J MEDICINE 2006; 7 : 70-72

CASE REPORTS

ADDISON'S DISEASE IN CHILDREN

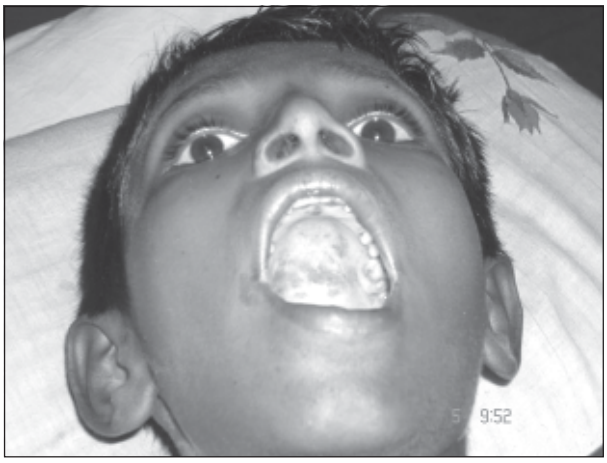
AKM MAMUNUR RASHID[1], MA HASSAN[2], A A MAHBOOB[2], ABDULLAH AL MAMUN[3], MD. BILLAL ALAM[4], KAK AZAD[5]

Introduction

Addison's disease is relatively uncommon disease in western countries. It can present as a life threatening crisis because it is frequently unrecognized in its early stage[1]. The prevalence of chronic primary adrenal insufficiency (Addison's disease) has been reported to be 39 to 60 per million population[2]. The most common cause was formerly tuberculous adrenalitis, but now it is auto immune adrenalitis (slow destruction of the adrenal cortex by cytotoxic lymphocytes), Sometimes accompanied by auto immune Thyroid disease and other autoimmune endocrine deficiencies (autoimmune polyglandular syndrome)[3]. Tuberculosis was overwhelmingly the commonest cause but 20 years later accounted for only 1 in 5 United Kingdom (UK) cases. As with other AI diseases there is a familial incidence and female preponderence[4]. If autoimmune adrenatitis is the underlying disorder the medulla is usually spared[5]. All causes of primary adrenal insufficiency involve the adrenal cortex as a whole resulting in a deficiency of cortisol and aldosterone (plus adrenal androgen), although the severity of the deficiency may vary [6,7]. We encountered a child suffering from Addison's disease recently in our ward. As this was a rare case particularly in the children we found interested to publish as a case report.

Case

A boy of 13 years was admitted with the complaints of abdominal pain for one and half years, black discoloration of whole body for 4 months and anorexia, weight loss, weakness for the same duration. Abdominal pain was not so severe in intensity initially but it increased in severity for the last four months. Black discoloration was first observed in the elbows and knees. No history of such disease in the family. There was no relevant drug history (Fig.-1).



On examination the boy had hyper pigmented lesion all over the body with pigmentation in the gum and soft palate. Blood pressure was 70/40mm of Mercury (Hg). There was no organomegaly but tenderness was present in the right hypochondriac region. There was no other systemic abnormality. The patient was clinically diagnosed incidentally as Addison's disease

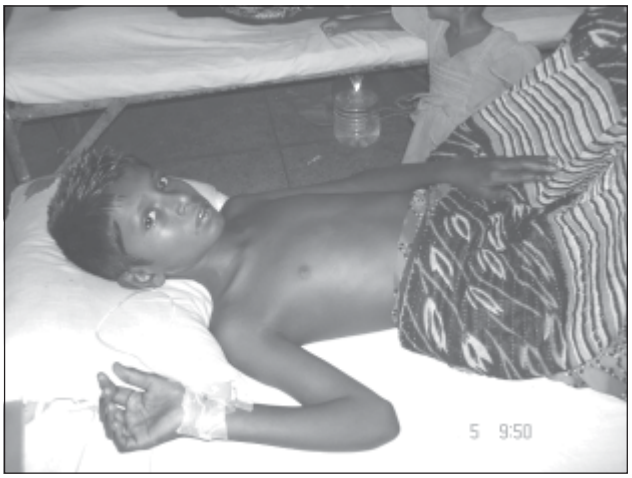


Fig.02.

On investigation Hemoglobulin (Hb%)-68%, Erytrhocyte Sedimentation Rate (ESR) 82mm in the

1. Associate Professor, Department of Paediatric, Khulna Medical College, Khulna.
2. Assistant Professor, Department of Paediatric, Khulna Medical College, Khulna.
3. Associate Professor, Department of Medicine, Khulna Medical College, Khulna
4. Associate Professor, Department of Medicine, Dhaka Medical College, Dhaka
5. Associate Professor, Department of Medicine, Bogra Medical College, Bogra

first hour, Total count of White Blood Cell (WBC) 10,700/cmm, Neutrophils 67%, Lymphocytes-30%, Eosinophils -3%. Urine for routine analysis was normal. Ultrasonogram (USG) of the whole abdomen, X-ray chest, X-ray skull was normal. Electrocardiograph (ECG) showed sinus bradycardia, Bacilli Culmitti Gurine (BCG) Acceleration test was negative. Serum Electrolyte was done and found Sodium (Na^+) 119.5mmol/L, Potassium (K^+)-6.2mmol/L, Chloride (Cl^-) 85.6mmol/L, Bicarbonate ($HCO3^-$) 25mmol/L, Random Blood Sugar (RBS) 4.8mmol/L, Hepatitis B Surface Antigen (HBsAg) was negative. Serum cortisol level was 40.71 ng/dl, normal value was (147-251ng/dl). The boy was finally diagnosed as Addison's disease after summarizing these clinical and investigation reports. The boy was treated accordingly with Prednisolone and Hydrocortisone. He was improved and discharged with these drugs and kept for follow up.

