Type 1 Diabetes Mellitus in Homozygous Sickle Cell Anaemia

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Abstract
For reasons unknown, the association of diabetes mellitus with sickle cell anaemia is uncommon. A patient of sickle cell anaemia with diabetes mellitus, complicated with ketoacidosis is being reported in view of its rarity.©

INTRODUCTION
The prevalence of diabetes mellitus is very low among patients with sickle cell disease. A number of studies failed to detect a single case of diabetes among patients with sickle cell anaemia.1,2 However, few case reports of combined presentation of both the diseases without any complication have been reported among black population.3,4 No such case has been reported from India till date. This case report describes a patient of sickle cell anaemia with Type-1 diabetes mellitus complicated with ketosis.

CASE REPORT
SRK, a 17 years girl admitted to the 4th unit of Department of Medicine, VSS Medical College Hospital, Burla, Orissa with the complaints of loss of weight for 5 months; fever, pain in abdomen, and burning sensation during micturation for 7 days. She is a known case of sickle cell anaemia, diagnosed at the age of 12 years. She had been hospitalized thrice for painful crisis. Her parents had sickle cell trait. There is no family history of diabetes mellitus.

On examination, the patient had average built, height 5ft. 3 inches, weight-34 Kg, pulse rate-120 per minute, blood pressure (right hand and supine)-110/70 mm of Hg. She was anaemic and icteric. On abdominal examination liver was enlarged 3cm below the costal margin. Spleen was not palpable. Investigations showed: Hb-8 gm/dl, differential count- N: 82%, L:10%, E:6%, M:1%, B:1%, total leukocyte count-16,000/cmm, fasting blood glucose-530.0 mg/dl, blood urea-23.0 mg/dl, serum creatinine-1.1mg/dl, serum bilirubin-2.6mg/dl, C-peptide-0.2 ng/ml, glycosylated haemoglobin (Hb.A1c)-11.5%, lipid profile: S. cholesterol-181.0mg/dl, triglycerides-174.0mg/dl, LDL-121.2mg/dl, HDL-25.0mg/dl, VLDL-34.8mg/dl. Urine examination showed the presence of reducing sugar, protein, ketone bodies, and clumps of pus cells. E. coli was grown in culture and was sensitive to Amikacin. Haemoglobin electrophoresis showed SS band. High performance liquid chromatography (Variant, β-thalassemia Short Program, Biorad) showed Hb S window-76.8%, Hb F-10.5%, Hb A2 – 1.9%.

The diagnosis of sickle cell anaemia with diabetes mellitus, complicated with urinary tract infection and ketoacidosis was made and treated with normal saline, Inj. Ceftriaxone 1000 mg I.V.daily, Inj. Amikacin (15mg/Kg) 250mg twice daily, human neutral insulin, and other supportive measures. Human neutral insulin was administered intramuscularly at the dose of 4 IU/hour. Blood glucose was normalized with 90 units/day on 4th day. She recovered and was discharged on the 12th day.

DISCUSSION
Concurrent sickle cell anaemia with diabetes mellitus is very rare and with ketosis, still rarer. Only few case reports of this uncommon association of these two diseases are available.3,5 A couple of homozygous sickle cell patients with pregnancy and insulin dependent diabetes mellitus had been reported earlier.3 Combination of these two diseases in two children had also been reported from Nigeria.4,5 All previously reported cases were not associated with any complication of diabetes mellitus. The present case reported here is the first case of sickle cell anaemia with Type-1 diabetes mellitus complicated with ketosis. This patient presented with fever and abdominal pain, hence initially a provisional diagnosis of sickle cell anaemia with crisis had been made. This is because abdominal pain is one of the presenting features of sickle cell crisis with or without acute pancreatitis.6 Diabetes mellitus was diagnosed in this patient only when fasting blood glucose was done as a routine investigation. Because of the young age at
onset, absent family history of diabetes, and the presentation with ketosis, the diagnosis of Type-1 diabetes mellitus was made.

To investigate the prevalence of diabetes mellitus among sickle cell anaemia patients, Morrison et al conducted a study with pregnant black patients with sickle cell anaemia. The authors selected pregnant patients because it is well known that pregnancy is diabetogenic and latent diabetes may be unmasked after 28 weeks of gestation. However, the authors could not detect a single case of diabetes as compared to 4% prevalence in the control.1 Another study, which had been conducted in Nigeria to detect the beneficial effect of glycosylated sickle Hb (HbSS) also failed to detect a single case of HbSS-diabetic.2 Similarly from Orissa, India where the frequency of sickle cell gene is very high (15.1%), diabetes has not been reported among patients homozygous and heterozygous for sickle cell gene.6,7

There are no satisfactory explanations for the uncommon association of these two diseases. One explanation is that majority of patients with sickle cell anaemia died early, therefore, relatively small number of patients survived for the clinical manifestation of diabetes.5 However, sickle cell anaemia found in India and Saudi Arabia (Asian haplotype) is less severe than African haplotype and a significant proportion of patients survived more than 30 years of age.6,8 In spite of longer survival, concurrent diabetes with sickle cell anaemia has not been reported from India.6,7 Hence, some other unknown factor(s) may be responsible for this rare association. Another explanation is genetic. In support of this hypothesis is the fact that both the b-globin and the insulin genes are present in short arm of chromosome 11.1 It is not known whether the genetic loci of insulin and b-globin have any inhibitory effect on the inheritance pattern or penetrance of the other. Therefore, the relation between diabetes mellitus and sickle cell anaemia needs further evaluation.

REFERENCES


Announcement

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