International Journal Dental and Medical Sciences Research
Volume 4, Issue 6, Nov-Dec 2022 pp 60-63 www.ijdmsrjournal.com ISSN: 2582-6018

Rare complication of Insulin dependent diabetes mellitus – Mauriac Syndrome

Dr. Prashanth Sondalagere Kapanaiah(1), Dr. Kurakula Rani (2), Dr. K Thulasiram(3).

(1) Postgraduate, General Medicine Department, Sri Venkateswara Medical College, Tirupathi.
(2) Postgraduate, General Medicine Department, Sri Venkateswara Medical College, Tirupathi.
(3) Professor, General Medicine Department, Sri Venkateswara Medical College, Tirupathi.

Submitted: 25-10-2022 Accepted: 04-11-2022

ABSTRACT
Mauriac syndrome is characterized by obesity, Abnormal lipid profile, growth failure, hepatomegaly and delayed pubertal maturation, nephropathy, retinopathy, cushingoid features such as moon facies, protruded abdomen in patients with Insulin-dependent diabetes mellitus (IDDM). It is associated with poor glycemic control. Currently the disease is a rare complication, but it is necessary to pay attention to the possibility of the existence of its individual characteristics. We hereby report a case of Mauriac syndrome in a 24year Male Patient.

Keywords: Mauriac Syndrome, Insulin dependent diabetes mellitus, rare complication, Poor glycaemic control, Hepatomegaly, Pubertal delay, Growth failure.

I. INTRODUCTION

Pierre Mauriac in 1930 described this syndrome[1] in children diagnosed as Insulin dependent diabetes mellitus(IDDM) who presented with classical features of growth failure, pubertal delay, protuberant abdomen, moon facies and hepatomegaly and who were treated with short-acting insulin with poor glycemic control[2]. Its incidence is equal in male and female population with adolescence being the most affected group[3].Its etiology is multifactorial such as decreased levels of insulin-like growth factor-1 (IGF-1) and growth hormone, defective or resistant hormone receptors or inadequate utilization of glucose in the tissues[4]. Hepatomegaly is thought to be due to glycogen deposition in the liver[5].Before the discovery of insulin, adolescents and young adults with diabetes had no chance of life. At that time, Mauriac syndrome was a common occurrence until the fifties and sixties of the last century[6-8]. The introduction of modern methods of insulin and glucose monitoring decisively reduced the frequency of occurrence of this syndrome. The incidence of Mauriac syndrome decreased dramatically, although it is still being reported. Often in these cases the patients present only some symptoms of this syndrome.A case of Mauriac Syndrome in a patient with poorly controlled IDDM has been presented in the following case report.

II. CASE REPORT

A 24year old male patient presented to emergency ward of SVRRGGH, Tirupathi with complaints of swelling of both lower limbs on and off since 6 months, complaints of visual disturbance since 3 months, complaints of decreased urine output of around 500ml/day since 10days, complaints of breathlessness since 7days associated with easy fatiguability, loss of appetite and generalized weakness. Patient was a known case of IDDM which was diagnosed at the age of 11yrs and on Insulin therapy, known case of systemic hypertension since 3months using antihypertensive medication. On examination patient had moon facies, underdeveloped secondary sexual characteristics tanner staging corresponds to stage 3, pallor present, bilateral pitting pedal edema extended up to knee joints, height 145cm, body weight 45kg, pulse 100bpm, regular, high volume, blood pressure 160/90mm Hg, temperature 37.6degree Celsius. Abdomen soft distended, non tender, no local raise of temperature, liver palpable with smooth margins(liver span=17cm) present, no evidence of peritoneal fluid and splenomegaly, On auscultation bilateral fine basal crepitations were present .No palmar erythema, spider angiomata, leg edema, petechia or purpura were found. On ophthalmic examination Bilateral moderate non-proliferative diabetic retinopathy(retinal hemorrhages, cotton wool spots) noted, Kayser Fleischer ring not found.

International Journal Dental and Medical Sciences Research
Volume 4, Issue 6, Nov-Dec 2022 pp 60-63 www.ijdmsrjournal.com ISSN: 2582-6018

Laboratory findings

Investigation	On day of admission	3rd day of admission	10th day of admission	Reference values
Hemoglobin(g%)	7.6	7.8	8.0	13-16
Total bilirubin(mg/dl)	0.8	0.9	0.7	0.2-1.2
Direct bilirubin(mg/dl)	0.1	0.2	0.2	<0.3
AST/ALT/(U/L)	167/184	160/130	145/130	1-40
ALP(IU/L)	186	220	200	30-120
Total protein(g/dl)	5.4	5.8	6	6.5-8
Serum Albumin(g/dl)	3.5	3.4	3.6	3.5-5.4
Serum Creatinine(mg/dl)	6.3	5.5	3.3	0.8-1.3
Blood urea(mg/dl)	128	106	63	17-45
Serum Na+/K+/Cl-(mEq/L)	137/5.4/97	138/5.2/95	142/4.8/102	135-145/3.5-5.5/96-110
RBS(mg/dl)	461	286	170	<200
HbA1c %		10.35		
Total cholesterol(mg/dl)	-------	230	210	110-199
Serum HDL(mg/dl)	-------	36	50	40-60
Serum LDL(mg/dl)	-----	150	200	62-128
24hrs Urinary protein	-----	3.2g/1000ml	------	-------
ABG=pH,HCO3 (mMol),CO2(mmHg)	7.2,16,35	7.3,18,40	7.4,20.6,40	7.35-7.45/22-26/35-45
Urine ketone bodies	Positive	-----	Negative	-----

