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An underrecognized cause of hepatomegaly in uncontrolled type I diabetes

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Introduction

A 29-year-old woman with a past medical history of poorly controlled type 1 diabetes and autoimmune hypothyroidism presented to the emergency department for evaluation of abdominal pain, nausea and vomiting. The patient was a cocaine user, but she did not use alcohol and did not smoke. Her current medications included Insulin aspart, Insulin degludec, L-thyroxine and Pantoprazole. She reported no use of over-the-counter medication or herbal and dietary supplements. There was no family history of diabetes, autoimmune diseases, or liver diseases.

Her vital signs included temperature 36,3°C; heart rate 99 bpm; respiratory rate 16 pm and blood pressure 92/58 mmHg. She was alert and oriented. Physical examination was notable for a thin, anicteric woman who was not in acute distress. The abdomen was nondistended and soft with diffusely tenderness and a smooth palpable liver 3 to 4 cm below the costal margin.

Laboratory studies was significant for glucose 539 mg/dL (normal 65-110), alkaline phosphatase 153 U/L (normal 35-105), gamma-GT 44 U/L (normal <36), lactate 42,1 mg/dL (normal 4,5-19,8), arterial pH 7,32 (normal 7,35-7,45) and HbA1c 11% (4,5-6).

A computed tomography (figure 1; panel A and B) and liver biopsy (figure 1; panel C and D) were performed.

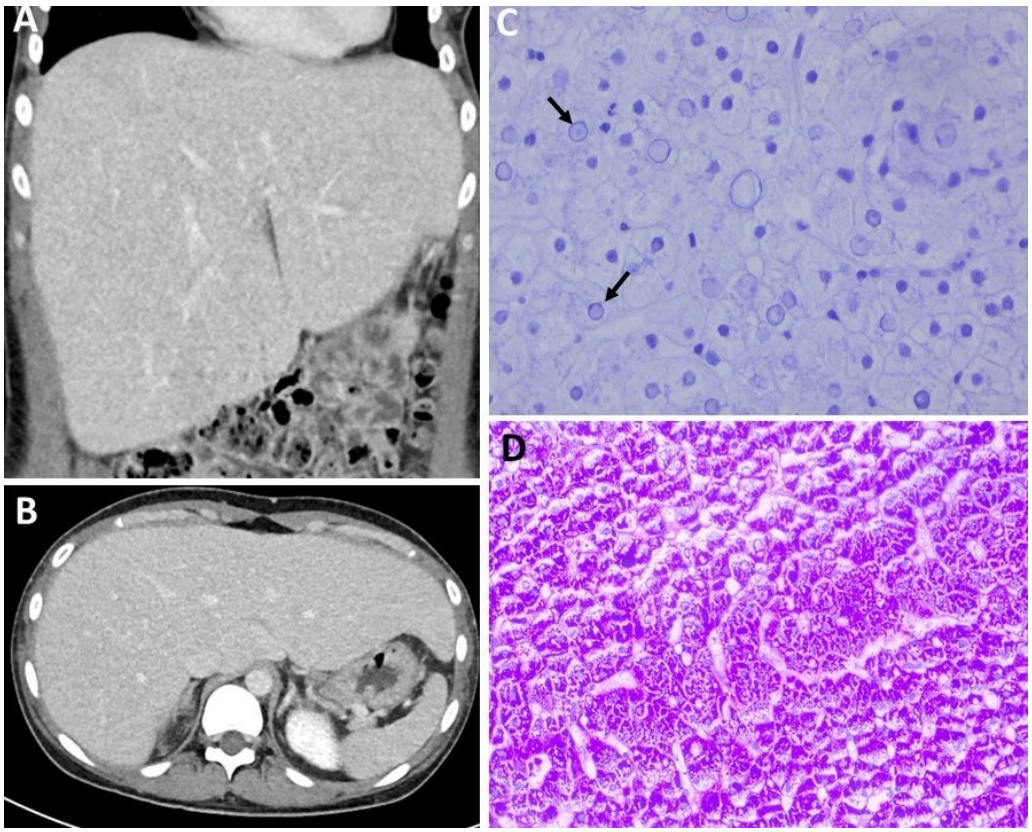

Figure 1. — (Panel A): Coronal section of the Computed Tomography. (Panel B): Cross-sectional Computed Tomo-graphy. (Panel C): Liver-biopsy specimen; Hematoxylin-and-eosin staining – arrow showing glycogenated nuclei. (Panel D): Liver-biopsy specimen; Periodic acid-Schiff staining.

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Question

What is the clinical problem?

Answer

Coronal section of the Computed Tomography (Panel A) showed a severe hepatomegaly with a midclavicular craniocaudal diameter of 18 cm. Cross-sectional Computed Tomography (panel B) showed a severe hepatomegaly reaching deep into the left hemi-abdomen.

A Hematoxylin-and-eosin (HE) staining on liver-biopsy specimen (Panel C) revealed glycogenated nuclei (arrows) and diffusely swollen hepatocytes with pale cytoplasm. Periodic acid-Schiff (PAS) stain (Panel D) shows hepatocytes strongly and diffusely positive, suggesting that the hepatocytes were swollen with glycogen, consistent with a diagnosis of glycogenic hepatopathy. There were no signs of non-alcoholic steatohepatitis, autoimmune or drug-induced hepatitis.

The patient was treated with aggressive intravenous fluid resuscitation and insulin therapy with improvement of the abdominal pain, lactate and blood glucose level after 24 hours.

This case calls attention to glycogenic hepatopathy as an underrecognized cause of hepatomegaly in patients with poorly controlled type 1 diabetes (1-3). Clinical symptoms may include abdominal pain, nausea, vomiting and hepatomegaly (1-3). The key histological findings on HE staining are swollen hepatocytes and on PAS staining glycogen accumulation in the hepatocytes, causing hepatomegaly and elevated liver enzymes (1-3).

The cornerstone of treatment is glycaemic control. Glycogenic hepatopathy is a reversible condition with an excellent prognosis once glycaemia is controlled (2).

Keywords: Glycogenic hepatopathy, hepatomegaly, type 1 diabetes.

Conflict of interest

The authors have no potential conflict of interest relevant to this clinical image to be reported.

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