Hepatocellular carcinoma in a non-failing Fontan circulation

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CASE REPORT

A 41-year-old woman with a tricuspid atresia and a pulmonary stenosis operated on at eight years old with a classic Fontan operation was referred for evaluation due to ascites without limb edemas. Cardiac magnetic resonance imaging (MRI) showed a rudimentary right ventricle (RV) and a giant right atria (RA) with the right atrial appendage (arrowhead) connected to the pulmonary artery (PA) (Fig. 1A). Abdominal MRI showed the liver with parenchymal heterogeneity, dilatation of the suprahepatic veins (SHV) and ascites (asterisk) (Fig. 1B). Hepatic MRI with and without contrast showed a 12 mm nodule in the liver suggestive of hepatocellular carcinoma (Fig. 2A-C, arrowhead) according to the European Association for the Study of the Liver (1), ascites (asterisk) and a dysplastic nodule (Fig. 2D, arrowhead). No significant gastro-esophageal varices were seen. Hemodynamic evaluation showed a SHV pressure of 14 mmHg and a hepatic venous pressure gradient of 2 mmHg with no stenoses in the Fontan pathway. Serum-ascites albumin gradient was < 1. Liver biopsy was rejected due to the high risk of bleeding. Unfortunately, whilst waiting for chemotherapy embolization the patient died.

DISCUSSION

Patients after a Fontan operation exhibit some degree of fibrosis which are primarily located in a sinusoidal or centrilobular pattern. Cirrhosis may develop years later and the incidence of cancer is up to 5% (2). That is why some authors recommend that patients who are > 10 years out from their Fontan operation should undergo cardiac assessment as well as the determination of alpha-fetoprotein level, the realization of liver imaging tests or even liver biopsy (3) to stay ahead of neoplastic transformation.

REFERENCES