Discussion

Addison's disease is rare in children. It is estimated to be 1 in 10,000.[4] The various etiologies of addison's disease can be grouped in to three categories: 1) Adrenal dysgenesis; 2) adrenal destruction; and 3) impaired steroidogenesis of the adrenal gland currently accounts for most of the cases presenting outside of the newborn period[8]. Recently discovered causes of primary adrenal insufficiency are the acquired immunodeficiency syndrome (AIDS), in which the adrenal gland may be destroyed by a variety of opportunistic infectious agents in up to 5 present of patients in late stage of the disease[9,10]. Our case is also thought to be caused by autoimmunity because we excluded all the probable causes including the tuberculosis. The boy was 13 years old, which was also similar in the age of incidence. Autoimmune disease is a familial incidence with female preponderance entity. The exception was that we encountered male child with no familial incidence of the disease.

Addison's disease presents with fatigue, Muscular weakness, abdominal pain, vomiting, Diarrhoea, salt craving, Behavior change. Headache, sweating, Depression, Muscle and joint pain, postural hypotension, weight loss, generalized pigmentation, darked skin creases, pigmentation of buccal mucosa and nail beds and associated vitiligo and / or goiter were the other presenting features[8].Presenting features of our case were abdominal pain hyper pigmentation, anorexia, wt loss, weakness, hypotension.

High plasma renin and / or increased night Adreno Cortico Tropin Hormone (ACTH) levels low ACTH stimulated cortisol responses, hyponatremia, hyperkalemia, hypoglycemia, Eosinophilia and lymphocytosis is found in Addison's disease[8]. Eosinophilia, lymphocytosis was not observed in our case but there was hyponetremin, hyperkalemia, hypoglycemia and low cartisol level. We could not investigate plasma renin and ACTH level for lack of facilities. X-ray skull and chest was done but no abnormalities were detected. The standard test for detecting antibodies against the adrenal cortex is the indirect immunofluorescence technique used on sections of bovine or human adrenal cortex cut in a cryostat. The sensitivity of this test in patients with autoimmune adrenalitis about 70 percent and the specificity is very high. Recently a simple binding assay that uses radio labeled recombinant human 21- hydroxylase was described[3]. Its sensitivity and specificity were higher than those of the older assay in patient with autoimmune adrenalitis. This assay is not feasible in our context but our case was labeled as autoimmune adrenalitis by method of exclusion of other causes of the disease and by supporting investigations. We suspected the patient when he seeked advice due to weakness and fatigability.

Early detection of the case is needed because primary adrenal insufficiency can become a life threatening disorder in any stressful situation, since cortisol secretion can not be increased at all. Treatment of adrenal crises with full recovery of a dangerously ill patient within a few day is one of the greatest achievements of modern medicine. Whenever a case presents with hyper pigmentation, abdominal pain and easy fatigability, we should consider clinically also as a Addison's disease although it is rare in our context. Thus we can reduce the morbidity and mortality from this disease to a great extent.

References:

1. Willis AC, Vince FI. The prevalence of Addison's disease in Country, UK. Postgrad Med J 1997;73: 286-88.

2. May ME, Vaughn ED, Carey RM. Adrenocortical

insufficiency- clinical aspects. In: Vaughan ED Jr, Carey RM, eds Adrenal disorders. New York Thieme Medical, 1989: 171-89

3. Falorni A, Nikoshkov A, Laureti S et al. High diagnostic accuracy for idiopathic Addison's disease with a sensitive radiobinding assay for autoantibodies against recombinant human 21-hydroxylase. J clin Endocrinol Metab 1995; 80: 2752-5

4. Christopher JH kelnor, Gary E Butter. Endocrine gland disorders and disorders of growth and puberty. In Neil Mcintosh, Peter Helms, Rosalind Synyth. In: Forfar & Arneil's text book of pediatrics (6th ed). Great Britain: Churchill Livingstone 2003: 443-559.

5. Amberson JB, Gray GF. Adrenal pathology. In: Vaughan ED Jr, Carcy RM, eds. Adrenal disorders. New York Thieme Medical. 1989: 13-36.

6. Tsigos C, Arai K, Laronico AC, Digeorge AM, Rapaport R, Chrousos GP. A novel mutation of the adreno corticotrophin receptor (ACTH – R) gene in a family with the syndrome of isolated glucocorticoid deficiency, but no ACTH-R abnormalities in two families with the triple A Syndrome. J clin Endocrinol Metab 1995; 80: 2186-9.

7. Moore PSJ, Couch RM, Perry YS, Shuckett EP, Winter JSD. Allgrove Syndrome: an autosomal recessive syndrome of ACTH insensitivity, achalasia and alacrima. Clin Endocrinol 1991; 34: 107-14.

8. Svetlana T, Maria N, Maclaren. Addison's disease 2001, Clinical review 130. J Clin Endocrinol Metab 2001; 86: 2909-22.

9. Bhatt HR, James VHT, Besser GM et al. Journal of Endocrinology, Thomas Addison society 1994: 69-80.

10. Freda PU, Wardlaw SL, Brudney K, Goland RS. Primary adrenal insufficiency in patients with the acquired immunodeficiency syndrome: a report of five cases. J Clin Endocrinol Metab 1994; 79: 1540-5.