Visual Vignette

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Case Presentation: A 25-year-old woman with a 6-year history of diabetes that was well controlled on sulphonylurea therapy with a hemoglobin A1c level of 6.5% presented with sticky stools, muscle aches, lower limb weakness, and a 5-kg weight loss over 1 year. She also had a 5-year history of intermittent abdominal pain. Her body mass index was 17.5 kg/m^2, her blood pressure was normal, and there was diffuse skin darkening without clinical features of insulin resistance. She appeared lipoatrophic and had fat malabsorption with a 72-hour stool fat level of 41 g (normal <18 g), and hemogram, liver, and renal function tests and abdomen ultrasonography were normal. Abdomen magnetic resonance imaging (MRI; A, T1-weighted; B, fat-suppression) is shown in Figure 1. There was no dilatation of intrahepatic biliary radicals or the common bile duct. **What is the diagnosis?**

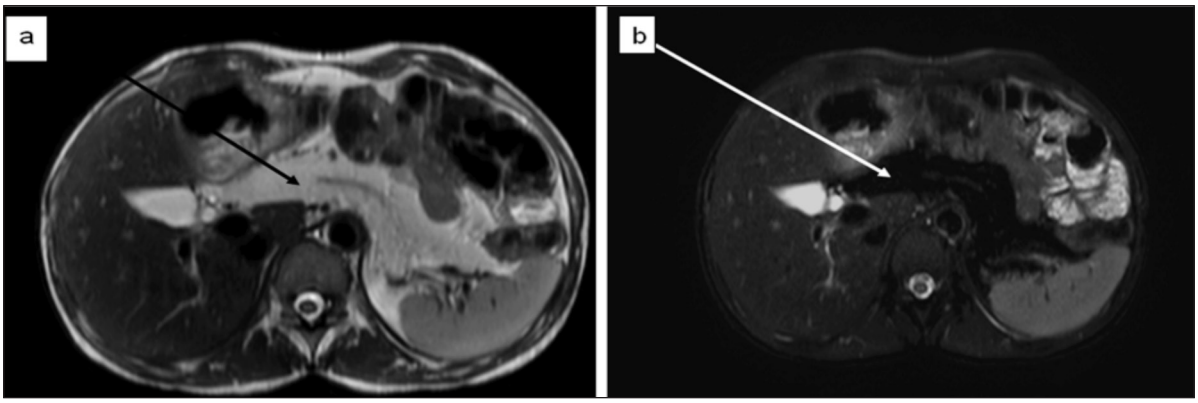

Fig. 1

Answer: Total pancreatic lipomatosis or lipomatous pseudohypertrophy of the pancreas. She was treated with pancreatic enzyme supplementation and glimepiride. Her weight increased by 5 kg in 6 months. Hemochromatosis, adrenal insufficiency, and vitamin B12 deficiency were ruled out with biochemical tests: serum ferritin, 41.2 ng/mL (10 to 290); vitamin B12, 731 pg/ml (200 to 950); and 8:00 AM serum cortisol, 11.79 mcg/dL (5 to 23). She had a family history of diabetes; her mother and sister were diagnosed in their 40s. The possibility of maturity onset diabetes of the young (MODY) was considered but was ruled out given the family member age of onset, and her clinical presentation and anatomical findings. Lipomatous pseudohypertrophy of the pancreas or total pancreatic lipomatosis is a rare disorder characterized by the replacement of pancreatic exocrine tissue with adipose tissue, although the pancreatic duct and islets remain intact (1). Initially described by Hantelmann as early as 1903, the specific etiology remains unknown. Several predisposing factors have been suggested, namely obesity, diabetes, and age-related pancreatic fat infiltration (2). However, imaging usually reveals remnant pancreatic parenchyma with uneven fat infiltration, which was different from the uniform fatty replacement in the present case. On the basis of the imaging findings, a diagnosis of diffuse pancreatic lipomatosis leading to fat malabsorption was made. The role of sonology was limited. Chemical shift MRI has an advantage over computed tomography in confirming focal fatty replacement of the pancreas (3). Pancreatic enzyme supplementation in combination with dietary counseling is the mainstay of therapy. Clinicians should consider total pancreatic lipomatosis as a possible differential diagnosis in patients with malabsorption. Abdomen MRI can reliably exclude the disease.

DISCLOSURE

The authors have no multiplicity of interest to disclose.

REFERENCES

1. **Yasuda M, Niina Y, Uchida M, et al.** A case of lipomatous pseudohypertrophy of the pancreas diagnosed by typical imaging. *JOP*. 2010;11:385-388.
2. **Olsen TS.** Lipomatosis of the pancreas in autopsy material and its relation to age and overweight. *Acta Pathol Microbiol Scand A*. 1978;86A:367-373.
3. **Kim HJ, Byun JH, Park SH, et al**. Focal fatty replacement of the pancreas: Usefulness of chemical shift MRI. *AJR Am J Roentgenol*. 2007;188:429-432.

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