Intrahepatic Cholangiocarcinoma in a patient with Alcoholic Liver Cirrhosis and markedly elevated serum Alpha-Fetoprotein.

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Introduction

- Intrahepatic cholangiocarcinoma (ICC) represents 5% of all primary liver malignancies, second only to hepatocellular carcinoma (HCC).
- Although risk factors such as primary sclerosing cholangitis, hepatolithiasis, and asbestos have been reported, the etiology of ICC is not well understood.
- In high-risk patients, such as those with chronic liver disease, AFP > 400 ng/mL has a high positive predictive value for HCC and a specificity of >95%.
- An elevated serum AFP greater than 20 ng/mL is seen in <25% of ICC.
- We report a case of a patient with alcoholic liver cirrhosis and a markedly elevated serum AFP found to have ICC.

Case description

- A 62-year-old Puerto Rican male with a past medical history of alcoholic liver cirrhosis, Child-Pugh Class C presented with an acute change in mental status.
- Physical exam was remarkable for spider angiomas, jaundice, and icteric sclera. The abdomen was diffusely tender and distended with shifting dullness and hepatomegaly.
- Initial laboratory investigations revealed an obstructive pattern of transaminitis with a normal ammonia level, negative hepatitis panel, and serum AFP of 1955.8 ng/mL.
- Ultrasound-guided liver biopsy and immunohistochemical analysis demonstrated positive expression for cytokeratin 20, and CA 19-9.

Imaging and laboratory

INITIAL LABORATORY VALUES		
Platelets	125	130-499 K/UL
Corrected calcium	13.8	8.9-10.3 MG/DL
Sodium	128	136-144 MMOL/L
AST	104	15-41 U/L
Alkaline Phosphatase	185	38-126 U/L
Total Bilirubin	6.4	0.4-2 MG/DL
Direct Bilirubin	3.3	0.1-0.5 MG/DL

TABLE 1: Laboratory findings.

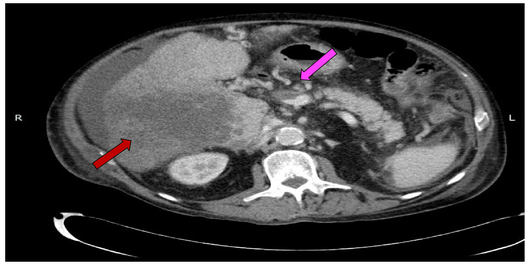

FIGURE 1: Computerized tomography (CT) scan of the abdomen with contrast showed a large infiltrative heterogeneous mass involving the entire right hepatic lobe with a non-occlusive thrombus extending to the portal vein.

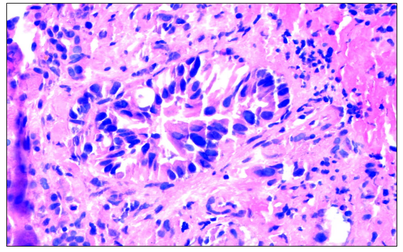

FIGURE 3: Immunohistochemical study shows positive CA 19-9 stain of adenocarcinoma.

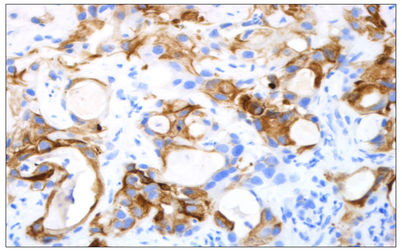

FIGURE 2: Histology of poorly differentiated adenocarcinoma composed of duct-like structures with high pleomorphism. H&E 400x.

Case description continuation

- The histological pattern was consistent with poorly differentiated adenocarcinoma, most likely pancreaticobiliary in origin consistent with probable intrahepatic cholangiocarcinoma (ICC).
- Due to the advanced nature of his disease, palliative management options were offered. However, the patient unfortunately expired.
- Neoplastic cells were negative for Hep-Par1, Glypican 3, and Arginase 1.

Discussion

- Elevated serum AFP accompanied by a space-occupying solid lesion in a cirrhotic patient is typically indicative of hepatocellular carcinoma (HCC).
- In this case, surprisingly, the histopathological report showed findings consistent with ICC and negative hepatic markers.
- Although unusual, this pathological entity should always be considered in a patient with alcoholic liver cirrhosis with a liver mass even in the presence of a markedly elevated serum AFP.
- Due to the aggressive nature of ICC most patients are diagnosed with lymph node involvement, intrahepatic metastasis, and peritoneal dissemination.
- Advanced ICC is refractory to chemotherapy and radiotherapy with a median survival rate of <2 years.

References

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