Insulinoma and extreme obesity in a patient with MEN1 syndrome

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To the Editor,

We report a patient aged 16 experiencing headache, galactorrhea and ammenorrhea. Laboratory examination showed an increased prolactin (PRL) level (> 400 ng/mL ; normal range 5-30 ng/mL) and computed tomography (CT) of pituitary gland revealed a tumor mass, 1.5 × 0.9 cm in size. Transnasal selective partial adenomectomy was performed, and postoperative excisional biopsy showed a prolactinoma. Postoperatively, she developed hypopituitarism, for which levothyroxin (100 μg/day) and hydrocortison (30 mg/day) replacement therapy were prescribed. Further laboratory tests showed decreased glucose levels (3.0 mmol/L ; normal range 3.5-6.5 mmol/L), never completely explained, because the patient left the hospital on her demand. She returned to our department at the age of 21, with long-standing symptoms of weakness, sweating, tremor and mental confusion. She was 150 cm high and had 158 kg (90 kg more than at previous hospitalization), BMI 70.2 kg/m^2. Laboratory examination revealed persistent hypoglycemia (2.5-3.1 mmol/L), hyperinsulinemia (105 pmol/L ; normal range < 5 pmol/L) and increased C-peptide level (95 μg/L ; normal range 1.1-5.0 μg/L). The serum levels of IGF-1, cortisol, vanillylmandelic acid, metanephrines, parathormone, PRL, thyroid stimulating hormone, growth hormone, follicle-stimulating hormone and luteinizing hormone were within normal limits. Abdominal ultrasonography revealed a tumor mass in the distal part of the pancreatic tail, 2.6 cm in size (Fig. 1).

We failed to perform abdominal CT and magnetic resonance imaging (MRI) due to patient's enormous obesity. Other methods were also excluded, and surgical procedure was indicated. Partial pancreatectomy (body and tail of the pancreas) and splenectomy (due to vascular irrigation) were carried out. Histopathological examination confirmed an insulinoma. Clinical symptoms resolveded after the operation, but she developed transient postoperative diabetes and she was treated with insulin for the following nine months. On the last testing, the laboratory results were within normal ranges and the patient felt quite well.

Furthermore, one year after the operation, body weight gradually decreased ; she lost 54 kg, with normal findings on postoperative MRI and CT scans.

The described case of the patient with MEN1 syndrome (prolactinoma and insulinoma) illustrates how unattended symptoms, with consecutive excessive weight gain, can be a diagnostic challenge for a clinician. Patients with insulinomas are frequently asymptomatic before onset of episodes of sudden confusion or unconsciousness recurrent over years and becoming more frequent. When laboratory testing establishes the diagnosis of insulinoma, localizing procedures should be performed. Depending on tumor localization, pancreaticoduodenectomy or partial (usually distal) pancreatectomy may be performed, as well as total pancreatectomy, if a single tumor cannot be found or multiple lesions exist (1). Insulinomas are, in most cases, identified at the initial exploration. A second exploration, using palpation and intraoperative ultrasound, bears more than 90% chance of identifying a resectable lesion. However, surgery carries a risk of inducing pancreatitis and other postoperative complications. Patients with a presumptive diagnosis should therefore be referred to a center for evaluation by experienced physicians prior to surgery (2). The goal of medical therapy is to prevent

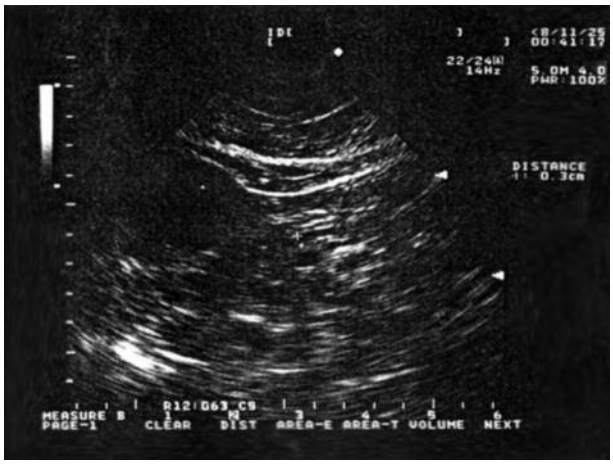

Fig. 1. — Abdominal ultrasonography imaging. Tumor size was 2.6 cm localised in the distal part of pancreatic tail.

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symptomatic hypoglycemia. Two drugs that inhibit insulin secretion are available, diazoxide and octreotide ; both have side effects, and a consultation with an experienced physican is recommended (3,4,5).

It can be concluded that appropriate preoperative evaluation in an experienced centre and thorough exploration of the pancreas during the surgery, are very important for the success of the treatment of insulinoma.

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