Case report on Addisons insufficiency

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Abstract
Addisons disease is a relatively rare condition. It as an endocrine disorder which can occur in all age groups, affecting both girls and boys. Clinical manifestations include fatigue, muscle weakness, hypoglycemia, hyperpigmentation, nausea, vomiting and diarrhea. We report a two months old female child who presented with recurrent episodes of hypoglycemia and convulsions.

Keywords: Primary adrenal insufficiency, cortisol deficiency, hypoglycemia

Introduction
Addison’s, also known as primary adrenal insufficiency, was first described by Thomas Addison in 1855 as a syndrome of weakness and hyperpigmentation associated with adrenal gland destruction.
Addison’s disease is an endocrine disorder that occurs in about, 1 in 1,00,000 children. It occurs in all age groups affecting both girls and boys. The two most common causes of Addison’s disease are autoimmune adrenalitis and tuberculosis.

The symptoms of adrenal insufficiency usually develop gradually with chronic fatigue, muscle weakness, loss of appetite, nausea, vomiting, and diarrhea.

Addison’s disease refers to primary hypo-adrenalism caused by a total or near total destruction or dysfunction of both adrenal cortices. A deficiency of ACTH can also produce hypo-cortisolism but this is known as secondary adrenal insufficiency.

In infants, acute adrenal insufficiency (Addison disease) may occur in the context of serious illness (eg, sepsis), prolonged and difficult labor, or traumatic delivery. Tuberculosis (TB), meningococcemia, or any severe septicemia may also result in adrenal insufficiency (Addison disease). However, adrenal insufficiency (Addison disease) may occur without concomitant illness when it is due to congenital adrenal hyperplasia or congenital adrenal hypoplasia.

Patients with adrenal insufficiency (Addison disease) may have hypoglycemia, and most have hypotension. Orthostatic changes in blood pressure and pulse are cardinal signs of adrenal insufficiency. Symptoms of hypoglycemia are common in small children. Altered mental status, even without hypoglycemia, is common in affected patients with acute adrenal insufficiency.

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.

Case report: A 2 months old only female child, presented to the pediatric OPD with h/o recurrent episodes of hypoglycemia since 1 month and 2 episodes of generalized tonic clonic convulsions since 2 days, 24 hrs apart lasting for about 15-20 min and associated with post ictal drowsiness for about 30 min. There was h/o vomiting (3-4 times/day, non projectile) and failure to thrive.
Birth history: Full term, vaginal forceps delivery, baby did not cry immediately after birth, h/o nicu admission for 20 days was present. Past history revealed h/o birth asphyxia and neonatal convulsions at the age of 2 days and the child was receiving gardenal. There was h/o repeated admissions in hospital for convulsions and hypoglycemia. No h/o tuberculosis in the family.

On Examination- Child was lethargic and moderately dehydrated. Temperature-98.6 degrees Fahrenheit, HR-134/min, RR-34/min, peripheral pulses –well felt, BP-76/54mm of Hg. Pallor and skin hyperpigmentation was present.

Systemic examination Respiratory System: Shape of chest was symmetrical, movements of chest were equal on both sides, trachea appeared central in position. Air entry was bilaterally equal, normal vesicular breath sounds heard, no adventitious sounds heard.

Cardiovascular System: No chest deformities seen, apical impulse was located in the left fourth intercostal space just lateral to the mid clavicular line. Heart sounds S1 and S2 heard, no murmurs heard.

Per Abdomen: shape of abdomen appeared normal, all quadrants of abdomen move equally on respiration. On examination of abdomen there was no tenderness, no guarding or rigidity. There was no organomegaly. Abdomen was normal. No calcification was seen in adrenal glands. TFF’s-TSH- 11.13 micro IU/l, FT4-1.22 and hence patient was started on Tab. Eltroxine. USG Cranium revealed changes of HIE grade 2 and MRI was suggestive of perinatal insult. Due to repeated episodes of convulsions and hypoglycemia and hyperpigmentation addisons disease was suspected and the child was investigated further. Serum progesterone levels were done which were 0.09ng/ml, 17OH progesterone-0.69ng/ml, serum cortisol-0.08microgm/dl and ACTH levels were >2500 which supported the diagnosis of addison’s insufficiency. Endocrinologist opinion was taken and patient was started on IV hydrocortisone and Tab. Fludrocortisone. On follow up after 3 weeks- hypoglycemic episodes were decreased, child responded well to the treatment, child was accepting feeds well and weight gain was present.

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 [1-3]. 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 stimulation test is required to determine the secondary cause of adrenal insufficiency [3, 4]. 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 [5, 6]. 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 [7, 8].

References