Hepatic subcapsular flow: An early marker in diagnosing biliary atresia

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ABSTRACT
We report an interesting sign in the sonological evaluation of neonatal cholestasis, which is hepatic subcapsular flow. Hepatic subcapsular flow is an early and useful marker in diagnosing biliary atresia.

Key words: Biliary atresia; hepatic artery diameter; hepatic subcapsular flow; triangular cord sign

Introduction
Even with all available investigations, it is very difficult to differentiate biliary atresia from non-biliary atresia cases. In this context, we report the usefulness of a color Doppler finding, that is, hepatic subcapsular flow, in diagnosing biliary atresia.

Case Report
An 11-day-old male baby presented with yellowish discoloration of eyes and dark urine and persistently pale-colored stools since birth. On clinical examination, the baby was active and weight gain was adequate, with hepatomegaly 3 cm below the costal margin, firm, and no splenomegaly.

On further evaluation, the baby was found to have conjugated hyperbilirubinemia with high gamma-glutamyl transpeptidase (GGT). Prothrombin time (PT) was normal and serum albumin 2.9 was g/dl; Ig M antibodies against Toxoplasma, Rubella, Cytomegalovirus, Herpes virus, sepsis, Inborn Error of Metabolism screening, serum ferritin, alpha-fetoprotein, and urine examination were normal. Hepatobiliary iminodiacetic acid (HIDA) scan showed no gut activity even after 24 h.

USG [LOGIC 500MD GE Japan] showed the following: Enlarged liver with diffuse mild increased echotexture, triangular cord sign: 3.2 mm, hepatic artery diameter 0.9 mm, and subcapsular flow was present [Figure 1].

In view of persistently pale stools in a well-thriving child, high GGT, non-excretory HIDA, and presence of subcapsular hepatic flow (though the triangular cord sign was negative), the baby was taken up for peroperative cholangiogram/Kasai’s procedure on 23rd day of life.

At surgery, gall bladder was found to be atretic and cord-like. Cannula could not be passed, hence complete peroperative cholangiogram could not be done, and we proceeded to do modified Kasai’s portoenterostomy. Liver was dark green, and the surface of the liver showed telangiectatic hepatic artery branches [Figure 2] and no micro/macro nodules. Portal dissection was done. No demonstrable bile duct was seen; fibrotic portal plate was excised and roux-en-Y jejunal loop was anastomosed to porta hepatis. Liver biopsy done showed features suggestive of biliary atresia with fibrosis score of 1/6 (Ishak’s score).

Discussion
No single test is confirmatory for diagnosing biliary atresia. USG is a useful noninvasive test used for the evaluation of
babies with neonatal cholestasis. Sonographic triangular cord sign, GB length and contractility, hepatic artery diameter, and other parameters have been extensively studied in differentiating biliary atresia from non-biliary atresia cholestasis cases. Stowen for the first time described about the hyperplastic and hypertrophic changes in the branches of hepatic artery in the intrahepatic portal areas of the patients with biliary atresia.[1] In hepatobiliary specimens obtained from patients with biliary atresia, enlargement of the hepatic artery had been noted.[2‑3] Enlargement may be a compensatory change to improve the blood supply for the biliary trees, a secondary change of liver cirrhosis, or an essential vascular malformation.[2‑3] In an angiographic study[6] of biliary atresia, all patients with biliary atresia had hepatic artery enlargement and the branches of the intrahepatic peripheral hepatic artery manifested with irregular contours suggestive of peripheral occlusion. However, pathogenesis of the hepatic arteriopathy still remains uncertain.

Recently, Lee et al. described the usefulness of subcapsular hepatic flow in diagnosing biliary atresia.[7] Color Doppler USG images demonstrated an enlarged hepatic artery and hepatic arterial flow that extended to the hepatic surface in all patients with biliary atresia. Hepatic subcapsular flow had a sensitivity, specificity, and positive and negative predictive values of 100%, 86%, 85%, and 100%, respectively, in their study. All patients with biliary atresia who had hepatic subcapsular flow on color Doppler USG images had subcapsular telangiectasia vessels at the time of the Kasai’s procedure.

In our case, though triangular cord sign was less than 4 mm, presence of subcapsular hepatic flow gave a clue to the diagnosis. Per-operative findings confirmed the presence of telangiectic hepatic artery branches. Histopathology showed mild fibrosis (according to Ishak’s scoring system for liver fibrosis).[8]

Conclusion

Hepatic subcapsular flow is an early sign and further studies are required to demonstrate the usefulness of this sign.

References


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