Isolated intraductal variant of hepatocellular carcinoma

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DESCRIPTION

A 70-year-old male teetotaller, with a history of chronic obstructive lung disease and hypertension, presented to the emergency department with a 2-day history of right upper quadrant pain and melaena. Investigations revealed total bilirubin 6.8 mg/dL, aspartate and alanine transaminases 88 and 122 IU/L, respectively, alkaline phosphatase 338 IU/L and gamma-glutamyl transpeptidase 223 IU/L. Tests for chronic hepatitis B, hepatitis C and HIV were negative. Serum alpha-fetoprotein was 22.8 ng/mL, whereas carcino-embryonic antigen (CEA), CA 19–9, prostate specific antigen and chromogranin levels were normal. Contrast CT of the abdomen revealed fatty liver with moderate dilation of bilateral intrahepatic biliary radicles (figure 1A) and dilated common bile duct (CBD) with hyperdense contents, without identifiable mass lesions in the liver or biliary tree. Upper gastrointestinal endoscopy revealed haemobilia, and cholangiogram showed normal calibre CBD with filling defects. Endoscopic intraductal ultrasound revealed focal thickening of segment three hepatic duct, and Spyglass cholangioscopy confirmed a white polypoidal lesion (figure 1B, black arrow) with abnormal surface vessels (figure 1B, white arrow) and few blood clots. Histopathology revealed few ductal cells with reactive atypia and streams of epithelial cells forming columns among vascular channels exhibiting mild variation in nuclear size (figure 1C). Immunohistochemistry showed pancytokeratin and hepatocyte antibody positive cells (figure 1D) lining vascular spaces that were negative for CD34, CK 7, CK 20, synaptophysin and chromogranin and exhibited high proliferation index, suggestive of hepatocellular carcinoma (HCC) of isolated intraductal type. Surgical options and palliation with biliary stenting and addition of sorafenib were suggested, the family opting for the latter. Eight weeks after stenting and initiation of sorafenib, liver functions normalised, while clinically, mild anorexia persisted.

We describe a patient with HCC, presenting as isolated intraductal mass in the absence of parenchymal mass lesions, tumour extension into the biliary tree or pure bile duct thrombus.

The earliest description of HCC of bile ducts without palpable surface liver mass on laparotomy was described by Sanford in 1952.[1] Thereafter, rare reports of such presentations were documented in the literature. In most cases, the liver was cirrhotic, and parenchymal lesions were identified on follow-up imaging. In all cases, the mass involved the extrahepatic bile ducts.[2] Our case is novel because HCC developed within intrahepatic bile ducts in a non-cirrhotic liver. Tsushimi *et al* hypothesised that HCC arising within bile ducts was from ectopic liver tissue.[3] We believe that ectopic theory along with epigenetic and genetic factors in the background of non-alcoholic fatty liver disease brings about variations in morphological and biological characteristics in HCC.

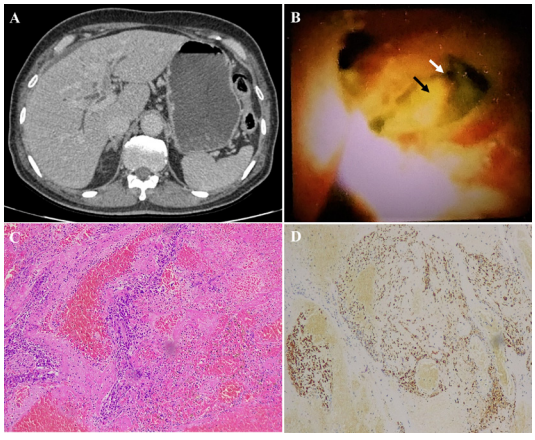

Figure 1 (A) Contrast CT of the abdomen showing dilation of intrahepatic biliary radicles without identifiable mass lesions in the liver or biliary tree. (B) Spyglass cholangioscopy of the biliary tree showing a polypoidal lesion in segment three bile duct. (C) Histopathology from the polypoidal lesion showing streams of epithelial cells forming columns and showing variation in nuclear size. (D) Immunohistochemistry of the intraductal lesion showing hepatocyte antibody positivity.

Learning points

- Presentation of hepatocellular carcinoma as an isolated intrahepatic intraductal mass in a non-cirrhotic liver has not been described before.
- In intraductal hepatocellular carcinoma, parenchymal lesions usually develop on follow-up imaging.
- Biopsy of biliary mass lesions presenting atypically, in cirrhotic or non-cirrhotic liver, could help in ideal treatment decision.
- Hepatocellular carcinoma must be kept as a differential diagnosis of isolated biliary mass lesions.

Contributors CAP designed the study and wrote the manuscript. RP and PM provided pertinent clinical images for the study. PA revised the manuscript and provided pertinent changes. All authors finalised the manuscript.

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