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Review Article

Glycogenic Hepatopathy in Children with Type 1 Diabetes: About 10 Cases

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Introduction

Acquired glycogenic liver disease (GH) is a rare disease that develops due to excessive accumulation of glycogen in hepatocytes, linked to high sugar intake requiring and very high insulin requirements leading to overload hepatomegaly in patients. Type 1 diabetic patients described for the first time in 1930 by Mauriac as part of a syndrome comprising growth retardation and delayed puberty in unbalanced diabetic children. Recent case reports have demonstrated that GH may be the only feature of Mauriac syndrome, GH is a benign and reversible disease with a good prognosis, unlike non-alcoholic fatty liver disease (NAFLD), although GH is well individualized, it remains underdiagnosed.

The objective(s) of the study:

In this study, we describe the clinical, biochemical and histopathological characteristics concerning 10 cases of diabetic children presenting with GH.

Methods

Retrospective descriptive study between 2019-2020 of 10 type 1 diabetic children presenting GH.

Results

The 10 patients had a median age of 13.5 years and a duration of diabetes of around 6 years. Diabetes mellitus control was poor and hemoglobin A1c averaged 12.7%. Insulin requirements exceeding 1.5 IU/KG/D in all patients. Half of the cases had a history of diabetic ketoacidosis. All patients presented hypoglycemic episodes, nausea and vomiting, abdominal distension and pain, lunar facies and edema, hepatomegaly was significant with a hepatic flexure of more than 10 cm in all patients, stature delay was not constant found in half of the patients. Laboratory examinations showed a moderate elevation of liver enzymes, an elevation of lactate, viral serologies were negative, autoimmunity assessment negative in all patients, the diagnosis time GH ranged between one and two years after the onset of the above GH symptoms, suggesting that it is underdiagnosed. 5 patients progressed well after dietary monitoring with reduction of insulin doses and improvement in liver enzyme levels and a progressive reduction in hepatic arrow.

Discussion

The discussion of hepatomegaly in these diabetic children led us to discuss autoimmune hepatitis taking into account the terrain of autoimmunity, hepatic steatosis, hepatic glycogenosis was suspected on a set of anamnestic arguments, clinico-biological confirmed by histology.

Conclusion

GH must be sought in any poorly controlled diabetic child in order to avoid progression towards irreversible stature delay which is Mauriac syndrome, even if the evolution of GH has a good prognosis without hepatic after-effects, this goal expected remains difficult to achieve given the difficulty of the treatment which aims to reduce.