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# Effective treatment of autoimmune hepatitis-primary biliary cholangitis overlap syndrome with obeticholic acid

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*Dear Editor,*

We present the case of a 43-year-old female who attended our clinic due to abdominal pain, asthenia and pruritus. Laboratory profile tests showed GGT 263 U/l, ALK Ph 253 UI/l, IgG 2,060 mg/dl, positive for ANA and anti-Sp100. A liver biopsy showed PBC (stage 2 fibrosis) and ursodeoxycholic acid (UDCA) was started, without a response according to the Paris II criteria, so fibrates were added. Liver enzymes did not improve and laboratory test showed anti-Ro52 antibodies and ANA 1/1280. A new liver biopsy identified autoimmune hepatitis-primary biliary cholangitis (AIH-PBC) overlap syndrome with moderate-severe interface hepatitis.

Due to the AIH component, corticosteroids and azathioprine were added without improvement. Therefore, both treatments and fibrate were discontinued. Obeticholic acid (OCA) was started as a rescue therapy. An improvement was observed after two weeks. The dose was increased seven months later. This improvement is still being maintained after 15 months (Fig. 1). The tolerance was excellent, with occasional pruritis. Cholesterol levels (LDL, HDL) and IgG decreased. The control transient elastography at 15 months showed stage 2 fibrosis.

**Discussion**

Our patient presented a definitive diagnosis (22 points) according to the new scoring classification for PBC-AIH overlap syndrome (1). This syndrome predisposes to a major risk of liver failure and progression to cirrhosis (2,3) and therefore, an effective treatment is crucial to increase survival.

Currently, there is no consensus treatment for patients with AIH-PBC overlap syndrome who do not respond to the conventional therapy. In our case, OCA was initiated due to a PBC dominant component (4). The patient showed improvement, with a progressive reduction in liver enzymes and IgG. No fibrosis progression (5) or adverse effects during follow-up were observed.

To the best of our knowledge, this case is the first report of AIH-PBC overlap syndrome with a predominant PBC component that was effectively treated with OCA. More studies are required to define the role of this therapy in this group of patients.

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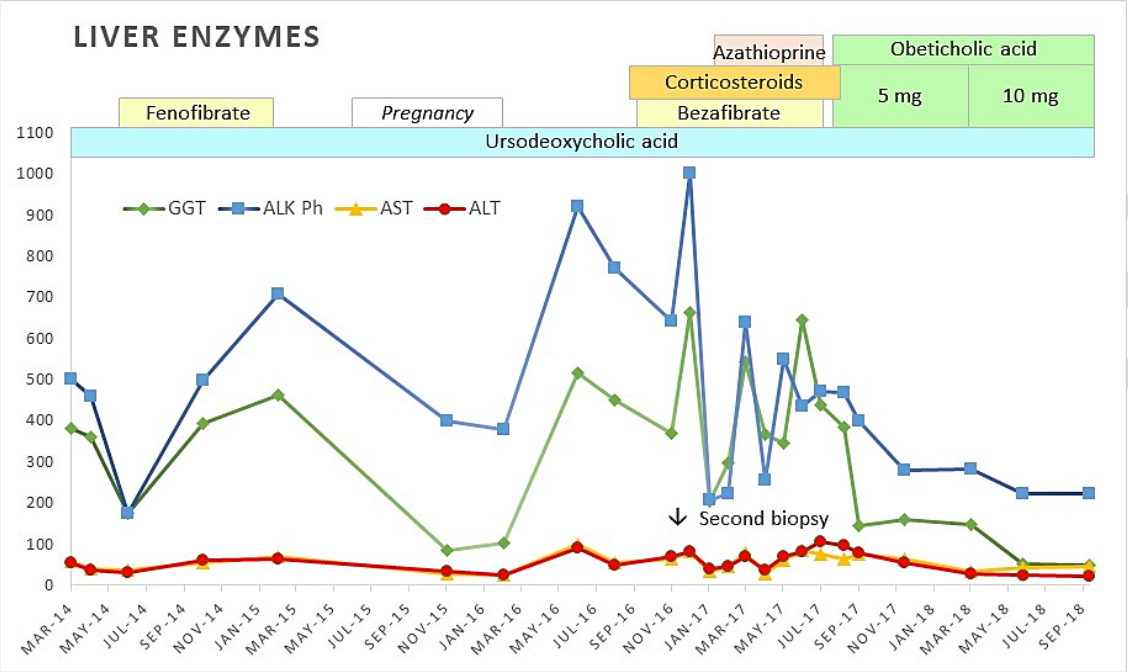


Fig. 1. Liver enzymes and treatment received during patient follow-up. Ursodeoxycholic acid (15 mg/kg/day) was prescribed with fenofibrate (160 mg/day) for eleven months and bezafibrate (400 mg/day) for ten months, with pregnancy between both treatments. During pregnancy, there was an improvement in the liver enzymes with a worsening after delivery (November 2015). Afterwards, she received corticosteroids, initially budesonide (9 mg/day) and then methylprednisolone (initial dose of 50 mg/day) following a liver biopsy result. Azathioprine was added (maximum dose of 2 mg/kg/day) along with methylprednisolone. Due to a lack of response, bezafibrate and azathioprine were suspended and corticosteroids were gradually reduced. Obeticholic acid was started at 5 mg/day for seven months and then increased to 10 mg/day until 15 months of follow-up with an excellent response.