SAS Journal of Medicine
Abbreviated Key Title: SAS J Med
ISSN 2454-5112
Journal homepage: https://saspublishers.com

OPEN ACCESS

Endocrinology

Mauriac Syndrome: A Rare Complication of Type 1 Diabetes Mellitus

H. Boualam[1*], S. Bammou[1], S.Rafi[1], G. El Mghari[1], N.El Ansari[1]

[1]Department of Endocrinology, Diabetes, Metabolic Diseases and Nutrition, Mohammed VI University Hospital of Marrakech
[1]Faculty of Medicine and Pharmacy of Marrakech, Cadi Ayyad University, Marrakech, Morocco

DOI: 10.36347/sasjm.2023.v09i05.005 | **Received:** 11.02.2023 | **Accepted:** 25.03.2023 | **Published:** 05.05.2023

***Corresponding author:** H. Boualam
Department of Endocrinology, Diabetes, Metabolic Diseases and Nutrition, Mohammed VI University Hospital of Marrakech

Abstract **Case Report**

The Mauriac syndrome is a rare complication of poorly controlled diabetes mellitus in adolescence. It is characterized by hepatomegaly, growth and puberty delay, and the presence of elevated transaminases and serum lipids. We report the case of a 17-years-old patient with type 1 diabetes who got admitted for evaluation of growth retardation. The clinical examination showed failure to thrive, hepatomegaly and abdominal distension. Blood sugar was very high. Blood transaminases were also high. Abdominal ultrasound showed homogeneous hepatomegaly. Viral hepatitis serologies and autoimmune study were negative. Liver histology analysis after liver biopsy comfirmed the hepatic glycogenosis. After glycaemic control was improved, liver enzymes normalized and the adbominal distension regressed.

Keywords: Mauriac Syndrome, Hepatic Glycogenosis, Type 1 Diabetes Mellitus.

INTRODUCTION

Mauriac syndrome (MS) is a rare complication of type 1 diabetes mellitus (DM1), characterized by hepatomegaly (hepatic glycogenosis), puberty and growth delay, dyslipidemia, transaminase elevation and reduction of IGF1 (insulin-like growth factor 1) [1]. Cushingoid features may also be present [2].

MS is more common in children and adolescents with poor glycemic control and increased susceptibility of complications [1], and is the commonest cause of hepatic dysfunction in children and adolescents with DM1 [3]. We present a case of Mauriac syndrome in a 17-year-old male patient.

CASE REPORT

A 17-years-old Type 1 diabetic boy was admitted for evaluation of growth retardation. He was diagnosed to have T1DM 15 years back. The patient maintained poor glycemic control since childhood, presenting an elevated glycated hemoglobin rate persistently higher than 10% and recurrent episodes of ketosis. Examination showed that height was 131 cm (less than 3rd percentile), weight 28 kg (less than 3rd percentile) and body mass index of 16.3. Tanner stage was G2P1. He had abdominal distension with hepatomegaly (figure1).

Investigations showed HbA1c 15,2% and IGF-1 65,7 ng/ml N (131-490). Bone age was 13 years. Liver function test showed AST 514 IU/l, ALT 277 IU/l and total bilirubin 3 mg/l. His renal function was normal. Viral hepatitis serologies and autoimmune study were negative. Ultrasound abdomen revealed hepatomegaly with a liver span of 15 cm. A liver biopsy revealed numerous hepatocytes with glycogenated nuclei, abundant cytoplasmic and nuclear glycogen deposits, and moderate portal fibrosis.

Based on the clinical history and investigations, the final diagnosis of Mauriac syndrome was made. Strict diabetic dietary management was started along with regular monitoring of blood sugar level. Long acting insulin Glargine injection was administered once daily along with regular insulin injection before breakfast, lunch and dinner (basal-bolus regimen). He was followed-up for 2 months. He had shown a reduction in hepatomegaly, a normalization of transaminase concentrations with a decrease of HbA1c concentration (from 15,2% to 11%).

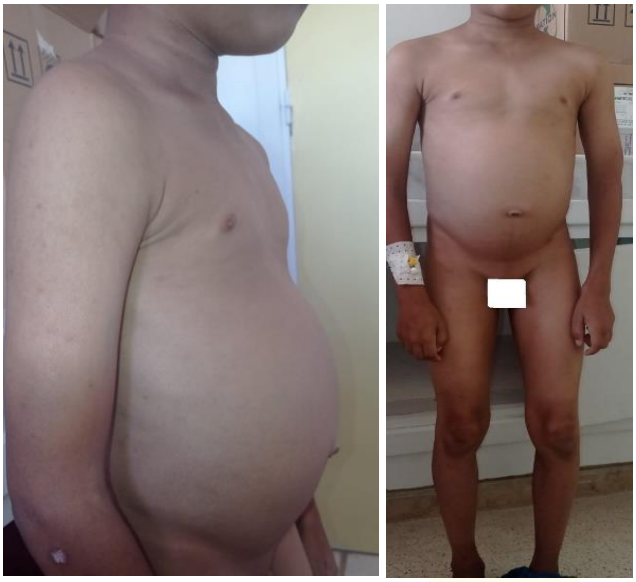

Figure 1 : abdominal distension, Short stature

DISCUSSION

Mauriac in 1930, described growth failure and maturational delay with hepatomegaly and abdominal distension in children with T1DM, who were treated with short-acting insulin [4]. In the late 1930s Joslin clinic reported a case series of 60 youngsters with hepatomegaly, growth failure, delayed sexual maturation, and severe uncontrollable diabetes [5]. Equal incidence is reported in males and females, with most of the cases occurring during adolescence [6]. MS can occur in any age if good metabolic control is not achieved [2]. With better control of sugar, the incidence of this syndrome has reduced rapidly and in the current era this is a very rare syndrome.

