Pancreatic Hepatoid Carcinoma: A Very Rare and Intriguing Entity

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Introduction:

Hepatoid carcinoma (HC) is a rare tumor with features morphologically and immunohistochemically like focal hepatocellular carcinoma (HCC). HC is extremely rare in pancreas compared to other organs with less than 50 cases reported in the literature so far.

Case description:

67-year-old male with compensated cirrhosis due to hepatitis C, was evaluated for gradual weight loss and poor appetite. Abdominal ultrasound (US) showed a pancreatic head mass only and a cirrhotic liver. Computed tomography scan of the abdomen & pelvis with contrast revealed a 10.1 cm pancreatic head mass and another mixed attenuation 4.18 cm mass in lateral right hepatic lobe suggesting a metastatic neoplastic disease. CA 19-9 levels were normal, and Alfa Fetoprotein (AFP) levels were significantly elevated. Fine needle aspiration (FNA) of the mass via endoscopic US was initially reported as pancreatic adenocarcinoma. An excisional supraclavicular lymph node biopsy had features consistent with metastatic hepatoid (hepatocellular) carcinoma. Additional immunohistochemical staining and evaluation by pathologists of the pancreatic FNA sample confirmed a hepatoid (hepatocellular) carcinoma. The neoplastic cells were strongly positive for Cam 5.2, Hep par-1, arginase-1, glypican-3, villin, beta-catenin and SMAD-4. The patient was started on atezolizumab and bevacizumab therapy.

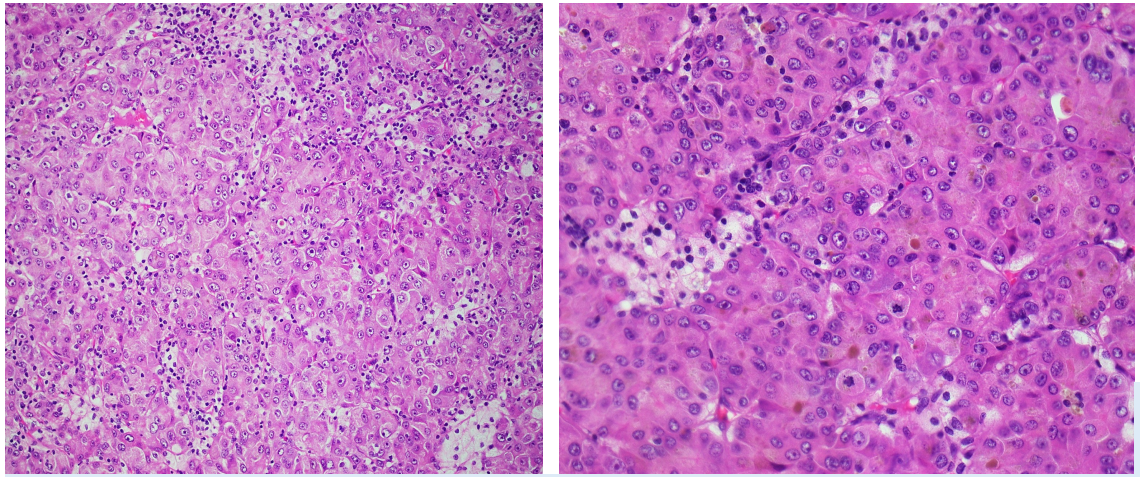

Figure 1 A and B: Lymph node Histopathology: Small and large nests of neoplastic cells with large amount of cytoplasm, prominent nucleoli and intranuclear inclusions. Few areas of tumor necrosis are seen.

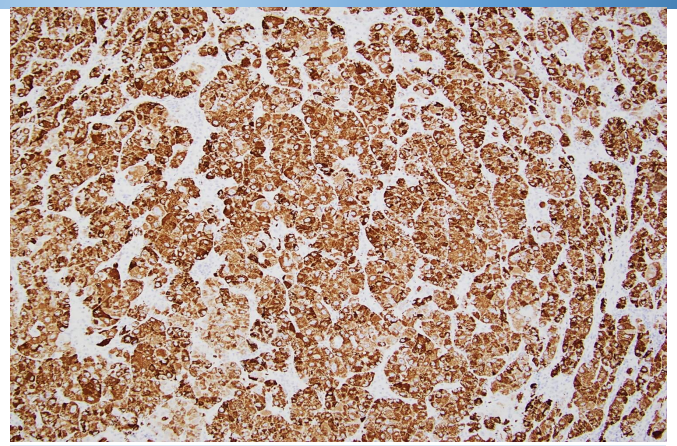

Figure 2: Pancreatic FNA specimen stained positive Hep par 1 staining

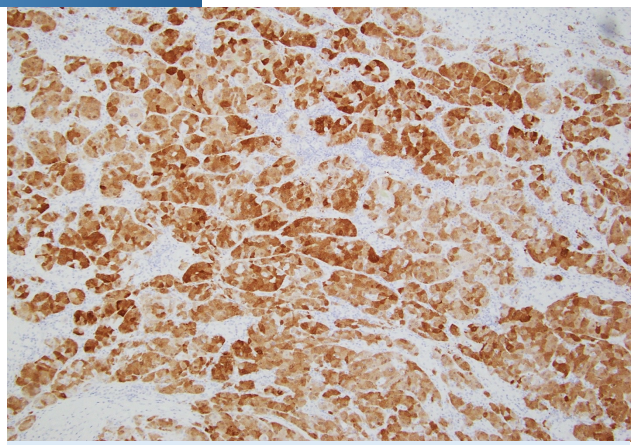

Figure 3: And the FNA specimen also stained positive Arginase 1.

Discussion:

The theories proposed to explain pathogenesis of pancreatic hepatoid carcinoma (PHC) are a) pancreas has ectopic liver tissue where an HC originates b) the pancreatic cells transdifferentiate into hepatocytes and c) there may be activation of the genes controlling hepatic differentiation of pancreatic cells during carcinogenesis, which are normally suppressed.

Four histological subtypes are noticed on a review of 41 cases: purely HCC-like morphology, with neuroendocrine differentiation, and with acinar or glandular differentiation.

Serum AFP levels are mostly elevated and used as a marker to determine the success of therapy. Hep Par-1 is thought to be the most sensitive among all the markers.

Prognosis is difficult to predict and there is no consensus on preferred chemotherapy due to limited data. Surgical resection is the preferred treatment option. Among different sub-types of PHC, pure HCC-like morphology has a better outcome than other sub-types . 1 year and 5 years survival in a case-series of 23 cases of HC were reported to be 71.1% and 40.4%, respectively.

References:

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