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Case Report Open Access

Mauriac Syndrome: Rare Complication in Type-1 Diabetic Children

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Abstract

Mauriac syndrome is rarely noted complication of type-1 diabetic children, which is typically associated with growth failure and delayed pubertal maturation. We have presented a case report of 13 years male adolescent who had multiple hospital admissions for diabetic ketoacidosis.

Keywords: Mauriac syndrome; Type-1 diabetes; Blood glucose; Hepatomegaly; Growth failure

Introduction

Mauriac syndrome is rarely noted complication of type-1 diabetic children characterised by dwarfism, osteopenia and glycogen laden enlarged liver [1]. It is typically associated with growth failure and delayed pubertal maturation, although these effects can be reversed with good glycemic control [2].

Case Presentation

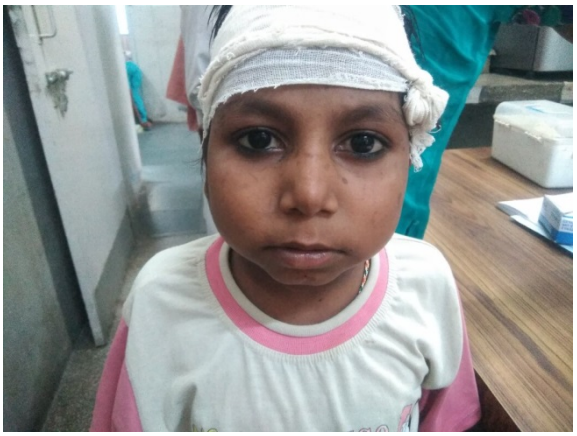

Figure 1: Male adolescent presented with complaint of scalp abscess.

We are presenting case report of 13 years male adolescent presented with complaint of scalp abscess (Figure 1) for 1 month, diagnosed at 2 years of age and was on mixtard insulin on an average dosage of 0.8 units per kg per day. Patient had multiple hospital admissions for diabetic ketoacidosis. His family history is noncontributary and immunized as per age. His anthropometry is suggestive of short stature, height/110.5 cm (<3rd percentile), weight 20 kg (<3rd percentile) bone age 7 years, US: LS ration/1.06, weight/20 kg, proportionate dwarfism. On examination vitals within normal limits, local examination of head showed 10 × 10 cm diffuse boggy swelling fluctuant. On systemic examination, abdominal distension present with hepatomegaly total span 14 cm nontender, smooth surface and rounded margins (Figure 2). CVS, RESP and CNS examination was normal.

Figure 2: Abdominal distension present with hepatomegaly.

On admission random blood sugar was not controlled, ketones were negative. Intially patient was switched from mixtard to regular insulin for proper control of random blood sugar. HbA1c was 8%. FSH/0.573 (1.4-1.8 mIU/ml), LH/0.152 (1.5-9.3 mIU/ml), Testosterone was zero (0.07 ng/ml) and RFT and serum electrolytes were within normal limit. LFT showed elevated liver enzymes SGOT/82 IU/l, SGPT/60 IU/l. Opthalmologist opinion was taken which showed bilateral cataract advised surgery. Scalp abscess was drained under all aseptic condition.

Discussion

Mauriac syndrome is an uncommon complication of uncontrolled type 1 diabetes reported in children belonging to age group of 13-17 years [3].

It is associated with hepatomegaly and diabetic dwarfism. The actual cause is unknown, but it is probably a combination of factors including inadequate glucose uptake and utilization in the tissues, decreased

insulin-like growth factor-1 and growth hormone levels, impaired bioactivity of these hormones, a circulating hormone inhibitor, resistant or defective hormone receptors, insulin deficiency, poor glycemic control, concurrent autoimmune diseases, decreased caloric intake and/or eating disorders. The periods of supraphysiological levels of insulin is associated with glycogen deposition in the liver leading to hepatomegaly [4,5]. Blood glucose passively enters the hepatocytes in which glycogen synthesis is promoted by high cytoplasmic glucose concentration reliant on the presence of insulin. Glycogen is then trapped within the hepatocytes as a result of a vicious cycle of hyperglycemia and insulin treatment [6]. Poor glycemic control due to hypoinsulinemia leads to lipolysis and ketone liberation. Ketosis activates cortisol synthesis promoting the release of fatty acids and hyperglycemia [7].

Conclusion

Growth failure, delayed puberty and hepatomegaly in Mauriac syndrome improve with glycemic control [8]. This is a case of adolescent male with poorly controlled DM who found to have short stature. With the additional findings of gross hepatomegaly, delayed puberty and a Cushingoid habitus, he was diagnosed with Mauriac syndrome. In this case, poor compliance because of poor literacy was the main cause behind poorly controlled diabetes.

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