EDITOR'S QUIZ: GI SNAPSHOT

Familial liver disease

Robin Spiller, *editor***Clinical presentation**

A 52-year-old man presented with malaise and abnormal liver function tests: alanine aminotransferase 71 IU/ml (0–50), γ -glutamyltransferase 465 IU/ml (0–60), alkaline phosphatase 167 IU/ml (30–140), bilirubin 19 μ mol (3–28). He had never consumed alcohol, was on no regular drug treatment and had no medical history of note. His 53-year-old brother had died of renal disease and his sister had presented with fatal portal hypertensive haemorrhage at the age of 41 years. Clinical examination was unremarkable, full blood count Hb 12.5 g/dl (13.5–17.5), white blood count 2.5×10^9 (4–11), platelets 81×10^9 /l (150–400), albumin, coagulation profile and immunoglobulins were normal. Viral markers for hepatitis and autoimmune serology were negative, abdominal ultrasound

Figure 1 Liver biopsy, H&E stain, low-power microscopy.

Figure 2 Liver biopsy, H&E stain, high-power microscopy.

examination showed coarse architecture of the liver with normal-sized spleen. Oesophagogastroduodenoscopy showed grade IV oesophageal varices, a liver biopsy was carried out (figs 1 and 2).

Question

What is the diagnosis?

See page 732 for answer

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