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Hirata Diseases: Autoimmune Hypoglycaemia

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Abstract

Insulin autoimmune hypoglycaemic syndrome is characterized by spontaneous symptomatic hypoglycaemia in a non-diabetic patient or without administration of exogenous insulin accompanied with significantly high serum content of total immunoreactive insulin and presence of insulin auto-antibodies in high titer.

Keywords: Insulin • Insulin antibody • Hypoglycaemia

Introduction

High concentration of serum insulin and insulin antibodies with fasting hypoglycaemia is known as insulin autoimmune syndrome. The first patient with insulin –binding autoantibody was reported by Hirata in 1970 [1].

Insulin Autoimmune Syndrome (IAS) also known as Hirata's, is not a common entity however it should be suspected in a non -diabetic patient with spontaneous symptomatic hypoglycemia in the absence of localization of an insulin secreting tumor in the body and excluding all those medications associated with the development of insulin autoimmune syndrome [2]. IAS accounts for 3.6% of all spontaneous hypoglycemic attacks in Japan [3]. Although IAS is rare in India, until now only 13 cases have been reported. We hereby report a typical case of IAS.

Case Presentation

A 48 years old male presented on 23rd August 2021 with complaints of recurrent giddiness, sweating, palpitation, tremors with a feeling of impending death since last 14 month. These symptoms were relieved after consumption food or sweets. He never forgot to keep a sweet candy in his pocket. Even during taking his history, he showed us the sweet chocolate from his pocket. This gave us a hint to the severity of his illness. He was examined by a neurologist and 6 other physicians. He underwent a brain MRI and was also put on anti- platelets drugs, with no benefits. He denied any family history of diabetes or consensual marriage. His blood pressure, serum electrolytes, thyroid stimulating hormone and electrocardiograph were normal. He denied history of consumption of anti-diabetic drugs, insulin therapy, anti- thyroid medications, herbal remedies, antacids and steroids. There was No history suggestive of hepatitis C infection or autoimmune diseases. His serum C-peptide level was 18.88 ng/mL (normal 0.90-7.1). Initially we thought of insulinoma, but the MRI of pancreas was normal (appendix) there was no acanthosis nigricans.

Blood was collected for serum insulin at time when blood sugar was 40 mg/dL. Serum insulin level was 13885.2 micro unit/mL (normal 6-55). Moreover serum insulin level in thousands excludes the possibility of a pancreatic insulinoma or insulin secretary tumours anywhere in the body. Serum insulin antibody was positive and >100 u/mL. D-hydroxybutyrate (ketone body) level was 0.02 mmo/L (normal 0.02-0.27). 78% of ketone body seen circulating in blood is in form of D-3 hydroxybutyrate which is the most sensitive marker for ketosis. Ketosis can be associated with diabetes mellitus, hypoglycemia, dieting, epilepsy and sepsis. He was further investigated for fasting, post breakfast, post lunch and bed time blood sugar levels for five days. Keeping autoimmune hypoglycaemia in mind, he was started on oral steroid therapy at 1 mg/kg body weight. The average before steroid therapy and after completion of steroid 1mg/kg body weight tapered over six weeks (Figures 1 and 2) [4-7].

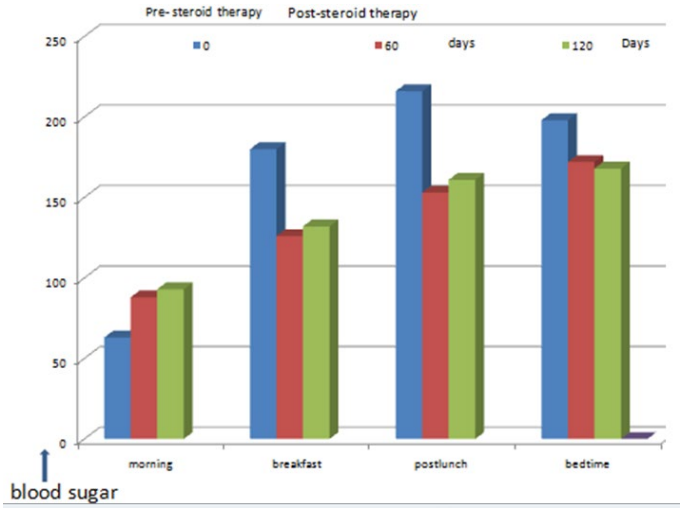

Figure 1. Average blood sugar fasting, breakfast, post lunch and at bed time studied for five days before steroid 9 pre-steroid) and after steroid (post-steroid therapy).

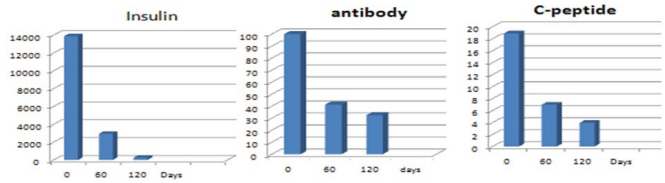

Figure 2. Showing the insulin, insulin antibodies and serum c-peptide levels after or before therapy, 60 and 120 days after steroid therapy.

Conclusion

These findings confirmed that our patient was suffering from Insulin Autoimmune Hypoglycemia (IAH). IAH is characterized by auto antibodies against beta cells of pancreas which secrete endogenous insulin without any prior exposure to synthetic exogenous insulin. Till now only 11 cases of IAH have been reported from India that too from tertiary care centres. This is the very first case to be suspected clinically and confirmed by laboratory

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investigations from a rural setting. In rural settings, insulin receptor antibodies and HLA analysis is not possible. Hirata disease is classically characterized by significantly raised insulin antibody, hypoglycaemia bur a comparatively lesser rise in C-peptide levels. High incidence of Hirata disease has been reported from Japan. Recently 7 cases of IHA were reported from a tertiary care hospital in Mumbai of which 2 had HLA-DRB1 × 03 and DRB1 × 04 and HLA-DRB1 × 04 respectively. In Japan, remissions achieved without any intervention. However in India remission is achieved by immunosuppressant's or oral steroids without antacid. Acanthosis nigricans accompanied with insulin resistant diabetes with raise in insulin receptor antibodies.

In conclusion recurrent sweating, giddiness, weakness improved with consumption of sweets must be investigated for fasting serum insulin levels, insulin antibodies and response to steroid therapy.

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