MAURIAC SYNDROME, A RARE COMPLICATION OF TYPE 1 DIABETES MELLITUS

Rivero Martín Mª José, Pérez Segura Mª Pilar, Alcázar Villar Mª José,
Montes Bentura David, Oros Milian Mª Eugenia
Department of Pediatrics, University Hospital of Fuenlabrada, Madrid, España

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 of hyperglycaemia, where glucose passes into the hepatocyte independent of insulin, followed by periods of insulin treatment which mediates 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 at 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 U/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 Lyphodistrophia in both arms. His target height was 165 ± 5 cm (p3, -1.95 SD).

Hemoglobin A1C 13.1%; fasting glucose 302 mg/dl; triglycerides 156 mg/dl; AST 43 U/L; ALT 51 U/L. No microalbuminuria. Bone age: 6 years (delayed; -4 to -5 SD). Treatment and diabetologic training were intensified.

5 years later his insulin regimen is glargine once a day and lispro at breakfast, lunch, night tea and dinner, total dose: 0.9 U/kg/day, and Hemoglobin A1C: 7.8% - 8%.

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.