A great mimicker of primary biliary cholangitis

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Introduction

A 59 year-old man without past medical history was referred with biochemical features of cholestasis (aspartate aminotransferase (AST) 117 U/L, alanine aminotransferase (ALT) 83 U/L, gamma-glutamyl transferase (GGT) 1307 U/L, alkaline phosphatase (AP) 1803 U/L, total bilirubin 0,7 mg/dL), a strongly positive anti-mitochondrial M2 antibody (AMA-M2) titer (88 U), weight loss and abdominal pain since several months. He did not take any medications, nor there was a history of alcohol abuse or sexual risk behavior. Upon presentation, clinical examination showed a rather cachectic patient (body mass index 19 kg/m²), without further abnormalities. As primary biliary cholangitis (PBC) was suspected, treatment with ursodeoxycholic acid had been initiated but did not improve the cholestasis nor the pain. Additional investigations didn't reveal any other irregularities, except for a mildly dilated aortic sinus root of 42 mm. The histopathological findings of a liver biopsy are shown below (Figure 1). This showed granulomas with cholangitis and increased presence of neutrophils, which raised suspicion for an infectious cause.

Question

Which infectious agent is re-emerging in human society and can mimic PBC?

Answer

Additional serology revealed a strongly reactive treponemal antibody test (TT) (signal-to-cutoff ratio 18,77), a negative rapid plasma regain test (RPR) and a reactive confirmatory TT. The patient was treated with benzathine benzylpenicillin 2,4 MIU given at weekly intervals during three weeks, which resulted in quick resolution of the long-lasting pain and significant reduction of liver biochemistry (AST 41 U/L, ALT 31 U/L, GGT 427 U/L, AP 881 U/L, total bilirubin 0,6 mg/dL on day 60 after treatment). Clinical improvement combined with serology and pathology lead to the final diagnosis of tertiary syphilis with hepatic involvement.

Syphilis is currently re-emerging at high speed (1). Liver involvement is rather uncommon and frequently overlooked as a cause of liver function abnormalities. The differential diagnosis with other causes of cholestatic liver injury is challenging as symptoms and signs are often non-specific (2). The presence of AMA-M2, usually highly specific for PBC, can be positive due to mimicry (3). A reactive TT is considered necessary for diagnosis, whereas a RPR can be non-reactive in long-standing untreated disease (1). Rapid remission of clinical and biochemical hallmarks after treatment is considered an important additional diagnostic clue (2).

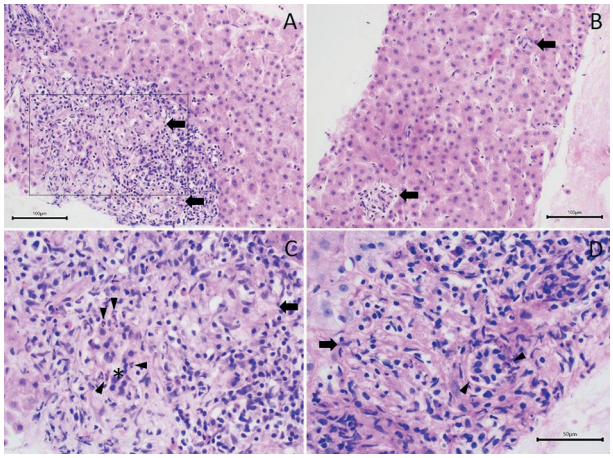

Figure 1. — Liver biopsy: A. Portal tract inflammation with lymphocytes, rare plasma cells, several eosinophils and neutrophils and non-necrotizing epithelioid granulomas (arrows) and very scant lobular lymphocytic infiltration (hematoxylin and eosin stain (H&E)). B. Lobular micro-granulomas (arrows) (H&E). C. (Higher magnification of inset in A) Portal tract with granuloma (arrow) and bile duct (asterisk in center of bile duct) with slightly damaged epithelium and several neutrophils (arrowheads) surrounding cholangiocytes (H&E). D. Portal tract with granuloma and bile duct (arrow) with sparse neutrophils located within the basement membrane (PAS-diastase).

Conflicts of Interest

The authors declare no conflict of interest.

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Submission date : 16/12/2020
Acceptance date : 14/01/2021