CXR P/A view: bilateral lower zone inhomogeneous opacites, USG abdomen and pelvis: enlarged liver measuring 17cm with increased liver parenchymal echogenicity, multiple calculi in gall bladder(cholelithiasis)

Serological findings (anti-HBc; negative HBs, HbeAg=non reactive) suggested recovery from hepatitis B infection. Other viral and parasitic markers were found to be negative (anti-HAV, anti-HCV, anti HEV, anti-HIV 1, anti-HIV 2, anti-rubella IgM, anti-CMV IgM, EBV VCA IgM, parvovirus B19 IgM, anti-toxoplasma IgM and IgG). Autoimmune hepatitis markers such as antinuclear antibody, anti-smooth muscle antibody, anti-liver-kidney-muscle, antimitochondrial antibody were negative. Thus, any possible infectious hepatitis, autoimmune hepatitis, and primary biliary cirrhosis were excluded. Serum Ferritin and Serum alpha1-antitrypsin were found to be in the normal range. No corresponding signs or symptoms of amyloid organ infiltration or Gaucher disease were seen. Possible diagnoses included nonalcoholic steatohepatitis, primary (congenital) glycogen storage disease, and secondary glycogen storage disease. Liver biopsy was performed, liver biopsy shows Periodic acid-Schiff (PAS)-positive granulesin enlarged hepatocytes, indicating the presence glycogen deposition.

International Journal Dental and Medical Sciences Research
Volume 4, Issue 6, Nov-Dec 2022 pp 60-63 www.ijdmsrjournal.com ISSN: 2582-6018

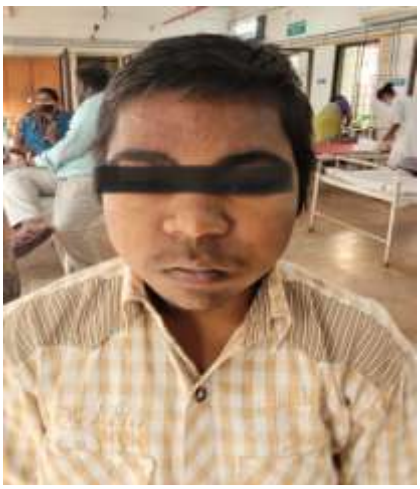

FIG-1: Mauriac syndrome patient with moon face and diminished facial hair growth

FIG-2: retarded growth, height measuring 145cm

III. DISCUSSION

IDDM is one of the common endocrinological morbidity in pediatric population and its various complications in this population are not unknown. Mauriac syndrome is one of such complications. The pathophysiologic process of glycogenic hepatopathy involves two components: hyperglycemia and over insulinization[9] In patients with poorly controlled IDDM, hyperglycemia increases the need for insulin. When insulin is administered to the patient in higher amounts, more quantities of active glycogen synthase are activated by the insulin. Increased activation of enzyme promotes hepatic glycogen storage by conversion of glucose-1-phosphate to glycogen. Because the entry of glucose into the liver via GLUT–2 mechanism is insulin independent, hyperglycemia itself also initiates glycogen synthesis.In glycogenic hepatopathy, hypercortisolism also contributes to glycogen storage in the liver[10], as was evident in this patient. Hypercortisolism also causes delay in sexual maturation and growth in patients with Mauriac syndrome[10]. Glycemic fluctuations may also cause hepatic glycogenosis, due to the discrepancy between insulin and glucose levels in the blood. The most common cause of these fluctuations is uncontrolled diabetes mellitus.

It is important to distinguish Nonalcoholic Steatohepatitis (NASH) from glycogenic hepatopathy. NASH warrants weight loss, correction of hyperglycemia, improvement of hypertriglyceridemia and therapy using insulin-sensitizing agents and Urso deoxycholic acid[11]. However, in glycogenic hepatopathy, glycemic control by adequate intensive insulin regimen reverses the condition of glycogen deposition and hepatomegaly. Although glycogenic hepatopathy does not progress to cirrhosis[12-14]. NASH is an established cause of cirrhosis and is frequently diagnosed worldwide[11]. The chief means of distinguishing between NASH and glycogenic hepatopathy is a liver biopsy.

