Proceeding S.Z.P.G.M.I vol: 15(2) 2001, pp. 103-104.

Mauriac Syndrome - A Case Report

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SUMMARY

This is the case report of a 16 year old diabetic child who presented with failure to gain height for 4 years and generalized swelling of the body for 1 week. He was diagnosed to have Mauriac syndrome.

INTRODUCTION

Mauriac syndrome is a complication of diabetes mellitus that is clearly related to under insulinization. Its features include hepatomegaly, limited joint mobility (LJM), short stature and maturation delay.

CASE REPORT

A 16 yrs old male presented with failure to gain height for 4 years and generalized swelling of the body for 1 week. The patient was a known case of Type-I diabetes mellitus for 13 years, diagnosed at the age of 3 years when he had polyuria. He was currently on combination insulin therapy but with poor glycaemic control and no regular follow-ups.

On examination he was found to be short statured. His height was 141 cm and weight was 42 kg,both of them were less than 5th centile for his age. His blood pressure was 140/100 mm Hg. He had stage 3 limited joint mobility, hyperpigmentation and hypertrophy of the injection site at left arm, periorbital swelling, ankle oedema and stage 2 sexual maturation. He also had hepatomegaly but rest of the examination including the eye was normal.

His investigations showed a blood glucose level of 402 mg/dl, with +3 proteinuria. His liver function tests were normal; BUN=40, creatinine=3mg/dl and S.total protein=5.2gm/dl, albumin=2.3 gm/dl. He also had high levels of S. Cholesterol= 355mg/dl, S. triglyceride=219, LDL=246 mg/dl. Thyroid function tests were normal. On X-rays, the bone age was found to be of 13 years. Ultra sound abdomen showed an enlarged liver with fatty changes. A final diagnosis of uncontrolled diabetes was made along with Mauriac syndrome, lipohypertrophy and suspected diabetic nephropathy.

DISCUSSION

Mauriac syndrome is rare nowadays because of the availability of longer acting insulins. Its features include a glycogen laden enlarged liver, syndrome of limited joint mobility, short stature, growth impairment and maturational delay[1].

Delayed linear growth and pubertal development is probably only seen with poor control. LJM is another feature which may be related to chronic hyperglycemia and has been correlated with early microvascular abnormalities such as retinopathy and nephropathy, which may appear before 18 yrs of age.

The syndrome of limited joint mobility is classified into six stages; stage 0-no abnormality, stage 1-skin thickening without contractures, stage 2-bilateral 5th finger contractures, stage 3-other fingers involved bilaterally, stage 4-fingers and wrist joint, stage 5-fingers, wrists and other joints are involved. The presence of LJM is an indication for critical appraisal of glycaemic control[2]. LJM is present in almost 15-30% of adolescents with IDDM. The earliest abnormality appears to be a sclerodermatous, tight, waxy skin consistency[3].

An increase in insulin dose and caloric intake may be sufficient to improve growth in some

children with Mauriac syndrome[4].

Lipohypertrophy probably reflects the anabolic effect of insulin on fat metabolism and thus results in fatty lumps in subcutaneous tissue at the site of repeated injection[5]. It should not occur if injection sites are regularly rotated. In our case, the child's dose of insulin was adjusted along with caloric intake with an improvement in the glycaemic control. Improvement in other features could not be recorded as the child did not report for follow up.

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