Comment

Female aged 20 with known T1D from the age of 3, currently with normal weight (weight 55.300 kg, height 153 cm, BMI 23.6, waist perimeter 83 cm) under treatment with diet, scheduled physical exercise and insulin NPH 28-14-18 IU/day, with regular insulin adjustments. The patient attended the clinic consultation due to dyslipidemia and a regular glycemic control, with self-monitoring between 200 and 220 mg/dL. As history, the patient referred multiple admissions due to recurrent serious hypoglycemia and diabetic ketoacidosis.

On physical examination, full moon facies stood out as well as rosy cheeks, increased distribution of fat in neck, trunk and abdomen, graceful limbs, and hepatomegaly of 2-3 cm. The following results were found in the performed analytical: hematocrits 35%, leukocytes 5,100/mm³ (neutrophils 80% eosinophils 1%, lymphocytes 15%, monocytes 4%, basophils 0%), fasting glycemia 89 mg/dL, HbA1c 6.91%, creatinine 6.8 mg/dL, total cholesterol 425 mg/dL, triglycerides 476 mg/dL, proteinuria 0.27 g/24 h, creatinuria 0.59 g/24 h (0.90-1.50), creatinine clearance 60 mL/min (75-135), TSH 0.83 mIU/L (0.5-4.5), T4L 1.1 ng/L (1.4 ng/100 mL) and negative anti-TPO antibodies. Other hormonal studies performed were: GH 3.5 ng/mL (3.3 ± 2.8), prolactin 9.2 ng/mL (10.2 ± 4.8) and cortisol 16.2 μg/100 mL (16.8 ± 5.9). Moreover, the presence of celiac disease was ruled out with negative IgA endomysial antibodies, antitransglutaminase-IgA antibodies 1 IU/mL (negative <4) and serum IgA levels of 117.400 mg/dL (90-300). The gynecological echography showed the presence of uterus and ovaries (right 20 × 11 mm, left 15 × 7 mm). A bone age of 15 years was determined through radiography of the carpus (left hand). An intensification of the insulin treatment was performed and treatment with simvastatin 10 mg + ezetimibe 10 mg (0-0-1) and enalapril 5 mg (1-0-1) was started.

The syndrome of Mauriac appears in children with inadequate controlled diabetes. At present, it is not so much observed, due to the improvement in the insulin treatment in this group of patients. The syndrome is characterized by the full moon face, short height, increase of the abdominal perimeter associated to hepatomegaly, infiltration of fat and glycogen and proximal muscular atrophy. Moreover, a limited joint mobility has been described associated to tense skin and waxy skin, osteopeny, growth impairment and delay of the bone maturation. The impairments of the joint mobility have been associated to the early development of diabetic microvascular complications, as the retinopathy and the nephropathy that might appear even before the age of 18.

References