Extrahepatic cholangiocarcinoma in a patient with situs inversus totalis diagnosed by endoscopic ultrasound

Fig. 1 Images from computed tomography (CT) scanning in a 67-year-old man with situs inversus totalis showing: **a** the mirrored anatomy of the chest, with the heart and descending aorta on the right side; **b** the mirrored anatomy of the abdomen, with the pancreas and spleen on the right side and the liver on the left side, along with dilatation of the extrahepatic bile duct.

Situs inversus totalis (SIT) is a rare congenital abnormality characterized by a left-right reversal of the positions of the internal organs. A 67-year-old man with SIT came to our attention with a recent onset of jaundice.

Computed tomography (CT) scanning showed dilatation of the biliary tree and truncation of the common bile duct (CBD) just above the papilla; no evidence of an ampullary or pancreatic mass was seen (Fig. 1).

Although to overcome potential technical limitations, it is advocated that patients with SIT lie in the right decubitus position, we performed radial scanning endoscopic ultrasound (EUS; Olympus GF UE160) without difficulties, keeping our

patient in the left decubitus position and under conscious sedation (Fig. 2a,b). The papilla of Vater appeared enlarged with normal overlying mucosa. EUS showed significant wall thickening and a 2-cm hypoechoic mass in the distal CBD, with infiltration of the papilla and of the pancreas (Fig. 2c); the proximal CBD was dilated (Fig. 2d). Because of the anatomical inversion, the echoendoscope had to be rotated clockwise rather than counterclockwise to explore the CBD from the papilla to the liver hilum (Fig. 2e). Subsequently, linear scanning EUS (Olympus GF UCT140) with fine needle aspiration (FNA) was carried out in the same fashion and without difficulties. Cytological examination of the periampullary

lesion revealed cholangiocarcinoma (Fig. 3). On the basis of the results from the EUS, the patient initially underwent endoscopic retrograde cholangiopancreatography (ERCP) with biliary stenting for palliation of jaundice. He then underwent a Whipple's procedure 2 weeks later, from which he made an uneventful recovery.

EUS is a very accurate technique for diagnosis and staging of most etiologies of extrahepatic biliary obstruction. However, reported cases of extrahepatic biliary tumor in patients with SIT have been investigated only by extracorporeal imaging [1–4]. A fear of technical difficulties due to the altered anatomy may account for the relative underutilization of EUS in this setting. To the best of our knowledge, EUS has been described only once before in a patient with SIT and extrahepatic cholangiocarcinoma but technical details were not provided [5].

In conclusion, using EUS performed under routine conditions, we were able to diagnose a small cholangiocarcinoma that had been missed on CT scanning. EUS proved to be feasible and accurate in this patient despite the associated SIT.

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Competing interests: None

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Fig. 2 Images from radial endoscopic ultrasound (EUS) showing: **a** the mirrored anatomy of the mediastinum with the left atrium (LA) and descending aorta (A) on the right, and the azygos vein (AZ) on the left, with the thoracic duct (TD) and the spine (S) also visible; **b** the mirrored anatomy of the retroperitoneum with the splenic vein (SV) on the right, the portal vein (PV) and hepatic artery (HA) on the left, with the superior mesenteric artery (SMA) also visible; **c** a small hypoechoic tumor (T) infiltrating the papilla of Vater and the juxta-ampullary pancreatic parenchyma (inset: endoscopic view of the enlarged papilla); **d** hypoechoic thickening (*) of the distal common bile duct (CBD); **e** the tumor (T), hypoechoic thickening of the distal CBD (*) and dilatation of the proximal CBD, all with inverted anatomy.

Fig. 3 Cytology of the specimen obtained during endoscopic ultrasound (EUS) showing gland-like clusters of small atypical epithelial cells with prominent nucleoli and moderate amounts of cytoplasm (Papanicolaou stain; magnification $\times 40$).

Bibliography

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