

Question 43 – June 1

Which of the following is true regarding the histologic pattern seen in patients with Wilson disease?

- A. The liver biopsy may show classic histological features of autoimmune hepatitis.
- B. Cirrhosis is usually micronodular but occasionally macronodular.
- C. Typically patients with Wilson disease and acute liver failure have marked hepatocellular degeneration and parenchymal collapse without evidence of cirrhosis.
- D. In the early stages of the disease, copper is mainly seen in lysosomes but then mainly in the cytoplasm in the later stages of the disease.
- E. The absence of histologically identifiable copper has a good predictive value for screening for Wilson disease.

Answer: A

The liver biopsy may show classic histological features of autoimmune hepatitis. With progressive parenchymal damage, fibrosis and subsequently cirrhosis develop. Cirrhosis is frequently found in most patients by the second decade of life. It is usually macronodular, although occasionally micronodular. In the setting of acute liver failure due to WD, there is marked hepatocellular degeneration and parenchymal collapse, typically on the background of cirrhosis. Apoptosis of hepatocytes is a prominent feature with acute liver failure due to WD. In early stages of disease, copper is mainly in the cytoplasm bound to metallothionein and is not histochemically detectable; in later stages, copper is found predominantly in lysosomes. The absence of histochemically identifiable copper does not exclude WD, and this test has a poor predictive value for screening for WD. Thus, of all the choices, choice A is correct.