Abernethy Malformation in Newborn. Ultrasound Diagnostic

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Abstract

Congenital absence of the portal vein (The Abernethy malformation – AM) is a rare anomaly in which the intestinal and the splenic venous drainage bypass the liver and drain into systemic veins through various venous shunts. The different variants of congenital portosystemic shunts are present in one in 30,000 children. Morgan and Superina proposed the following classification of portosystemic anomalies; either the liver is not perfused with portal blood because of a complete shunt (Type I) or the liver is perfused with portal blood due to the presence of a partial shunt (Type II). Additional anomalies are common in AM: more then 22 % of patients had congenital heart disease; hyperammonemia/neurological abnormalities (9 – 35 %), neonatal cholestasis (13 %) and pulmonary hypertension or hepatopulmonary syndrome (18 %). Other commonly found abnormalities of the spleen, urinary and male genital tract, brain and skeletal anomalies. Hepatic changes such as focal nodular hyperplasia, hepatocellular carcinoma, and hepatoblastoma are diagnosed in more then 40 % of patients. In ours cases four different varieties of congenital portosystemic venous shunts are described in newborn during a 10-year period, age from 1 to 17 days: 1 had a complete shunt (Type I) and 3 — Type II (larges intrahepatic shunts). Only 1 preterm boy (33 week, 1900 g) with a duodensl atresia had great hyperbillirubinemia, the other 3 where asymptomatics, the AM was found fortuitously on an abdominal ultrasound. The intrahepatic shunts resolved spontaneously within one year of age. This article also illustrates the ultrasound findings of AM.

Key words: Ultrasonography, Newborn, Abernethy Malformation.

References

1. *Olkhova E. B.* The viscerals pseudoaneurisms in the childhood: the variants of ultrasonography findings. Radiology — practice. 2013. No. 2. P. 32–41.
2. *Alonso-Gamarra E., Parrón M., Pérez A., Prieto C., Hierro L., López-Santamaría M.* Clinical and radiologic manifestations of congenital extrahepatic portosystemic shunts: a comprehensive review. Radiographics. 2011. V. 31. No. 3. P. 707–722.
3. *Barchetti F., Pellegrino L., Al-Ansari N., De Marco V., Scarpato P., Ialongo P.* Congenital absence of the portal vein in a middle-aged man. Surg. Radiol. Anat. 2011. V. 33. No. 4. P. 369–372.
4. *Bernard O., Franchi-Abella S., Branchereau S., Pariente D., Gauthier F., Jacquemin E.* Congenital portosystemic shunts in children: recognition, evaluation, and management. Semin. Liver Dis. 2012. V. 32. No. 4. P. 273–287.
5. *Blanc T., Guerin F., Franchi-Abella S., Jacquemin E., Pariente D., Soubrane O., Branchereau S., Gauthier F.* Congenital portosystemic shunts in children: a new anatomical classification correlated with surgical strategy. Ann Surg. 2014. V. 260. No. 1. P. 188–198.
6. *Guérin F., Blanc T., Gauthier F., Abella S. F., Branchereau S.* Congenital portosystemic vascular malformations. Semin. Pediatr. Surg. 2012. V. 21. No. 3. P. 233–244.
7. *Kim M. J., Ko J. S., Seo J. K., Yang H. R., Chang J. Y., Kim G. B., Cheon J. E., Kim W. S.* Clinical features of congenital portosystemic shunt in children. Eur. J. Pediatr. 2012. V. 171. No. 2. P. 395–400.

8. *Kobayashi N., Niwa T., Kirikoshi H., Fujita K., Yoneda M., Saito S., Nakajima A.* Clinical classification of congenital extrahepatic portosystemic shunts. Hepatol. Res. 2010. V. 40. No. 6. P. 585–593.
9. *Kong Y., Zhang H., Liu C., Wu D., He X., Xiao M., Zhao G., Zhang H.* Abernethy malformation with multiple aneurysms: incidentally found in an adult woman with Caroli's disease. Ann. Hepatol. 2013. V. 12. No. 2. P. 327–331.
10. *Lisovsky M., Konstas A. A., Misdraji J.* Congenital extrahepatic portosystemic shunts (Abernethy malformation): a histopathologic evaluation. Am. J. Surg. Pathol. 2011. V. 35. No. 9. P. 1381–1390.
11. *Lu J., Lin Z., Liu H., Liu Z.* An unusual presentation of type II Abernethy malformation. Ann. Vasc. Surg. 2014. V. 28. No. 6. P. 1567.
12. *Mistinova J., Valacsai F., Varga I.* Congenital absence of the portal vein Case report and a review of literature. Clin. Anat. 2010. V. 23. № 7. P. 750–758.
13. *Scalabre A., Gorincour G., Hery G., Gamerre M., Guys J. M., de Lagausie P.* Evolution of congenital malformations of the umbilical-portal-hepatic venous system. J. Pediatr. Surg. 2012. V. 47. No. 8. P. 1490–1495.
14. *Schaeffer D. F., Laiq S., Jang H. J., John R., Adeyi O. A.* Abernethy malformation type II with nephrotic syndrome and other multisystemic presentation: an illustrative case for understanding pathogenesis of extrahepatic complication of congenital portosystemic shunt. Hum. Pathol. 2013. V. 44. No. 3. P. 432–437.
15. *Sokollik C., Bandsma R. H., Gana J. C., van den Heuvel M., Ling S. C.* Congenital portosystemic shunt: characterization of a multisystem disease. J. Pediatr. Gastroenterol. Nutr. 2013. V. 56. No. 6. P. 675–681.
16. *Sood V., Khanna R., Alam S., Rawat D., Bhatnagar S., Rastogi A.* Ductal paucity and Warkany syndrome in a patient with congenital extrahepaticportocaval shunt. World. J. Hepatol. 2014. V. 27. No. 5. P. 358–362.
17. *Talwalkar J. A., Swanson K. L., Krowka M. J., Andrews J. C., Kamath P. S.* Prevalence of spontaneous portosystemic shunts in patients with portopulmonary hypertension and effect on treatment. Gastroenterology. 2011. V. 141. No. 5. P. 1673–1679.
18. *Witjes C. D., Ijzermans J. N., Vonk Noordegraaf A., Tran T. K.* Management strategy after diagnosis of Abernethy malformation: a case report. J. Med. Case Rep. 2012. V. 28. No. 6. P. 167.
19. *Yi J. E., Jung H. O., Youn H. J., Choi J. Y., Chun H. J., Lee J. Y.* A case of pulmonary arterial hypertension associated with congenital extrahepaticportocaval shunt. J. Korean. Med. Sci. 2014. V. 29. No. 4. P. 604–608.

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