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Case Report

Diabetic Glycogenosis or Mauriac Syndrome: A Case Report

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Abstract

The Mauriac syndrome is a rare complication of poorly controlled diabetes mellitus in adolescence. We report the case of a 17-year-old patient with type 1 diabetes who got admitted for severe hyperglycemic imbalance. The clinical examination showed failure to thrive, hepatomegaly. Biological analyses showed hyperglycaemia, major cytolysis, and anicteric cholestasis. The management was based on insulin and fluid therapy. The etiological investigation of the liver dysfunctions was negative. Clinical and biological evolution was favorable. The diagnosis of hepatic glycogenesis was retained based on a bundle of anamnestic and clinical arguments and the absence of other abnormalities causing by hepatic disorders. Blood sugar was very high. Blood transaminases were also high. Abdominal ultrasound showed homogeneous hepatomegaly. Viral hepatitis serology, immunological tests were negative, liver biopsy was in favor of hepatocyte ballooning. Given the favorable evolution under intensive insulin therapy, the diagnosis of Mauriac syndrome was retained.

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

Introduction

Poorly controlled diabetes represents a major concern in underdeveloped countries. It exposes to many complications related to insulin deficiency, and hepatocyte glycogen overload, previously referred to as Mauriac syndrome, is one of them [1]. Mauriac first described the syndrome in 1930. It is a rare syndrome described in children with type 1 diabetes mellitus in which significant hyperglycemia is followed by the administration of high doses of insulin [2]. In young adults with type 1 diabetes, the syndrome is not complete and in fact only hepatomegaly with increased liver enzymes is often present. The latter alterations are often unrecognized or confused with hepatic steatosis or nonalcoholic steatohepatitis (NASH), which is common in type 2 diabetes [3]. The exact incidence of Mauriac syndrome is uncertain because there are only a few cases reported in the literature [4]. Here we report a case of Mauriac syndrome in a young diabetic.

Observation

The patient was K.Z, 17 years old, type 1 diabetes for 3 years; she was hospitalized in the endocrinology department for the management of a staturo-ponderal delay and an unbalanced type I diabetes with fortuitous discovery during the exploration of a non severe acute cytolytic hepatitis. Clinically, the patient was conscious, in good general condition, anicteric, with a delay in weight and height, impuberty and hepatomegaly with a hepatic arrow at 16 cm. Diabetes was very unbalanced (HbA1c 14%), transaminases were more than 10 times normal with anicteric cholestasis (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).

The hemogram showed no abnormalities on the 3 lines; hemoglobin, platelets, leukocytes were correct with an abnormal hemostasis test (prothrombin level 100%).

Abdominal ultrasonography showed a liver of homogeneous echostructure increased in volume, VSH permeable non-dilated with permeable portal trunk.

Viral serologies were negative (Ac anti HVA IgM: negative, Ac anti Hbc IgM: negative, Ac anti Hbs: positive (vaccinated), Ag anti hbs: negative and the immunological assessment in search of antinuclear antibodies, anti KLM 1, anti-smooth muscle, anti-mitochondria M2, anti sp100, anti SLA, anti gp210 was normal.

Celiac disease and dysthyroidism were ruled out.

A liver biopsy showed normal liver architecture, hepatocyte ballooning with signs of hepatocyte regeneration without necrosis or inflammatory infiltrate, no histological cholestasis, no fibrosis.

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

Age	17 years
Duration of diabetes	14 years
HbA1c %	14 %
ASAT	81*N
ALAT	31*N
PAL	191,0 UI/l
GGT	1484,0 UI/l
Total Bilirubin	11,0 mg/l
Prothrombin time	100 %
Autoimmunity test Antinuclear antibodies Anti smooth muscle antibodies Anti-mitochondrial antibodies Anti LKM1 Anti gp210 Anti sp100 Anti SLA	Negative
Viral serologies	Negative
TSH	Normal
AC anti transglutaminase type IGA and IGG	Négative
Hepatomegaly	Homogenous with liver arrow at 16 cm
Liver biopsy	Hepatocyte ballooning, without signs of cholestasis

Table 1: *Summary of the results of the paraclinical examinations.*

Discussion

The observation of this young type I diabetic patient with very poor glycemic control describes a clinical picture of delayed weight and height, impuberty and hepatomegaly. This clinical syndrome is associated with hepatic cytolysis. Several complementary examinations focused on the liver were performed. They came back without any particularities apart from the hepatomegaly.

And we emphasize that after a well conducted insulin therapy, a favorable clinico-biological evolution. Secondary glycogenosis [hepatic glycogenosis] is less described in the literature, but it can be frequently observed and unrecognized in type 1 diabetes [5]. Its pathophysiology is imperfectly known. It seems to be related to the combined excess of insulin and hyperglycemic episodes. The mechanisms that contribute to hepatic glycogenosis in cases of insulin overuse associated with hyperglycemic phases consist of excessive storage of circulating glucose as intrahepatic glycogen by hyperstimulation of glycogenesis and inhibition of glycogenolysis; insulin activates glucokinase and glycogen synthetase and inhibits glucose-6-phosphatase [6]. The pathogenesis of growth retardation and pubertal delay is not clear, but rather appears to be multifactorial: insufficient tissue glucose, defective insulin as a growth factor, and hypercorticism may contribute. The cushingoid signs present during glycogenosis are classically described in children [7,8]. The better the glycemic control of type I diabetic patients, the more minimal hepatic glycogenosis should be. Unlike NASH, hepatic glycogenosis is completely reversible with good metabolic control [9,10]. Appropriate management of glucose and insulin levels can lead to complete remission of clinical, biological and histological abnormalities [11].

Conclusion

Hepatic glycogenosis must be evoked in front of the discovery of hepatomegaly with staturopunderal delay in a type 1 diabetic but it remains a diagnosis of elimination. Before accepting it, it is imperative to eliminate viral, metabolic, obstructive and autoimmune causes. The evolution is most often favorable thanks to glycemic control, and the hepatic involvement usually disappears in two to four weeks.

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