A RARE CASE OF SMALL CELL CHOLANGIOCARCINOMA

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Introduction

- Small cell cholangiocarcinoma is a rare entity and is encountered much less frequently than most other forms of cholangiocarcinoma.
- This case describes a 63-year-old female who presented with abdominal pain, fatigue, and nausea and was found to have a 1.4 cm mass at the confluence of the left and right intrahepatic bile ducts.
- This was deemed to be unresectable small cell cholangiocarcinoma, and she was ultimately provided palliative chemotherapy.

Case Presentation

- A 63-year-old female with a history of chronic obstructive pulmonary disease presented to the hospital with right upper quadrant pain, fatigue, nausea, and bloating.
- Laboratory studies revealed an alkaline phosphatase (ALP) of 917 U/L, total bilirubin of 3.2 mg/dL, alanine aminotransferase (ALT) of 552 U/L, aspartate aminotransferase (AST) of 285 U/L, and a normal lipase.
- A magnetic resonance cholangiopancreatography (MRCP) showed a 1.4 cm mass at the confluence of the left and right intrahepatic bile ducts leading to biliary obstruction.
- Endoscopic retrograde cholangiopancreatography (ERCP) demonstrated a stricture of the common bile duct for which a stent was unable to be deployed. The patient underwent biliary decompression via percutaneous drain placement.
- A biopsy was obtained during an attempt at surgical resection, but the mass was unable to be removed due to the involvement of the main portal vein.
- The AFP level returned at 7 ng/mL, CA 19-9 was 6 U/mL, and CEA was 1.6 ng/mL. Biopsies showed positivity for CAM 5.2, keratin AE1/AE3, CD56, synaptophysin, and TTF1. Ki67 index was >95%. Chromogranin was negative.
- The patient was treated with etoposide, atezolizumab, and carboplatin, however, the disease course was complicated by refractory disease and brain metastasis.

Discussion

- Small cell cholangiocarcinoma is much less prevalent than most other forms of cholangiocarcinoma.
- Symptoms and signs typically mimic the more common variants of cholangiocarcinoma and often include right upper quadrant discomfort, nausea, dyspepsia, jaundice, fatigue, and weight loss.
- Laboratory findings include elevations in ALP and bilirubin. ALT and AST may also be elevated, especially when the tumor is intrahepatic.
- Treatment depends on if the tumor is amenable to surgical resection. If it is unresectable, treatment with a platinum agent + etoposide +/- immunotherapy is often used.

Pathology

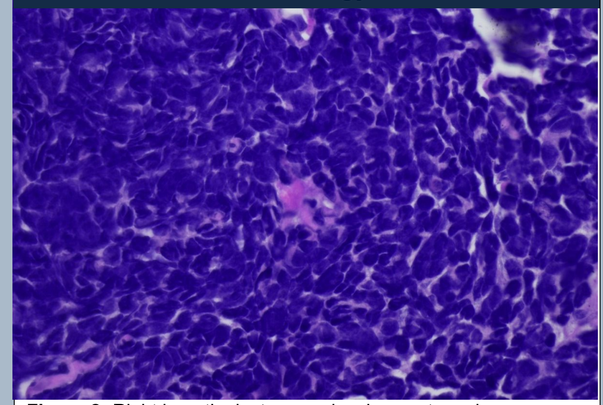

Figure 2: Right hepatic duct mass showing nests and sheets of malignant cells with small-to-intermediate-sized nuclei, fine chromatin, and scant cytoplasm consistent with small cell carcinoma (H&E 200X) *Pathology image courtesy Dr. Pham

Radiology

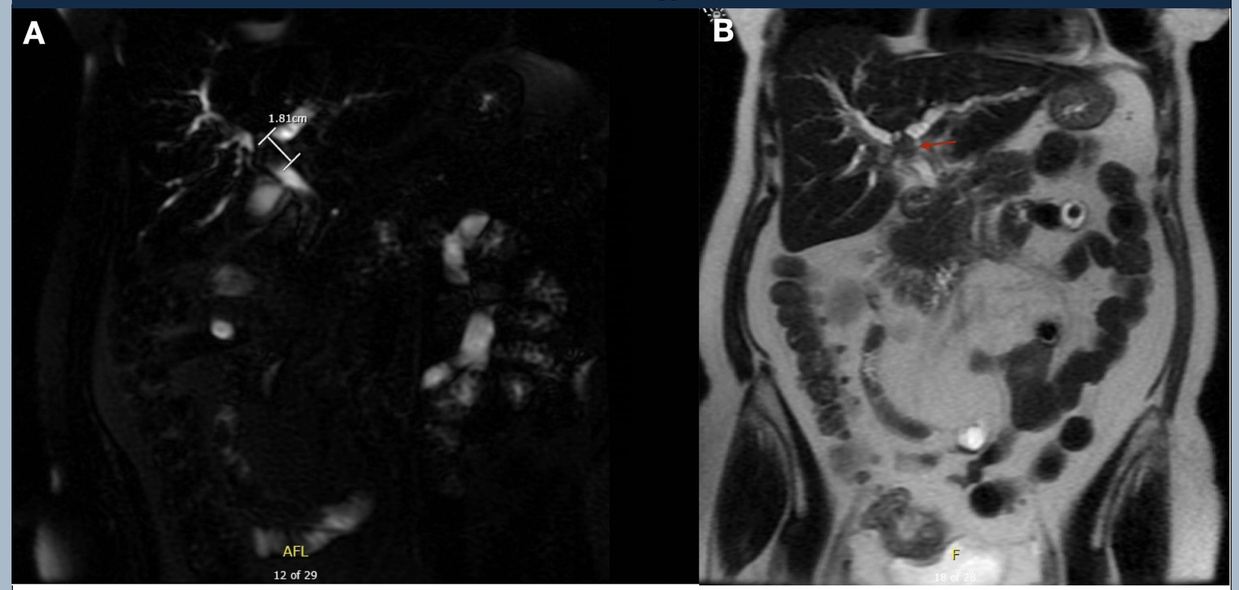

Figure 1A,B: MRCP showing an intrahepatic mass at the confluence of the right and left biliary ducts

Conclusion

- Neuroendocrine tumors originating from the biliary system are quite rare, and the prognosis is generally poor.
- Treatment options are like that of small cell pulmonary carcinoma and typically include systemic chemotherapy with a platinum agent and etoposide.
- We hope this case brings attention to this rare diagnosis so further research into this condition can be performed.

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