IMAGE OF THE MONTH

Peripheral Arrangement of Steatosis Microvacuoles in Wilson's Disease

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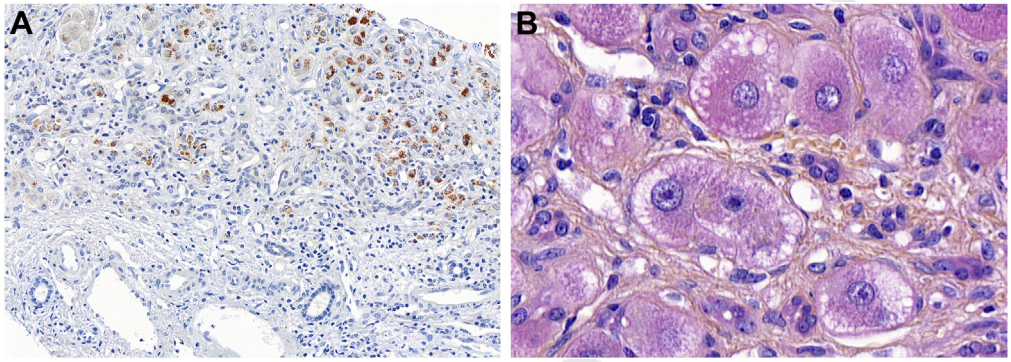

A 17-year-old girl was referred for jaundice and severe liver failure (international normalized ratio, 2.4; V, 19%, and total bilirubin level, 85 μmol/L). Transaminase and GGT levels were discretely increased. There was hemolytic anemia with a negative Coombs test and thrombocytopenia. The liver was dysmorphic with splenomegaly. There was no viral hepatitis; the IgG level reached 21 g/L without autoantibodies. Her urinary copper level was 1779 μg/24 hours (N $<$ 0.6), ceruloplasmin level was 0.12 g/L (N $>$ 0.2), and a high exchangeable copper to total copper ratio (23.4%; N $<$ 8.0) and hepatic copper level (844 μg/g) supported the diagnosis of Wilson's disease. A transjugular liver biopsy specimen showed cirrhosis with moderate periseptal activity (lymphocytes without a significant plasma cell contingent). Microvacuolar steatosis, anisokaryosis, glycogenated nuclei, and abundant hepatocellular deposits of copper (HES staining) were present in the nodules, confirmed by red deposits after rhodanine staining (Figure *A*). Strikingly, the steatotic microvacuoles were arranged at the periphery of hepatocytes, along the cytoplasmic membrane (Figure *B*).

An early diagnosis of Wilson's disease can be difficult and based on a variety of arguments. Its anatomopathologic features are not very specific and rhodanine staining may be absent. As a reference center for Wilson's disease, we have observed this peripheral arrangement of steatosis microvacuoles on liver samples from most of our patients.

Conflicts of interest
The authors disclose no conflicts.

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