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PICTURES IN DIGESTIVE PATHOLOGY

Glycogenic hepatopathy: A rare and reversible cause of elevated transaminases in diabetic patients. Case report

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BACKGROUND

Glycogenic hepatopathy (GH) is a quite unknown cause of elevated transaminases in type 1 diabetic patients with poor glycemic control characterized by glycogen overload hepatocytes.

CASE REPORT

We report a 28-year-old female, previously diagnosed long lasting type 1 diabetes, treated with insulin and with badly controlled glucose levels (Hb A1, 10.5 %).

She was sent to our service because of abdominal pain and altered liver function tests. She was not referring toxic habits, other drugs consumption or other interesting facts.

Examination showed weight 54 kg, height of 1.64 cm and less than 80 cm (BMI 20) abdominal perimeter. Painful hepatomegaly (3-4 cm) on palpation was noted, without any other interesting changes.

Laboratory tests showed a significant alteration of serum transaminases with AST 1600 IU/l, ALT 534 IU/l, FA 44 IU/l, GGT 661 IU/l with normal bilirubin and other liver function paramethers. In addition, she presented slight hypertriglyceridemia (250 mg/dl) and elevation (200 mg/dl) of LDL cholesterol with normal HDL (66 mg/dl). Ultrasound observed hepatomegaly and an increase in liver echogenicity suggesting steatosis.

Complete study of liver disease was made with negative results, ruling out the existence of entities such as viral hepatitis, autoimmune, hemochromatosis, alpha 1 antitrypsin deficiency, and Wilson's disease. The most likely differential diagnosis was established between glycogenic liver disease and non-alcoholic steatohepatitis.

Therefore, it was decided to perform an ultrasound-guided liver biopsy for an accurate diagnosis.

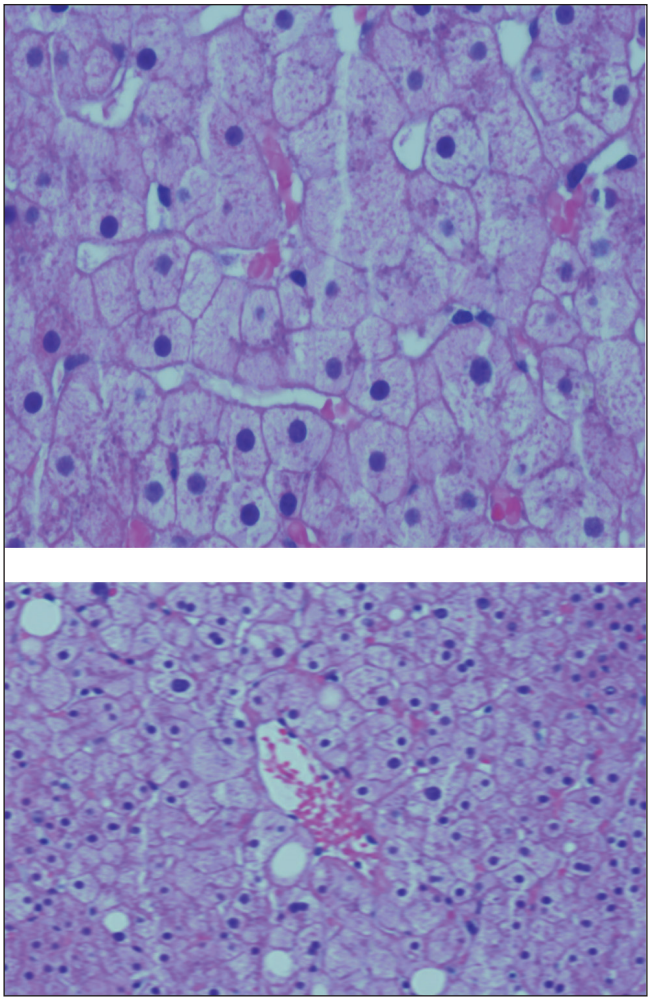

Fig. 1. Hematoxylin-eosin staining shows pal enlarged hepatocytes with sinusoidal compression. Mild steatosis with no fibrosis.

Histology showed pale appearance hepatocytes, swelling of cytoplasm with sinusoids compression and nucleus with glycogen in a large number of them (Fig. 1). Furthermore, it was described the presence of mild steatosis without necrosis,

inflammation and structural damage. Periodic Acid-Schiff staining (PAS) showed abundant intracytoplasmatic glycogen (Fig. 2). All the facts described above were consistent with the diagnosis of liver glycogen.

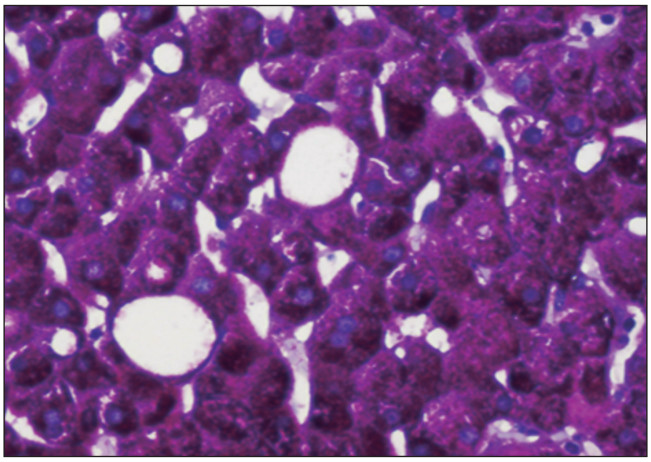

Fig. 2. Periodic acid-schiff (PAS) staining shows glycogen overload in hepatocytes.

DISCUSSION

Glycogen liver disease is an infra-diagnosed pathology, associated with poor glycemic controlled type 1 diabetes and characterized by glycogen overload in hepatocytes (1).

It usually appears with abnormal transaminases, quite relevant in our case, hepatomegaly and abdominal pain. It is a benign and reversible entity if good glycemic control is achieved; so it is important to distinguish it from other entities such as non-alcoholic steatohepatitis because treatment and prognosis are very different.

Although liver biopsy is the only way to confirm the diagnosis definitively, clinical and laboratory improvement might be enough in the rutinary practice.

Glycogenic liver disease was first described by Mauriac in 1930 (2). The syndrome was originally diagnosed in children with poor control of their diabetes and included hepatomegaly, Cushing-like face, short height, delayed sexual development and hyperlipidemia associated with glycogen accumulation. Afterwards, more cases have been reported in adults without these abnormalities, as occurs in our case (3-5). Moreover, some cases with acute abdomen requiring urgent laparotomy have been reported (6).

In a published report, 13 out of 14 patients type 1 diabetes patients whose liver biopsy was compatible with glycogen liver disease had impaired function test (1). The prevalence of this condition is unknown.

In glycogenic hepatopathy, insulin presence and high levels of glucose increase liver's glycogen load. So glycogenic-synthase-phosphatase is activated. It dephosphorylates and activates glycogen synthase, which is the enzyme required to transform glucose 1-phosphate into glycogen. This activation stimulates glycogen storage in the liver resulting in a blockade of further glycogenolysis (7).

It is important to distinguish this entity from fatty liver disease associated with metabolic syndrome because different prognosis and treatment. In the last one, disease progression can cause inflammation and sometimes fibrosis with development of nonalcoholic steatohepatitis resulting in a more difficult treatment and in an uncertain prognosis (8). On the other hand, glycogenic hepatopathy has a good prognosis as long as adequate glycemic control is achieved. As being previously described, alterations were totally reversible in our patient in which an acceptable glycemic control was reached and, subsequently, transaminases were normalized.

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