT volumetry estimated the residual left hemiliver volume including the middle hepatic vein to be 1300 cc. Serum level of alpha-fetoprotein (AFP) was slightly elevated (2.3 ng/ml). The patient had normal liver enzymes, total serum bilirubin of 1.2 mg/dl, serum albumin of 3.3 mg/dl and INR of 1.1. CA19-9 and CA125 were normal. CT of chest and extra-hepatic abdomen showed no other lesions.

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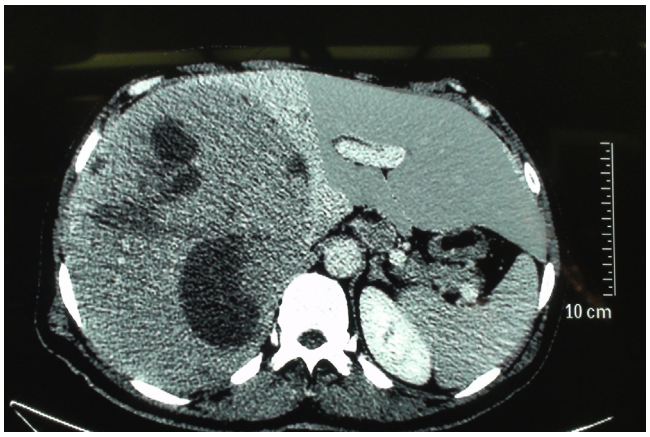

Fig. 1. Arterial phase abdominal CT scan showing large right hemiliver mass with areas of cystic degeneration. The residual left hemiliver including the middle hepatic vein being shadowed.

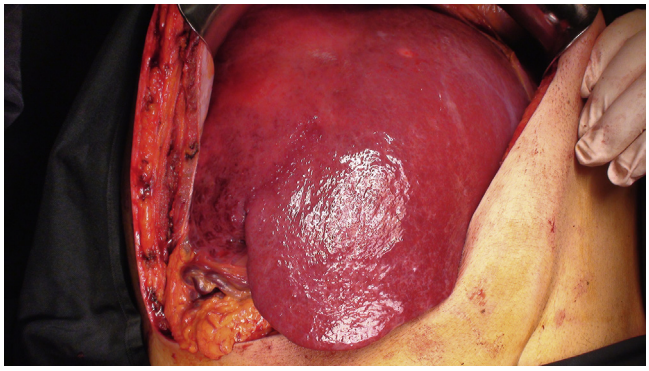

Fig. 3. Operative photography of the mass showing dilated veins extending from the mass to the hepatic flexure of the colon.

The patient underwent laparotomy through a J shaped incision (Fig. 3). On exploration the mass was found to be capsulated, 20 cm × 15 cm occupying most of the right hemiliver with multiple dilated collaterals extending from the mass to the diaphragm and the hepatic flexure of the colon. No ascites was found. No peritoneal nodules or significant lymph nodes. Right hepatectomy was done. Blood loss was 2500 cc and the patient received 4 units of blood intraoperatively due to presence of collaterals and large size of the mass making mobilization and resection of the right lobe technically challenging. The patient had an eventless post-operative course and was discharged on the 6th post-operative day.

The resected specimen measured 20 cm × 15 cm and weighed 4500 g. Cut section revealed a well-defined heterogeneous gray-white to brown friable mass 7.5 cm in diameter (Fig. 4). The surrounding liver was non-cirrhotic.

Microscopically, the tumor consisted predominantly of monotonous small- to medium-sized neoplastic cells arranged in trabeculea separated by sinusoidal spaces. Tumor cells had round to oval punctuate nuclei within conspicuous nucleoli and scanty cytoplasm (Fig. 5A). The tumor was accompanied by wide areas of geographic necrosis with high mitotic rate that ranged between 2 and 20/10 HPF. The whole morphological picture was reminiscent of high grade NEC. The surrounding liver tissue revealed chronic hepatitis without cirrhotic changes.

Immunohistochemically, the tumor cells were strongly positive for synaptophysin (Fig. 5D) and focally positive for chromogranin-A. Interestingly, the tumor cells showed patchy positive coarse granular staining for HerPar-1 in less than 1% of tumor cells (Fig. 5B and C) while Glypican-3staining was negative. The immunohistochemical findings supported the diagnosis of combined high grade NEC and HCC. Follow up period was short but no recurrence detected up to 6 months after surgery.

3. Discussion

Cases of primary neuroendocrine tumors in the liver combined with hepatocellular carcinoma are scarce.[2,4] It can present either as combined-type or collision type.[5] We herein describe a case of combined primary high grade NEC and HCC of the liver in which the NEC component comprised more than 99% of the tumor area, and a small nest of HCC was detected only by immunohistochemical staining for HepPar-1. In the case reported by Yang et al.,[5] the tumor consisted predominantly of small- to medium-sized

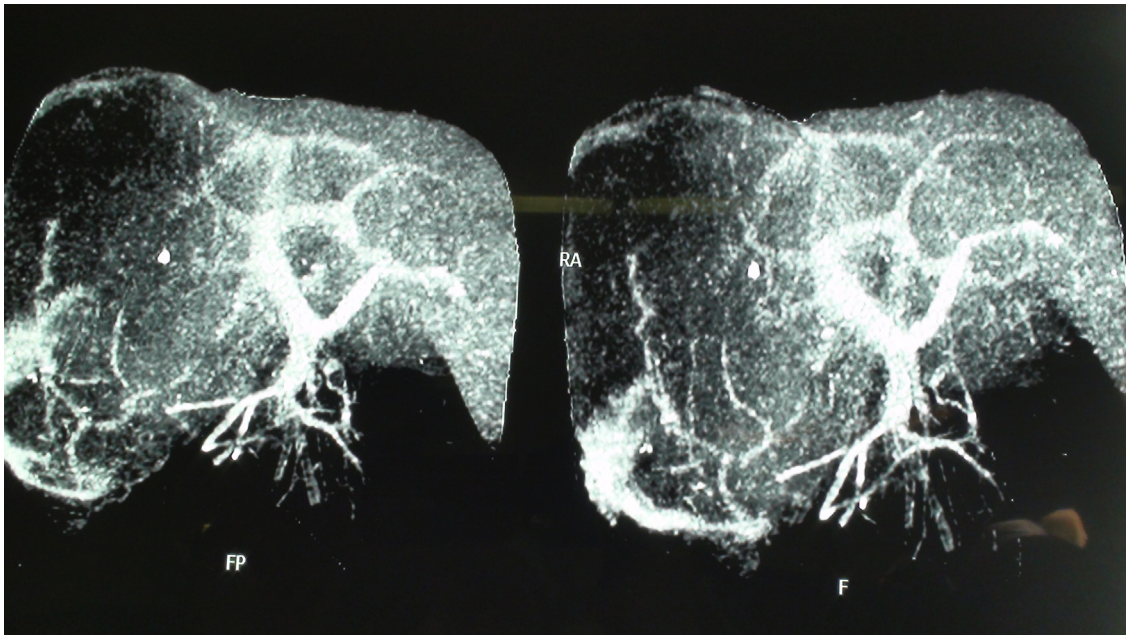

Fig. 2. CT portography and hepatic venography showing the relation of the mass to the portal and hepatic veins.