The pathogenesis of growth retardation is not clear but is thought to be multifactorial. Inadequate glucose to the tissues, decreased Insulin-like growth factor-1 and GH levels, hypercortisolism, resistant or defective hormone receptor action contribute to stunted growth and delay in puberty [6]. Hepatomegaly is a typical sign of Mauriac syndrome and appears in the majority of affected patients [7,8], its cause is thought to be due to the deposition of glycogen in the liver [9], and similar subcutaneous deposition gives rise to the round moon like facies [10]. In addition to glycogen accumulation, steatosis with varying degree of fibrosis may be evident in liver biopsy [11].

It is imperative to exclude other causes of hepatomegaly and elevated transaminase levels, including autoimmune hepatitis, celiac disease, viral hepatitis, hemochromatosis, and Wilson disease [12]. Growth failure, delayed puberty and hepatomegaly in Mauriac's syndrome improves with glycemic control [13].

CONCLUSION

Although Mauriac syndrome is rare, it should be still considered in Type 1 diabetic children with growth impairment and liver disease in order to achieve an earlier diagnosis of mild forms and timely insulin therapy adjustment.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study

REFERENCES

1. Maia, F. F., & Araújo, L. R. (2005). Pancreas transplantation in Mauriac Syndrome: clinical and biochemical parameters after one year follow up. *Arquivos Brasileiros de Endocrinologia & Metabologia*, *49*, 455-459.
2. Mahesh, S., Karp, R. J., Castells, S., & Quintos, J. B. Q. (2007). Mauriac syndrome in a 3-year-old boy. *Endocrine Practice*, *13*(1), 63-66.
3. Flotats Bastardas, M., Miserachs Barba, M., Ricart Cumeras, A., Clemente León, M., Gussinyer Canadell, M., Yeste Fernández, D., ... & Carrascosa Lezcano, A. (2007). Hepatomegalia por depósito de glucógeno hepático y diabetes mellitus tipo 1. *An. pediatr.(2003, Ed. impr.)*, 157-160.
4. Mauriac, P. (1930). Gros ventre, hepatomegalie, troubles de las croissance chez les enfants diabetiques traits depuis plusieurs annes par l'insuline. *Gax Hebd Med Bordeaux*, *26*, 402-410.
5. Marble, A., White, P., Bogan, I. K., & Smith, R. M. (1938). Enlargement of the liver in diabetic children: I. Its incidence, etiology and nature. *Archives of Internal Medicine*, *62*(5), 740-750.

6. Ferry Jr, R. J. (Ed.). (2011). *Management of pediatric obesity and diabetes*. Springer Science & Business Media.
7. Munns, C. F. J., McCrossin, R. B., Thomsett, M. J., & Batch, J. (2000). Hepatic glycogenosis: reversible hepatomegaly in type 1 diabetes. *Journal of paediatrics and child health*, *36*(5), 449-452.
8. Fitzpatrick, E., Cotoi, C., Quaglia, A., Sakellariou, S., Ford-Adams, M. E., & Hadzic, N. (2014). Hepatopathy of Mauriac syndrome: a retrospective review from a tertiary liver centre. *Archives of Disease in Childhood*, *99*(4), 354-357.
9. Lee, R. G., & Bode, H. H. (1977). Stunted growth and hepatomegaly in diabetes mellitus. *The Journal of Pediatrics*, *91*(1), 82-84.
10. Klein, R., Marks, J. F., Sherman, F. E., & Fetterman, G. H. (1962). The occurrence of peripheral edema and subcutaneous glycogen deposition following the initial treatment of diabetes mellitus in children. *The Journal of Pediatrics*, *60*(6), 807-814.
11. Ikarashi, Y., Kogiso, T., Hashimoto, E., Yamamoto, K., Kodama, K., Taniai, M., ... & Tokushige, K. (2017). Four cases of type 1 diabetes mellitus showing sharp serum transaminase increases and hepatomegaly due to glycogenic hepatopathy. *Hepatology Research*, *47*(3), E201-E209.
12. Sherigar, J. M., Castro, J. D., Yin, Y. M., Guss, D., Mohanty, S. R. (2018). Glycogenic hepatopathy: a narrative review. *World Journal of Hepatology*, *10*(2), 172–185.
13. Haller, M. J., Silverstein, J. H., Rosenbloom, A, L. (2007). Type 1 diabetes in the child and adolescent. In: Lifshitz F, editor. Pediatric Endocrinology. 5th ed. Vol. 1. Switzerland: Informa Healthcare;. p. 70.