

UNUSUAL PRESENTATION OF ADDISON'S DISEASE

Zaman Shaikh¹, Muhammad Umar², Mehwish Nisar³

Abstract

Addison's disease usually presents with non-specific symptoms like fatigue, nausea, vomiting, hyperpigmentation and generalized weakness. These symptoms are most often ignored or misinterpreted with other more common diseases. This is the major reason that this disease is under-diagnosed. Therefore, to establish a diagnosis, high index of suspicion is needed. We are reporting a case of six years old boy who presented with recurrent episode of severe hypoglycemia, so much so that he was receiving regular IV fluids including dextrose water & normal saline. While walking he was supported by his parents all the time because of recurrent attacks of fall due to extreme degree of hypoglycemia and subsequent weakness. His blood sugar was in the range of 40-90mg/dl, as a result, he was hospitalized several times. He was already worked up to exclude insulinoma before presenting to our institute. Eventually he was diagnosed as having Addison's disease.

KEYWORDS: Hypoglycemia, Addison's disease, Adrenal insufficiency.

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INTRODUCTION

Addison's disease refers to primary hypo-adrenalism caused by a total or near total destruction or dysfunction of both adrenal cortices.^{1,2} A deficiency of ACTH can also produce hypo-cortisolism but this is known as

secondary adrenal insufficiency. Addison's disease is an endocrine disorder that occurs in about, 1 in 1,00,000 people. It occurs in all age groups and afflicts men and women equally.³ The two most common causes of Addison's disease are autoimmune adrenalitis and tuberculosis. Other causes include invasion of glands by neoplastic cells, CMV virus, HIV, hemochromatosis, amyloidosis, hemorrhage (Water House Frederickson syndrome) and surgical removal of glands.^{4,5}

The symptoms of adrenal insufficiency usually develop gradually with chronic fatigue, muscle weakness, loss of appetite, nausea, vomiting, and diarrhea. In about 50% of cases, blood pressure is low and falls further in erect posture, causing dizziness or fainting. Skin changes are also common with the areas of hyperpigmentation, more on exposed parts of the body. Addison's disease can cause irritability and depression because of salt depletion, resulting in craving for salty foods. In women, menstrual periods become irregular or stop altogether; loss of pubic and axillary hair may also occur.^{1,6}

The combination of autoimmune disorder and Addison's disease is described as

1. Dr. Zaman Shaikh,
Professor of Medicine,
Director,
 2. Dr. Muhammad Umar Khan,
Clinical Registrar,
 3. Dr. Mehwish Nisar,
Resident Medical Officer,
- 1-3: National Institute of Diabetes and Endocrinology,
Ojha Campus,
Dow University of Health Sciences,
Karachi - Pakistan.

Correspondence

Prof. Zaman Shaikh
Director
National Institute of Diabetes and Endocrinology,
Ojha Campus,
Dow University of Health Sciences,
Karachi - Pakistan.
E-mail: drzamanshaikh@yahoo.com

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polyglandular autoimmune disorders, type I (PGA I) and type II (PGA II). In PGA I, there is associated hypothyroidism and mucocutaneous candidiasis and in PGA II there are hypo- or hyper-thyroidism, type I diabetes mellitus, primary ovarian or testicular failure and pernicious anemia.^{1,4}

CASE REPORT

A six years old boy, Master JA was brought by his parents in the National Institute of Diabetes and Endocrinology (NIDE), Ojha Campus, Dow University of Health Sciences, Karachi, with the complaints of recurrent episodes of hypoglycemia and generalized weakness for four months. Hypoglycemia usually occurred daily. These episodes of hypoglycemia were associated with anorexia, vomiting and generalized weakness. Vomiting occurred 4-6 times a day, non-projectile and containing mainly food particles. There was history of weight loss of approximately 5kg in four months. Generalized weakness was so severe that it resulted in difficulty in walking for which he was being supported by his parents.

