International Journal of Medical and Pharmaceutical Case Reports
4(5): 117-120, 2015; Article no.IJMPCR.18813
ISSN: 2394-109X

SCIENCEDOMAIN *international*
www.sciencedomain.org

Mauriac Syndrome in a 35 Year Old Woman With Type One Diabetes Mellitus

Ferdane Sapmaz[1*], Sebahat Başyiğit[1], Ismail Hakki Kalkan[2], Sefa Güliter[2] and Işilay Kalan Sari[3]

[1]Department of Gastroenterology, Keçiören Education and Training Hospital, Ankara, Turkey.
[2]Department of Gastroenterology, Kırıkkale University Faculty of Medicine, Kırıkkale, Turkey.
[3]Department of Endocrinology, Kırıkkale University Faculty of Medicine, Kırıkkale, Turkey.

Authors' contributions

This work was carried out in collaboration between all authors. Author FS wrote the draft of the manuscript. Authors SB and IHK managed the literature searches. Author IKS, managed literature searches and contributed to the correction of the draft. Author SG provided the case and supervised the work. All authors read and approved the final manuscript.

Article Information

DOI: 10.9734/IJMPCR/2015/18813
Editor(s):
(1) Jignesh G. Patel, Department of Pathology, University of Texas Medical Branch at Galveston, Texas, USA.
Reviewers:
(1) Anonymous, University of Arkansas, USA.
(2) Parineeta Samant, Department of Biochemistry, MGM Medical College, Navi-Mumbai, India.
(3) A. Papazafiropoulou, Department of Internal Medicine and Diabetes Center, Tzaneio General Hospital of Piraeus, Greece.
Complete Peer review History: http://sciencedomain.org/review-history/9966

Case Study

Received 11th May 2015
Accepted 10th June 2015
Published 29th June 2015

ABSTRACT

Mauriac syndrome is a rare complication of type 1 diabetes mellitus. It is characterized by hepatomegaly, growth delay and the presence of elevated transaminases and serum lipids. Mauriac syndrome occurs in males and females equally, and is most common in adolescence, although there are reports in children as young as toddlers and in adults.
Here in we report a 35 year old type 1 diabetic patient was admitted to our clinic with elevated transaminases and hepatomegaly and diagnosed with Mauriac Syndrome.

Keywords: Mauriac syndrome; adult; type 1 diabetes mellitus; hepatomegaly.

**Corresponding author: E-mail: ferda-sapmaz@hotmail.com;*

1. INTRODUCTION

Mauriac syndrome is an uncommon syndrome associated with poor control of type 1 diabetes mellitus in children. It presents as obesity, cushingoid faces, hepatomegaly and elevated transaminases. It is typically associated with delayed pubertal maturation and growth failure. The incidence of this syndrome has decreased significantly with introduction of long acting insulin and better control of blood sugar [1]. Altough this syndrome is diagnosed in childhood, it had been reported cases in adults. Here in we report a 35 year old type 1 diabetic patient presented to clinic with elevated transaminases and hepatomegaly and which diagnosed with Mauriac Syndrome.

2. CASE REPORT

A 35 year old was referred to the gastroenterology clinic due to elevated liver enzymes noted during standard screening blood tests. She was diagnosed to have type 1 diabetes mellitus (DM) 20 years back and was put on premix insulin (70/30). She had poor control of diabetes with no regular follow- ups and she was hospitalized for diabetic ketoacidosis (DKA) for 5 times till date.

Physical examination showed that she was significantly short (143 cm), her weight was 38 kg. Her Body Mass Index (BMI) was 18.6 kg/m2. She had retarded growth and development. She had moon faces, cold skin and thin scalp hair. Liver was palpable clinically 6 cm below costal margin with no splenomegally. Laboratory investigation were ; hemoglobin 10.6 g/dl, blood glucose: 575 mg/dl, HBA1c:%13, alanine aminotransferase (ALT):311 U/L, aspartate aminotransferase (AST):205 U/L, alkaline phosphatase (ALP): 434 U/L, total protein: 6.5 g/dl, albumin: 3 g/dl, blood urea :36 mg/dl, serum creatinine :0.9 mg/dl, total cholesterol: 270 mg/dl, low density lipoprotein-cholesterol: 193 mg/dl. Her thyroid profile (free T3, free T4 and thyroid stimulating hormone) was within normal limits.

Ultrasonography revealed hepatomegaly and grade 2 fatty change of the liver. Bu she didn't have liver biopsy. Celiac disease was ruled out with endoscopy and anti transglutaminase negative. We evaluated other reasons of liver disfunction. Her test results were as follows: Hepatitis B Surface Antigen, HBsAg (-), antibody to hepatitis B immunoglobulin M, Anti HBc IgM (-), antibody to hepatitis C virus, Anti HCV (-), antibody to hepatitis A virus immunoglobulin M, Anti HAV IgM (-),antibody to hepatitis E virus, Anti HEV(-) antibody to cytomegalovirus, Anti CMV IgM (-), antibody to Epstein-Barr virus immunoglubluin M, Anti EBV IgM (-), venereal disease research laboratory, VDRL (-), antibody to herpes simplex virus immunoglobulin M, Anti HSV IgM (-), Anti rubella IgM (-), Anti toxo IgM (-), IgA, M, and G levels were normal, antinuclear antibody, ANA (-),anti-double stranded DNA, Anti dsDNA (-), Anti-mitochondrial antibodies, AMA (-), liver kidney microsomal antibody, LKMA (-), anti-smooth muscle antibody, ASMA (-), urine Legionella antigen (-), H1N1 PCR (-),seasonal influenza PCR(-), Brucella rose bengal and tube agglutination (-), and seruloplasmin level was normal. There was no history of alcoholism or primary malnutrion.