In our case there was a significant history early onset of IDDM with inadequate insulin therapy resulting in poor glycemic control and clinical findings of short stature, moon facies, underdeveloped secondary sexual characters, protruded abdomen with hepatomegaly and laboratory findings suggestive of dyslipidemia, elevated liver enzymes and liver biopsy reported glycogen deposition, all the above features suggestive of Mauriac syndrome, along with these our patient developed diabetic nephropathy,retinopathy and systemic hypertension. Patient was treated with regular insulin,after glycemic control given diuretics, iron supplementation and advised diabetic diet.

In patients with Mauriac syndrome, all the clinical features regress with optimum insulin therapy and strict control of blood glucose levels. During follow up, in patients with glycogenic hepatopathy, hepatomegaly and elevated liver enzymes generally return to normal with tight metabolic control[15,16]. In this patient, hepatomegaly and elevated liver enzymes returned to normal after one month of discharge. In a patient with IDDM, if hepatomegaly persists for a period longer than four weeks, other causes must be investigated [16].

IV. CONCLUSION

Severe growth retardation with pubertal delay, obesity and hepatomegaly such as in Mauriac syndrome, have been reported often in IDDM patients belongs to adolescents and young adult age group. These conditions are now rarely seen due to improvements in insulin therapy and glycemic control. However, one should look into the possibility of occurrence of individual features of Mauriac syndrome. One should also remember that the Mauriac syndrome may occur in other types of diabetes. It is essential to be aware of Mauriac syndrome's importance as most of the clinical features are reversible with good metabolic control.

REFERENCES

[1]. Mauriac P. Gros ventre, hepatomegalie, troubles de croissance chez les enfants diabetiques traits depuisplusiersannee par l'insuline. Gaz Hebd Med Bordeaux 26 (1930): 402-410.

[2]. Mandell F, Berenberg W. The Mauriac syndrome. Am J Dis Child. 1974; 127(6): 900–902, doi: 10.1001/archpedi.1974.02110250126021, indexed in Pubmed: 4834782.

[3]. Madhu SV, Jain R, Kant S, et al. Mauriac Syndrome: A rare complication of type 1 diabetes mellitus. Indian J Endocrinol Metab 17 (2013): 764-765.

[4]. Kocova M, Milenkova L. Old syndrome-new approach: Mauriac syndrome treated with continuous insulin delivery. SAGE open medical case rep 6 (2018): 2050313X18785510.

[5]. Lee RG, Bode HH. Stunted growth and hepatomegaly in diabetes mellitus. J Pediatr 91 (1977): 82-84.

[6]. Najjar S, Ayash MA. The Mauriac syndrome. Clin Pediatr (Phila). 1974; 13(9): 723–725, doi: 10.1177/000992287401300904, indexed in Pubmed: 4416595.

[7]. Olszowska L, Ludwiczak H. Mauriac syndrome — an ever present problem in infantile diabetes. Pediatr Pol. 1976; 51(8): 931–935, indexed in Pubmed: 967595.

[8]. Otto-Buczkowska E, Rzepka J, Sońta-Jakimczyk D, et al. Somatic development of diabetic children. Pediatr Pol. 1973; 48(3): 307–315, indexed in Pubmed: 4693964.

[9]. Rogal SS, Ukomadu C, Levy BD, Loscalzo J. Clinical problem-solving. A sweet source of abdominal pain. N Engl J Med. 2011;364:1762-1767.

[10]. Flotats Bastardas M, Miserachs Barba M, Ricart Cumeras A, Clemente León M, GussinyerCanadell M, Yeste Fernández D, Albisu Aparicio MA, Carrascosa Lezcano A. [Hepatomegaly due to glycogen storage disease and type 1 diabetes mellitus]. An Pediatr (Barc). 2007;67:157-160.

[11]. Smith BW, Adams LA. Nonalcoholic fatty liver disease and diabetes mellitus: pathogenesis and treatment. Nat Rev Endocrinol. 2011;78:456-465.

[12]. Chatila R, West AB. Hepatomegaly and abnormal liver tests due to glycogenosis in adults with diabetes. Medicine (Baltimore). 1996;75:327-333.

[13]. Torbenson M, Chen YY, Brunt E, Cummings OW, Gottfried M, Jakate S, Liu YC, Yeh MM, Ferrell L. Glycogenic hepatopathy: an underrecognized hepatic complication of diabetes mellitus. Am J Surg Pathol. 2006;30:508-513.

[14]. von Sengbusch A, Tannapfel A, Klein HH, Hering S. [Hepatomegaly und elevated transaminases in a 24 year-old with type 1 diabetes mellitus]. Internist (Berl). 2010;51:84-87.

[15]. Munns CF, McCrossin RB, Thomsett MJ, Batch J. Hepatic glycogenosis: reversible hepatomegaly in type 1 diabetes. J Paediatr Child Health. 2000;36:449-452.

[16]. Abaci A, Bekem O, Unuvar T, Ozer E, Bober E, Arslan N, Ozturk Y, Buyukgebiz A. Hepatic glycogenosis: a rare cause of hepatomegaly in type 1 diabetes mellitus. J Diabetes Complications. 2008;225:325-328.