He attained his milestones at appropriate age but did not get any type of childhood vaccination. There was no history of tuberculosis or any other major illnesses in his family. On examination the boy was looking ill and lethargic. He had short stature with ectomorphic built. His vitals showed a significant postural drop from 90/60 mmHg in lying position to



Fig-1: Patient showing hyper-pigmentation & supported by his mother because of generalized weakness



Fig-2: Hyper-pigmentation over the tongue

75/50mmHg in standing posture. He was anemic and dehydrated. There were also areas of hyper-pigmentation in the buccal mucosa, gums, and on palmer creases. His height was 100cms which was less than 3rd percentile and he was weighing 11kg (Fig- 1 to 3)

For these complaints patient visited different family physicians and was admitted three times in private hospitals. His minimum blood sugar recorded during these hypoglycemic episodes was 30mg/dl on one occasion, when he was brought to a private hospital in stuporous condition. His blood sugar was first measured by glucometer and then confirmed by laboratory. When he was referred to NIDE, his Hb was 11.5g/dl, showing normocytic normochromic picture. Fasting blood sugar was 65 mg/dl, serum sodium 130mEq/L and potassium 4.8mEq/L. X-Ray chest revealed prominent bilateral hila with calcification mainly on right side and microcardia.



Fig-3: Hyper-pigmentation over the gums and lips



Fig-4: Patient is now able to stand without support with reduced body pigmentation after initiation of treatment

His ultrasonography and CT scan of abdomen were normal. No calcification or any other abnormality was seen in adrenal glands. Mantoux test was negative. Fasting serum insulin was less than 1.0 micro unit already advised by a general physician suspecting insulinoma because of recurrent episodes of hypoglycemia. Considering the clinical diagnosis of Addison's disease, Short Synacthen test was performed with 250 μ gm of ACTH and result showed serum cortisol baseline of 1.2 μ g/dl, after 30 minutes it was 2.0 μ g/dl and after 60 minutes it was less than 1.1 μ g/dl, thus confirming



Fig-5: Weight increased with reduced body pigmentation after treatment.

the diagnosis of Addison's disease. Antimicrosomal and antithyroglobulin antibodies were negative and thyroid function tests were normal.

The patient was diagnosed as a case of Addison's disease. After extensive workup no definite etiology could be detected in this patient. He was treated with Tab. Hydrocortisone 10mg, two tablets in the morning and one tablet in the evening, this dose was tapered off after three days. Tab. Fludercortisone was given in a dose of 50mcg / day, with this drug therapy, after 6 weeks patient gained 5kg weight. He is now able to walk, even run and there is no postural drop. Severity of hyperpigmentation is reduced. He is looking healthy and cheerful, and at present he is on a maintenance dose of 5mg in the morning and 2.5mg in the evening. (Fig-4 to 6)

DISCUSSION

A medical history of the symptoms is often sufficient to raise a suspicion of Addison's disease. Quite often the first clue is from the abnormal laboratory tests, like hyponatremia, hyperkalemia, hypoglycemia, eosinophilia, neutropenia and hypercalcemia.^{1,2} The most specific test for diagnosis is ACTH stimulation (Synacthen) test. In the short or rapid, stimulation test, measurement of cortisol in blood is repeated 30 and 60 minutes, after an intravenous injection of synthetic ACTH. The patients with adrenal insufficiency responds poorly or do not respond at all. Sometimes a long ACTH



Fig-6: Pigmentation over tongue is reduced after successful therapy.

stimulation test is required to determine the secondary cause of adrenal insufficiency.^{4,7} Since all of the manifestations of Addison's disease are caused by the lack of cortisol and aldosterone, the treatment is to replace these with similar steroids. Cortisol is usually replaced orally by hydrocortisone tablets and aldosterone is replaced by an aldosterone like synthetic steroid, fludrocortisone (Florine) tablets. The doses of each of these medications are adjusted according to the individual's response and any co-existing medical condition. Response may be seen clinically by observing blood pressure, postural drop, reduction in the hyper-pigmentation and bio-chemically may be seen by improvement in the imbalance of the serum electrolytes, blood sugar and serum renin. In emergencies or during surgery, hydrocortisone must be given intravenously.^{4,8} Patients with Addison's disease should be taught to treat minor illnesses with extra salt and fluids. A person who has adrenal insufficiency should always carry identification card, stating his or her condition, with full address & contact numbers. As long as the proper dose

of replacement medication is taken every day, an Addisonian can have a normal crisis-free life.³

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