Congenital absence of the intrahepatic portion of the portal vein in an adult male resected for hepatocellular carcinoma

Abstract We report a case of congenital absence of the portal vein (CAPV) in an adult male who underwent resection for hepatocellular carcinoma. CAPV is a rare anomaly; only 29 cases, mostly in children, have been presented to date. An association between CAPV and cardiac abnormalities, polysplenia syndrome, skeletal abnormalities and liver tumors has been seen in children. To the best of our knowledge this is the first case of CAPV in an adult male resected for hepatocellular carcinoma. However, since CAPV can be without symptoms, the number of undiagnosed cases is not known.

Keywords Liver vascular supply · Portal vein anomalies · Congenital absence of portal vein · Hepatocellular carcinoma

Introduction Congenital absence of the portal vein (CAPV), a rare anomaly, was first reported by Abernathy in 1793 [1]. A short main stem of the portal vein or a separate entrance of the superior mesenteric and splenic veins into the supra- or intrahepatic portions of the inferior vena cava (IVC), the left renal vein or iliac veins, as well as portal blood diversion through collaterals to the ayzygos vein has been described [2, 3, 4, 5, 6, 7, 8, 9, 10, 11, 12, 13, 14, 15, 16]. Most cases (25 of 29) have been in children; up to now only four patients more than 18 years old at diagnosis have been reported.

We present herein a case of CAPV, with the portal vein emptying into the suprahepatic IVC, in a 51-year-old male with a hepatocellular carcinoma. To the best of our knowledge, this is the first reported case of an adult male with CAPV presenting with a malignant liver neoplasm.

Case report A previously healthy, 51-year-old male sought medical attention because of fatigue, dizziness and 10 kg weight loss. Ultrasonography and CT showed a > 10-cm-sized tumor originating in the left liver lobe (Fig. 1).
A laparoscopy was performed in order to obtain biopsies and to make a resectability assessment. The tumor seemed to grow in the left liver lobe only, and there was no evidence of extrahepatic tumor growth. When the surgical biopsies were obtained from the tumor, a marked arterial bleeding was seen from the biopsy site and diathermy had to be used to stop the bleeding. The biopsies showed a hepatocellular tumor, either an adenoma or a highly differentiated hepatocellular carcinoma. No portal venous structures could be seen in the biopsy specimens.

At a preoperative abdominal US, the right liver lobe appeared normal, and the tumor was present in the left lateral segment of the liver only. The main stem of the portal vein could not be seen in the liver hilum, and the hepatic artery was wider than usual. The hepatic veins and the inferior vena cava showed no abnormality.

The bilirubin and S-glutamyltransferases (S-GT) were within normal limits. The alkaline phosphates (ALP) was 8.7 µmol/l (normal 0.8–4.6), aspartat-aminotransferases (S-ASAT) 1.2 µmol/l (normal < 0.7), alalin–aminotransferas (S-ALAT) 0.68 µmol/l (normal < 0.7). The blood chemistry values were all within normal limits, including P-ammonium. No clinical or serological evidence of hepatitis was present.

There was no clinical evidence of encephalopathy and the electro-encephalogram (EEG) was normal.

At the subsequent laparotomy, the tumor was seen to occupy the lateral segment of the left liver lobe only, and a radical resection was performed. The rest of the liver was without abnormalities. During hilar dissection and with peroperative US it was noted that the main portal vein, instead of passing in the hepatoduodenal ligament to the liver hilum, emptied directly into the inferior vena cava (IVC; Fig. 2). The hepatic artery were wider than usual, almost 1 cm in diameter.

Except for slight postoperative fever, due to resorption of a hematoma, the postoperative course was uncomplicated. Postoperative CT verified the portal vein entrance into the infrahepatic IVC, normal splenic (SV) and superior mesenteric veins (SMV), and the wide hepatic artery (HA; Fig. 3). The patient has remained well for 2 years without evidence of tumor recurrence.

The final pathological diagnosis was quite vascular, radically resected 12-cm-sized, moderately differentiated hepatocellular carcinoma. The liver was nodular and there were positive hepatitis-B markers (core antigen) in the non-tumorous hepatocytes suggesting a previous hepatitis-B infection.

**Discussion**

Congenital absence of the portal vein is a rarely reported anomaly. To date, only 29 cases have been presented including the original paper by Abernathy in 1793. The development of the portal venous system, with the formation of paired communications between the right and left vitelline veins, form the embryological base of variations in the portal venous system, like a preduodenal portal vein, CAPV, portal vein atresia and duplications, as well as communications between the portal venous system and the pulmonary veins [2, 4, 6].

Congenital absence of the portal vein with either end-to-side or side-to-side diversion of the portal blood flow has been named the Abernathy malformation [11]. It has been suggested that the side-to-side shunts should be surgically corrected to prevent the development of liver encephalopathy [11], but it has also been stated that most cases with CAPV would be undiagnosed if it were not for the concomitant occurrence of other diseases such as cardiac defects and liver tumors [8].

The portal venous blood in CAPV has been reported to enter the IVC through a short common portal vein trunk in 18 of 29 cases [1, 3, 6, 7, 8, 11, 12, 13, 14, 15, 16], or with a separate entrance of the SV and SMV into the IVC in 3 of 29 cases [15]. In 4 patients the portal blood flow drained through the left renal vein [4, 5, 9, 15], in