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Mauriac Syndrome: a diagnosis that still exists

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Introduction

Mauriac syndrome is one of the complications of poorly controlled type1 diabetes mellitus (T1DM). It is characterized by poor glycemic control, hepatomegally, growth disorders with Cushinoid features and delayed puberty. Since the introduction of new forms of insulin cases of Mauriac syndrome are rarely reported.

Objective

To report a case of T1DM complicated with Mauric syndrome from Eastern Sudan.

Case Report

A 14- year old boy, was referred for evaluation of short stature and poor diabetes control. He was diagnosed as T1DM at 9-years of age. Since then he was on the same dose of premixed insulin, 4 units in the morning and 2 units in the evening. The doses were omitted

several times, either due to lack of insulin or poor compliance and was admitted many times with DKA. The last one was 10 days prior to referral. Clinical examination revealed height 116 cm (less than 3rd percentile), weight 20.3 kg (<3rd perenentile), BMI of 15.8. He had distended abdomen with hepatomegaly, muscle wasting and no signs of puberty. Investigations revealed normal haemoglobin, liver renal and thyroid function tests. He had no microalbuminuria and coeliac screening was negative. Blood glucose ranged between 270-400 mg/dl and HbA1c was 11%. Based on the history, clinical features and investigations, the patient was diagnosed as Mauriac syndrome.

Conclusion

The incidence of Mauric syndrome has significantly declined with improvement of the glycemic control, however cases are still seen in low resources countries as the optimum management is not possible for all patients.



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