A Newborn with Congenital Intrahepatic Arterioportal Fistula

Zhiqiu Ye¹ · Jianlan Wang² · Zhuxiao Ren² · Jie Yang²

Received: 2 September 2020 / Accepted: 24 November 2020 / Published online: 7 January 2021
© Dr. K C Chaudhuri Foundation 2021

To the Editor: A male neonate presented with hepatomegaly and tachypnea soon after birth. He needed low-flow oxygen support and treated in local hospital. Fetal ultrasound indicated suspicious enlargement of the heart and liver. On the 14th day of life, he was transferred to our hospital complaining of hepatomegaly, tachypnea, and prominent abdominal distension of 3 d duration. On physical examination, the epigastric veins were slightly engorged and the lower margin of the liver was found at 3 cm below the xiphoid process. Computed tomography (CT) scan and angiography revealed multiple hepatic hemangiomas, hepatic arterioportal fistula, significant ascites, and enlarged celiac shaft (Supplementary Fig. 1a and Fig. 2a). He was diagnosed as congenital intrahepatic arterioportal fistula (IAPF) and accepted coil embolization on day 24 of life (Supplementary Fig. 2b). Immediate improvement of symptoms including hepatomegaly, tachypnea, and prominent abdominal distension was noticed and he was discharged home 6 d after coil embolization. He remained asymptomatic and had satisfactory result of repeated CT on one-year follow-up (Supplementary Fig. 1b) and kept healthy on two-year follow-up. Congenital IAPF, one of the causes of portal hypertension, is uncommon in neonate and interventional radiological procedures are the preferred treatment for most cases with IAPF [1].

Several cases of congenital IAPF had been reported. In 1998, a similar case was reported by Cheung et al. [2], which

described a neonate presented tachypnea and feeding intolerance and diagnosed with hepatoportal arteriovenous fistulas, and then the baby was treated with transcatheter coil embolization and remained asymptomatic on follow-up. In another case, an infant with IAPF was not identified until about 80 d after birth because of septic shock. However, the infant presented with dehydration and hyperbilirubinemia on day 3 of age, and pneumonia and constipation at 1 mo of age [3]. The above cases of congenital IAPF show these baby had abnormal digestive systems and suggest that vascular malformations should be considered if puzzling abdominal symptoms or signs appear during the neonatal period without any obvious cause. To our knowledge, early coil embolization for neonates with congenital IAPF is effective and safe.

Supplementary Information The online version contains supplementary material available at <https://doi.org/10.1007/s12098-020-03587-3>.

Compliance with Ethical Standards

Ethics Approval It is approved by the ethics committee: Guangdong Women and Children Hospital Ethics committee.

Conflict of Interest None.

Informed Patient Consent Obtained.

References

1. Norton SP, Jacobson K, Moroz SP, et al. The congenital intrahepatic arterioportal fistula syndrome: elucidation and proposed classification. *J Pediatr Gastr Nutr.* 2006;43:248–55.
2. Cheung YF, Leung MP. Coil embolization of hepatoportal arteriovenous fistula in a neonate. *Clin Cardiol.* 1999;22:675–6.
3. Bogert JN, Potter DD, Crow S, Arteaga GM, Freese DK. An unusual presentation of a congenital intrahepatic arterioportal fistula in an infant with down syndrome. *J Pediatr Surg.* 2011;46:252–5.

Publisher's Note Springer Nature remains neutral with regard to jurisdictional claims in published maps and institutional affiliations.

Zhiqiu Ye and Jianlan Wang have equivalent contribution to the study.

✉ Jie Yang
jiejyang0830@126.com

¹ Department of Invasive Technology, Guangdong Women and Children Hospital, Guangzhou Medical University, Guangzhou, China

² Department of Neonatology, Guangdong Women and Children Hospital, Guangzhou Medical University, Guangzhou 510010, China