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Case Report

Mauriac syndrome; a rare complication of type 1 diabetes mellitus

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ABSTRACT

Mauriac syndrome is a rare complication of poorly controlled Type 1 diabetes mellitus.It is characterized by, truncal obesity, delayed puberty, cushingoid features, hepatomegaly. Although rare in developed countries, cases are seen in developing countries like India due to lack of awareness and poor socio-economic status. We report a case of 5 years old male child with type 1 diabetes mellitus and poor glycemic control with features of Mauriac syndrome from Jhalawar district, Rajasthan.

Keywords: Mauriac syndrome, Type I diabetes mellitus, Hepatomegaly, Rare complication

INTRODUCTION

The Mauriac syndrome is a rare complication of type 1 diabetes related to chronic under insulinisation that is characterized by growth failure and hepatomegaly due to excess glycogen accumulation in liver.[1] Clinical features include Moon face, protuberant abdomen, proximal muscle wasting , enlarged liver from fat and glycogen infiltration.[1] Although rare in developed countries, cases are seen in developing states where patients are not well aware about self-management of type I diabetes mellitus.[2]

We present a case of type 1 diabetes mellitus with chronic poor glycemic control with features in spectrum of Mauriac syndrome.

CASE REPORT

We describe a 5-year-old male child with insulin dependent diabetes diagnosed at 1 year of age, on human mixtard insulin. He presented with breathing difficulty and high blood sugar. From history we understood that the treatment compliance was not good because of improper injection technique, poor blood sugar monitoring. On examination, he was conscious, oriented, febrile, growth retardation (Height for age <3rd centile, weight for age <50th centile) was present, system examination showed abdominal distension with enlarged liver. At admission RBS was 426mg/dl and urine positive for ketone. Hemogram was normal, HbA1c was high 10%, triglyceride 261 mg/dl, cholesterol 210 mg/dl, aspartate amino transferase 20 IU/L, alanine amino transferase 17 IU/L, with normal serum bilirubin. Serum urea, creatinine were 33 and 0.6 respectively. Thyroid functions were normal. Ultrasound confirmed moderate hepatomegaly and normal spleen. Urine examination revealed 3+ sugar and 8-10 RBCs.

Past history of Type 1 Diabetes Mellitus since 5 years and hospitalization with Diabetic ketoacidosis at least once in a year. Father was the primary care taker and illiterate, they had issues with insulin availability and storage due to poor socio-economic status. Patient was managed with IV fluids, insulin infusion and IV antibiotics. As the patient was unable to maintain target blood sugar level despite maximum insulin dose and strict dietary adherence, he was referred to Endocrinologist. This patient presented with clinical spectrum of Mauriac syndrome, which is less commonly seen in developed countries now due to better insulin

regimen still is present in developing countries due to lack of awareness and poor socioeconomic status.

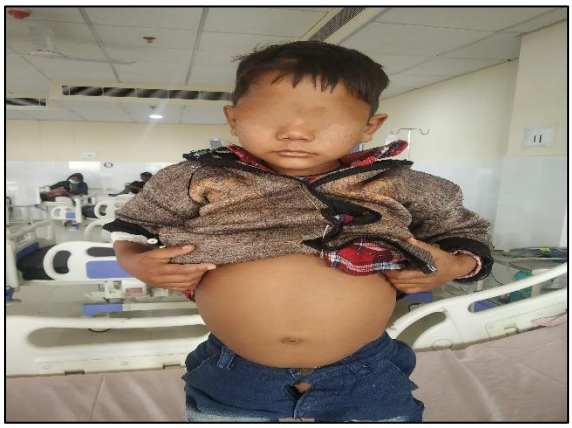

Figure 1: 5 year old boy with type 1 diabetis mellitus and features of mauriac syndrome.

DISCUSSION

Pierre Mauriac in 1930 described this syndrome in children diagnosed as type 1 diabetes mellitus who presented with growth failure, pubertal delay, abdominal distension and hepatomegaly and who are treated with short acting insulin.[3]

The age of presentation is commonly in adolescence; however, there have been reports on toddlers (1-5), young children (16-18) and adults (19,20). Sood et al reported a case of 19 year old child while Kim et al4 reported a case of 13.5 year old child.[2] Our patient was of 5 years age, there were no sex predeliction.[4] Its aetiology 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.[5] Hepatomegaly is thought to be due to glycogen deposition in the liver.[6]

We described a patient with the similar clinical picture from the rural region of Jhalawar, Rajasthan. Although this condition is commonly seen in areas like this, it is underdiagnosed due to lack of awareness among people.

Few studies suggested that over insulinisation may lead to deranged liver enzymes and liver enlargement and irregular dosing of insulin causing vicious cycle of hyperglycemia and hypoglycemia leading to glycogen deposition and hepatomegaly is associated with the syndrome.[7,8] Decrease in liver span and normalization of deranged live enzymes is reported with achieving good glycemic control. It may take few months or even years to achieve normalization of liver.

CONCLUSION

Mauriac syndrome is rare complication of insulin dependent diabetes mellitus. It is commonly present in adolescence but few reported cases showed its presence in younger children too. Long term poor glycemic control is the prime cause. Strict glycemic control can cause regression of symptoms. The condition often goes undiagnosed due to lack of awareness.

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