Acta Scientific Women's Health (ISSN: 2582-3205)

Volume 6 Issue 5 May 2024

Review Article

An Uncommon Case Report of Mauric Syndrome or Diabetic Glycogenosis in Girl

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Received: April 10, 2024

Published: April 18, 2024

Abstract

Mauriac syndrome is an uncommon side effect of poorly controlled adolescent diabetes mellitus. We present the case of a 17-year-old patient with type 1 diabetes who was admitted to the hospital because of a serious hyper glycemic imbalance. A failure to thrive and hepatomegaly were noted during the clinical assessment. Biological investigations revealed anicteric cholestasis, severe cytolysis, and hyperglycemia. The foundation of the care was provided by insulin and hydration therapies. The etiological examination for liver dysfunction produced negative results. Biology and medicine have positively evolved. The diagnosis of hepatic glycogenesis was upheld in light of several anamnestic and clinical evidences, as well as the absence of further abnormalities linked to hepatic disorders. The outcome was elevated blood sugar. Blood transaminases were also increased. An abdominal ultrasound indicated homogeneous hepatomegaly. Viral hepatitis serology and immunological testing came back negative, and a liver biopsy confirmed the hepatocyte ballooning. Given the favorable outcomes observed during high-dose insulin treatment, the diagnosis of Mauriac syndrome was retained.

Keywords: Mauriac Syndrome; Hepatic Glycogenosis; Diabetes Mellitus; Failure to Thrive; Hepatomegaly

Introduction

Inadequate management of diabetes is a major issue in poor countries. Hepatocyte glycogen overload, or Mauriac syndrome, is one of the many complications linked to insulin deficiency that it exposes patients to [1]. Mauriac described the syndrome for the first time in 1930. Babies with type 1 diabetes mellitus who have significant hyperglycemia followed by high insulin dosages have a rare condition [2]. In reality, type 1 diabetes in young adults usually only manifests as hepatomegaly with increased liver enzymes, leaving the diagnosis incomplete. Type 2 diabetes is commonly linked to nonalcoholic steatohepatitis (NASH), a disorder that is commonly misdiagnosed or confused for the latter alterations. The exact incidence of Mauriac syndrome is uncertain due to the limited number of cases that have been reported in the literature [4]. Here, we present the case of a young patient with Mauriac syndrome and diabetes.

Observation

The patient, who was 17 years old and had type 1 diabetes for three years, was admitted to the endocrinology department for treatment of an unbalanced type 1 diabetes and staturo-ponderal delay. This unplanned diagnosis of non-severe acute cytolytic hepatitis happened by chance. Clinically, the patient was anicteric, cognizant, in good overall health, hepatomegaly with a 16-centimeter hepatic arrow, delayed in weight and height, and impuberty. The patient has extremely balanced diabetes (HbA1c 14%), and transaminases (ASAT 3481 (81*N), ALAT 1331 (31*N), GGT: 1 484,0 UI/l, PAL: 191,0 UI/l, BT: 11,0 mg/l, BD: 8 mg/l) were more than ten times normal with anicteric cholestasis. Hemoglobin, platelets, and leukocytes were all correct with an abnormal hemostasis test (PT 100%); the hemogram revealed no abnormalities on the three lines. The liver appeared on abdominal ultrasonography to have a homogenous echo structure that had grown in volume, was non-dilated, VSH-permeable, and had a permeable portal trunk. The immunological assessment, which looked for antinuclear antibodies, anti KLM 1, anti smooth muscle, anti mithochondria M2, anti sp100, anti SLA, and anti gp 210, was normal. The viral serologies were negative (Ac anti HVA IgM:negative, Ac anti HbcIgM: negative, Ag anti Hbs: negative, and Ac anti Hbs: positive (vaccinated)). Dysthyroidism and celiac disease were ruled out. A liver biopsyre-vealed no histological cholestasis or fibrosis, normal liver architecture, and hepatocyte ballooning with indications of hepatocyte regeneration devoid of necrosis or inflammatory infiltration.

The evolution after correction of the glycemic figures under basal-bolus diet was favorable.

Discussion

Mauriac syndrome (MS) is a rare complication of type 1 diabetes mellitus (DM1). It is related to low insulin concentrations and is less common since longer-acting insulins became available. It is characterized by hepatomegaly, growth and puberty delay, and the presence of elevated transaminases and serum lipids. Mauriac syndrome is characterized by dwarfism, obesity and hepatomegaly in patients with insulin-dependent diabetes mellitus. It is associated with poor control of type 1 diabetes mellitus (T1DM) in adolescents, and may present as obesity, hepatomegaly, cushingoid facies and elevated transaminases. It is typically associated with growth failure and delayed pubertal maturation, which should alert the physician over insufficient management of diabetes mellitus and the related development of Mauriac syndrome, although these can be reversed with good glycemic control. This young child with type I diabetes and extremely poor glucose control presents a clinical picture of impuberty, hepatomegaly, and delayed weight and height. Hepatic cytolysis is linked to this clinical condition. There were multiple hepatic-focused supplementary exams conducted. All of the usual symptoms returned, with the exception of hepatomegaly. We further stress that there is a positive clinico-biological evolution following well-executed insulin therapy. Hepatic glycogenosis, or secondary glycogenosis, is less well-described in the literature but is frequently seen in type 1 diabetes yet goes undiagnosed [5]. Its pathogenesis is not fully understood. It appears to be connected to both the excess insulin and the periods of hyperglycemia. Insulin activates glucokinase and glycogen synthetase and inhibits glucose-6-phosphatase, resulting in excessive storage of circulating glucose as intrahepatic glycogen through hyperstimulation of glycogenesis and inhibition of glycogenolysis. These mechanisms contribute to hepatic glycogenosis in cases of insulin overuse associated with hyperglycemic phases [6]. The etiology of pubertal delay and growth retardation is unclear, however it seems to be multifaceted. Possible contributing factors include hypercorticism, inadequate tissue glucose, and faulty insulin as a growth factor. Children's cushingoid symptoms are traditionally characterized in relation to glycogenosis [7,8]. Hepatic glycogenosis should be as little as possible in type I diabetic patients with better glycemic control. Hepatic glycogenosis is totally reversible with adequate metabolic management, in contrast to NASH [9,10]. Complete remission of clinical, biochemical, and histological problems can result with appropriate glucose and insulin management [11].

Conclusion

When hepatomegaly with staturopunderal delay is found in a type 1 diabetic, hepatic glycogenosis must be suspected, but this is still an elimination diagnosis. It is critical to rule out autoimmune, metabolic, obstructive, and viral reason before adopting it. Because of glycemic management, the evolution is typically good and the hepatic involvement goes away in two to four weeks.

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