Based on history, examination and investigation findings, final diagnosis of Mauriac syndrome was made. Patient was shifted to strict dietary management and high dose premixed insulin. After 2 months of therapy, premeal sugar came down to 100 - 150 mg and HbA 1C: % 9 and there was reduction in liver function tests (ALT: 50 U/L, AST:45 U/L, ALP:150 U/L). Renal function tests were normal (blood urea :27 mg/dl, serum creatinine: 0.8 mg/dl). But there wasn't any reduction in hepatomegaly. Hepatomegaly often cannot be stretched after treatment. Therefore the cause of hepatomegaly has not been studied.

3. DISCUSSION

Mautiac syndrome is a condition which is characterized by hepatomegaly and growth failure in diabetic patients with poor metabolic control may be reversible with improvement of metabolic control is presented.

Mauriac syndrome was first described by Mauriac in 1930 in children with type 1 DM presenting with clinical features of growth failure, maturation delay, hepatomegaly and abdominal distension [2].

Table 1. Cases of adult onset Mauriac Syndrome

Patient	Sex	Age	Age of Type 1 DM diognosis	BMI (kg/m^2)	ALT (U/L)	HbA1c (mmol/mol)	Clinical presentation
1	F	20	3	17.2	1471	14.7	Nausea vomiting
2	F	26	5	N/A	158	12	Nausea, vomiting, abdominal pain
3	F	22	N/A	N/A	N/A	N/A	N/A

The features of Mauriac syndrome are mostly related with deficient insulinization. Hepatomegaly can be developed in patients due to intrahepatic glycogen deposition ; if these patients also have elevated liver enzymes, cushing features, dyslipidemia and delayed growth maturation, Mauriac syndrome can be diagnosed [3].

Two different forms of Mauriac syndrome was defined by the presence of obesity. The first form which is treated with regular insulin alone is associated Cushingoid obesity with secondery hyperadrenalism [4]. Mauriac syndrome has been reported in patients who are not obese and it occurs in patients who have given regular, under the dose insulin [5].

Mauriac syndrome is more common in children and adolescents with poor glycemic control and increased susceptibility of complications and is the commonest cause of hepatic dysfunction in patient with Type 1 DM. This is a case of adult female with poorly controlled DM.

Growth failure, hepatomegaly and elevated liver enzymes can reverse with improved glisemic control. But in our case despite liver function tests have been normalized, hepatomegaly didn't reverse.

With the additional findings of gross hepatomegaly, growth failure and elevated liver function tests , she was diagnosed with Mauriac syndrome. However children and adolescents are more vulnerable to this disease, Mauriac syndrome may be seen in adults [6]. There are only few cases were reported as Mauriac Syndrome during adulthood (Table.1) above. Mauriac syndrome should be considered in type 1 diabetic patients with growth failure and hepatomegaly.

4. CONCLUSION

Mauriac Syndrome is a rare complication of Type 1 DM. Although the disease was observed more frequently in children, may be seen in adults. With aggressive glycemic control, the manifestations of this syndrome can be reverted.

CONSENT

It is not applicable.

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

REFERENCES

1. Haller MJ, Silverstein JH, Rosenbloom AL. Type 1 diabetes in the child and adolescent. In: Lifshitz F, editor. Pediatric Endocrinology. 5th ed. Switzerland: Informa Healthcare. 2007;1:70.
2. Mauriac P. Gros ventre. Hepatomegalie, troubles de croissance chez les enfants diabetiques traites depuis plusiers annee par l'insuline. Gaz Hebd Med Bordeaux. 1930;26:402–10.
3. Flotats M, Miserachs M, Ricart A, Clemente M, Gussinyer M, Yetse D, et al. Hepatomegaly due to glycogen storage disease and type 1 diabetes mellitus. An Pediatr. 2007;67:157-60.
4. Gutch M, Philip R, Saran S, Tyagi R, Gupta KK. Re-emergence of a rare syndrome: A case of mauriac syndrome. Indian J Endocrinol Metab. 2013;17 (Suppl 1):S283-5.
5. Re-emergence of a rare syndrome: A case of mauriac syndrome. Gutch M, Philip R, Saran S, Tyagi R, Gupta KK. Indian J

Endocrinol Metab. 2013;17(Suppl 1):S283-5.

6. Madhu SV, Jain R, Kant S, Prakash V. Mauriac syndrome: A rare complication of type 1 diabetes mellitus. Indian J Endocrinol Metab. 2013;17(4):764-5. DOI:10.4103/2230-8210.113780.

© 2015 Sapmaz et al.; This is an Open Access article distributed under the terms of the Creative Commons Attribution License (http://creativecommons.org/licenses/by/4.0), which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.

Peer-review history:
The peer review history for this paper can be accessed here:
http://sciencedomain.org/review-history/9966