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Pediatric hepatocellular adenoma: Benign hepatic tumors that may transform

Scheers I., Stephenne X., Smets F., Komuta M., Reding R., Sempoux C., Sokal E.

Abstract

Objectives and study: Hepatocellular adenoma (HCA) is a benign tumor occurring in a non cirrhotic liver. HCA may be symptomatic due to hemorrhage, compression or malignant transformation; however, the tumor is mostly found incidentally. In children, HCA develops in patients with underlying diseases such as MODY3, portosystemic shunt, glycogenosis or sexual hormone imbalance. In adults, HCA can be subclassified according to their immunophenotype and genotype. However exceptionally described, HCA might complicate with malignant transformation. Risk factors for transformation are poorly understood in pediatrics. We aimed to investigate the immunophenotype and natural evolution of HCA in pediatric patients. We further searched predictors of malignant transformation according to tumor immunophenotype. Finally, we assessed the usefulness of alpha-fetoprotein and imaging techniques in early diagnosis of HCC. Methods: Our retrospective, single center study included 13 children with HCA. Clinical, biochemical, imaging and histologic characteristics were reviewed on admittance and during follow-up. Results: Thirteen patients were followed for HCA of whom 2 girls and 11 boys. Seven had multiple lesions. Eight HCA were discovered during routine ultrasound follow-up of the underlying liver disease, 4 by chance and 1 had an enlarged liver on physical examination. All patients presenting HCA had an underlying risk factor for developing adenomas: 1 was diagnosed with MODY3, 4 with glycogenosis, 7 with portosystemic shunts or intrahepatic vascular anomalies and 1 had Mc Cune Albright. HCA could be categorized into 4 distinct groups after immunophenotyping: inflammatory (n=1), beta-catenin mutated (B-HCA, n=2), HNF1alpha mutated (n=4) and unclassified adenoma (n=1); three of the tumors had a combined inflammatory and B-HCA phenotype. HCA transformed in hepatocellular carcinoma (HCC) in three patients, at a mean age of 19 years (range 11-24 years). All these patients had a B-HCA immunophenotype and only one of them had a high alpha-fetoprotein. Ultrasound was poorly sensitive to early

diagnose HCC. Contrast enhanced MRI was the most accurate imaging modality to distinguish HCA from HCC. Conclusion: HCA in children could be classified according to immunophenotype and may transform. Children with beta-catenin mutated HCA are the most at risk to evolve to hepatocellular carcinoma. Contrast MRI imaging is recommended to follow-up HCA as it is the most sensitive technique, and neither alpha-fetoprotein and ultrasound are sufficiently accurate to formally rule out HCC.

Drug terms

alpha fetoprotein beta catenin sex hormone

Disease terms

liver tumor **adenoma** malignant transformation neoplasm glycogen storage disease liver cell carcinoma congenital blood vessel malformation liver disease benign neoplasm hepatomegaly diseases endocrine disease bleeding

Other terms

European **society** **gastroenterology** **nutrition** human immunophenotyping patient child imaging follow up ultrasound portosystemic anastomosis risk factor physical examination boy liver girl early diagnosis pediatrics nuclear magnetic resonance imaging compression genotype adult male phenotype risk female

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Author addresses

Correspondence address

Scheers I.

Clinique Universitaire St Luc, Pediatric Gastroenterology, Hepatology and Nutrition, Brussels, Belgium

Author addresses

Scheers I., Sokal E.

Clinique Universitaire St Luc, Pediatric Gastroenterology, Hepatology and Nutrition, Brussels, Belgium

Stephenne X.

Cliniques Universitaires St Luc, Pediatric Gastroenterology and Hepatology, Brussels, Belgium

Smets F.

UCL, Cliniques Universitaires Saint-Luc, Irec, Pediatrics, Brussels, Belgium

Komuta M.

Clinique Universitaire St Luc, Pathology, Brussels, Belgium

Reding R.

Clinique Universitaire St Luc, Pediatric Liver Transplantation and Surgery, Brussels, Belgium

Sempoux C.

Centre Hospitalier, Universitaire Vaudois, Pathology, Lausanne, Switzerland