

Image in cardiology

Fontan-associated liver disease after heart transplant

Evolución postrasplante de la hepatopatía asociada a Fontan

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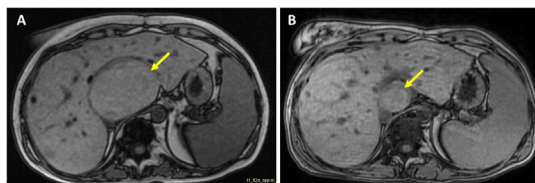


Figure 1.

The present report concerns a 32-year-old woman with complex univentricular congenital heart disease (pulmonary and tricuspid atresia) treated with an extracardiac Fontan procedure who was being assessed for heart transplant (HTx) suitability; all relevant consent forms were signed. Vena cava and pulmonary conduit and artery pressures were 15 mmHg while the transhepatic gradient was 4 mmHg. Angiography revealed multiple venovenous and aortopulmonary collaterals that were not amenable to percutaneous occlusion due to their small size. The patient had moderate Fontan-associated liver disease (Child-Pugh B8) and posthepatic portal hypertension (small esophageal varices, splenomegaly, and thrombocytopenia). Liver magnetic resonance showed multiple hepatocellular adenomas; the largest was 60 × 80 × 78 mm (figure 1A, arrow). Biopsy demonstrated METAVIR stage F3–4 fibrosis in the liver parenchyma and no malignancy of the main adenoma. After evaluation by the hepatology department, the patient was considered suitable for isolated HTx.

The transplant was conducted via the bicaval technique but was complicated by significant surgical bleeding that required delayed sternal closure after 48 hours. Subsequently, the patient was stable with a well-functioning graft.

Liver resonance 1 year after the transplant showed a significant reduction in adenomas; the largest measured 35 mm in size (figure 1B). Three years after the transplant, the main adenoma measured 30 mm and there was no significant portal hypertension while liver function was similar (Child-Pugh A6). The adenoma regression was probably at least partly due to the restoration of the biventricular circulation and normalization of the posthepatic pressure. Although the liver biopsy was not repeated, it seems that the Fontan-associated liver disease probably stabilized after the HTx and even showed signs of reversing.

FUNDING

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ETHICAL CONSIDERATIONS

International recommendations on clinical research (Declaration of Helsinki) have been respected. Sex and gender variables have been considered, in accordance with Sex and Gender Equity in Research (SAGER) guidelines. The patient provided signed informed consent.

STATEMENT ON THE USE OF ARTIFICIAL INTELLIGENCE

No artificial intelligence was used in the preparation of this work.

AUTHORS' CONTRIBUTIONS

All authors meet the requirements for authorship and contributed equally.

CONFLICTS OF INTEREST

None.

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