MAURIAC SYNDROME, A RARE COMPLICATION OF TYPE 1 DIABETES MELLITUS

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BACKGROUND

Mauriac syndrome (MS) classically involves hepatomegaly (hepatic glycogenosis), growth impairment, delayed puberty and cushingoid features in a patient with poorly controlled type 1 diabetes mellitus (T1DM). The typical age of presentation is adolescence.

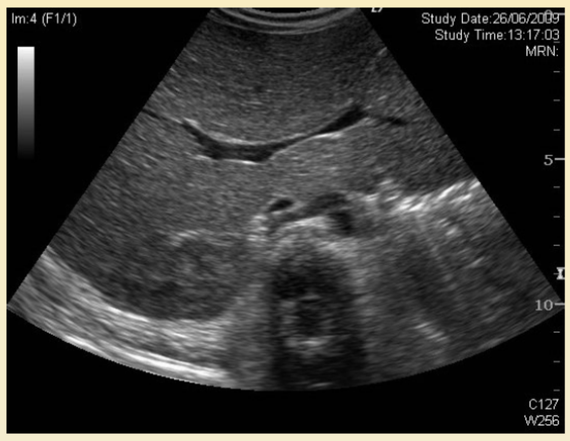
There is an accumulation of glycogen in the hepatocyte caused, in part, by prolonged periods iof hyperglycaemia, where glucose passes into the hepatocyte independent of insulin, folowed by periods of insulin treatment which mediatrws the conversion of the entrapped glucose to glycogen.

Case reports and small case series have been reported. With the advent of improved insulin regimens, MS has seen less frequently. However, new cases appear each year in medical literature.

CLINICAL CASE

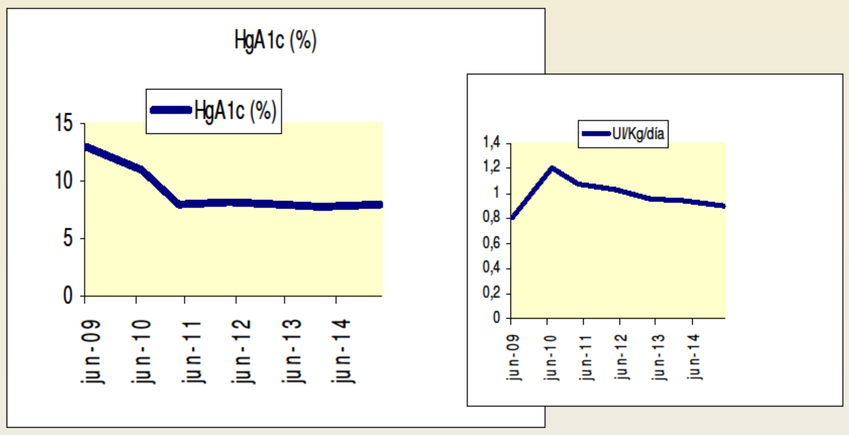
We report the case of a 9 years and 9 months of age male, with T1DM onset al 20 months of age. During the last 2 years he hadn't had medical control of T1DM; he was receiving treatment with NPH human insulin and short-acting insulin analogue (lispro), total dose: 0,8 UI/Kg/day.

Physical examination: Weight: 22.7 kg (-1.5 SD). Height: 117 cm (-3.58 SD). Rounded face. Abdominal swelling, liver 3 cm below RCM, he had no splenomegaly Lypodistrophia in both arms. His target height was 165 ± 5 cm (p3, -1.95 SD).

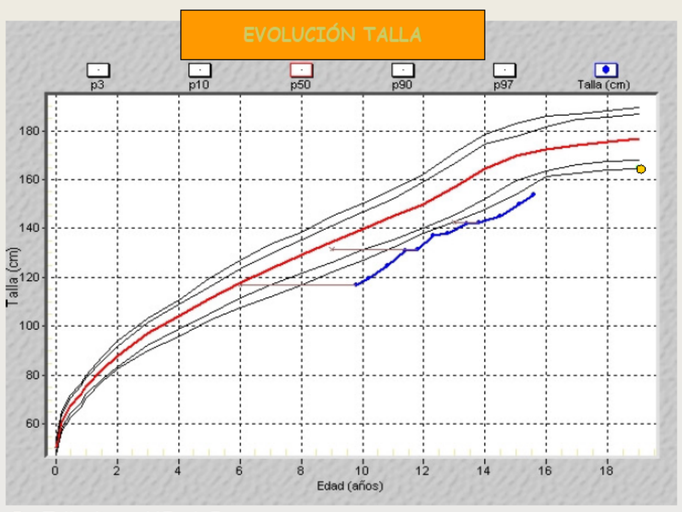


Abdominal ultrasound: diffuse hepatomegaly with homogeneous echogenicity. No intrahepatic focal lesions, normal-sized gallbladder with thin and smooth walls.

Hemoglobin A1C 13,1%; fasting glucose 302 mg/dl; triglycerides 156 mg/dl; AST 43UI/L; ALT 51 UI/L. No microalbuminuria. Bone age: 6 years (delayed; - 4 - 5 SD). Treatment and diabetologic training were intensified.



5 years later his insuline regimen is glargine once a day and lispro at breakfast, lunch, night tea and dinner; total dosis: 0.9 UI/Kg/day, and Hemoglobin A1C: 7.8% - 8%;



Age: 14.5 years

Physical examination: Weight: 53.8 kg (-0.82 SD). Height: 152 cm (-2.63 SD), growth rate: 7.6 cm/y (+3.6SD). Normal phenotype, no goiter, No abdominal masses or hepatomegaly. Pubertal stage: Tanner III (Right teste of 12 cm3 and left 10 cm3; adrenarche II-III. Left arm and left thigh lypodistrophia.

Bone age according to chronological age (BA: 14y CA: 15y)

CONCLUSIONS

Despite improvements in the therapeutic treatment of type 1 diabetes mellitus, Mauriac syndrome continues to appear in cases of poor control, even before puberty. Most of the clinical findings are reversible with a good metabolic control.

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