A newborn liver mass that never existed: a somber reminder of embryonic ties between umbilical vein and portal venous system

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Pediatric gastroenterology was consulted to evaluate abnormal liver tests and upper gastrointestinal (GI) bleeding in a 6-day-old Caucasian male, known to have transposition of the great vessels, requiring emergent atrial septostomy on the first day after birth. Before this procedure, at a referring hospital, the infant suffered a significant hypoxic event requiring dopamine, prostaglandin E1, sodium bicarbonate, and normal saline administered through umbilical venous catheter. After the septostomy, his neonatal intensive care unit course was relatively stable except abdominal distention and recovery of small quantities of bright red blood recovered through nasogastric tube left on intermittent suctioning. The abdomen was soft but distended without hepatosplenomegaly. Initial laboratory investigations revealed albumin at 2.7 g/dL, total bilirubin 5.2 mg/dL (normal < 0.8 mg/dL). The direct bilirubin was 0.9 mg/dL, alanine transaminase of 842 U/L and the alanine transaminase of 195 U/L (normal 45 for both), alkaline phosphatase 146 U/L, platelet count 115 × 10⁹/L and an international normalized ratio of 5.9. Ultrasound of the liver revealed a heterogeneous 3.7 × 2.9 cm “mass lesion” in the left hepatic lobe (Figure 1B) prompting further work up to rule out hepatoblastoma, hemangiendothelioma or mesenchymal hamartoma. Computed tomography scan of the abdomen revealed hyper-perfusion in the left lobe (Figure 1C) with areas of hypo- and hyper-attenuation (Figure 1D) suggesting compromised blood flow throughout the left lobe as the etiology of the lesion. Poor visualization of the left portal venous system and a clot in a branch of left portal vein with periportal edema (Figure 1D) supported the impression of thrombosis of left portal system. GI bleeding resolved when thrombocytopenia and coagulation parameters were optimized. All biochemical and hematological abnormalities resolved in 2 weeks. Three weeks latter, prior to reconstruction of the great vessels, a CT scan of the liver revealed resolution of the left liver lobe lesion except for minimal residual hypo-attenuation at the liver margin below the diaphragm.
This case illustrates the significance of correct positioning of umbilical venous catheters (UVC) to avoid compromised portal vascular flow. Although liver-related complications are uncommon, liver abscess, hepatic laceration, total parenteral nutrition ascites, and hepatic necrosis have been reported following hepatic placement of UVC. We attributed GI bleeding to tube-trauma in the setting of thrombocytopenia, because, without any additional therapies it resolved once the suction was discontinued. Similarly, although some degrees of portal hypertension is expected with thrombosis of portal venous system, the temporal profile (less than a week) is not supportive of portal hypertensive GI bleeding incidental to longstanding hepatic vascular or parenchymal diseases leading to portal hypertension. In the case of a neonate with abnormal liver tests, physicians should carefully review if a UVC was placed. If so, the correct placement of the catheter should be determined. The UVC initially noted to be at hepatic level was appropriately repositioned to lower cavo-atrial junction before atrial septostomy in this case. Hepatic abnormalities persisted for 2 weeks, however, presumably because of use during initial resuscitative efforts.

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References