Salt and sour craving and diffuse hyperpigmentation in a 13 year old boy - a case report

A J M Kolitha Jayasundara[1], S Prasad W Kumarasinghe[2]

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Introduction

Primary adrenal insufficiency or Addison's disease is caused by slow destruction of adrenal cortex bilaterally and is a relatively common endocrinopathy with a prevalence of about 40-60 per million of adults[1]. It is rare in children. It causes considerable morbidity and mortality. In contrast, early diagnosis and treatment with appropriate corticosteroid replacement restore normal health and life expectancy.

Case report

A 13 year old schooling teenager had generalized, progressive darkening of skin for 3 years duration which was associated with craving for salt and sour. He had easy fatigability, generalized weakness, tiredness, diffuse arthralgia and myalgia, loss of appetite, nausea, loss of weight, on and off mild generalized abdominal pain not associated with vomiting, diarrhea or constipation. There was no postural dizziness or syncope, no contact history of tuberculosis or past history of pulmonary tuberculosis, malaria or episodes of loss of consciousness. There was no history of exposure to heavy metals. Family members were not affected. His skin colour was much darker than that of family members. He had poor memory in the last several years; it had affected his school work as well.

On examination, he was thinly built, with a weight of 32.5 Kg (< 10th percentile), and a height of 152 cm (75th percentile), generalized hyperpigmentation, pigmentation of; mucosal surfaces, palmar creases, nails with longitudinal bands of darkening, and tongue. Figure 1 shows hyperpigmentation of the skin, lips, gums and the finger nails. There was no vitiligo, oral thrush, dental caries or enlarged tonsils. Hydration was normal. There was no pallor, generalized lymphadenopathy or thyroid gland enlargement. BCG scar was evident There was no axillary or pubic hair. Blood pressure was low but there was no significant postural drop (supine BP 90/50 mmHg, standing BP 80/50 mmHg). Abdominal, respiratory and central nervous system examination was clinically normal. Mini mental score was 19 out of 30.

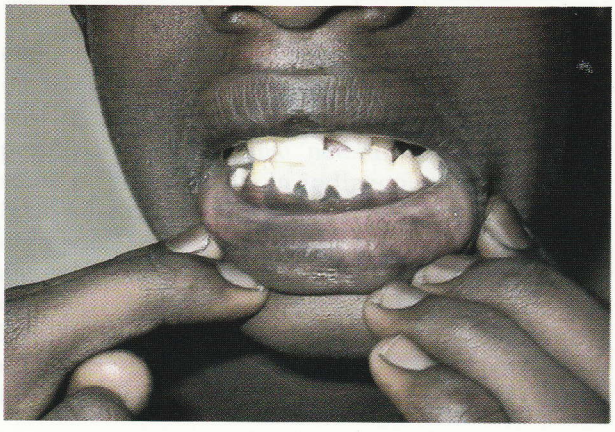


Figure 1. *Diffuse hyperpigmentation of the skin, lips, gums and finger nails.*

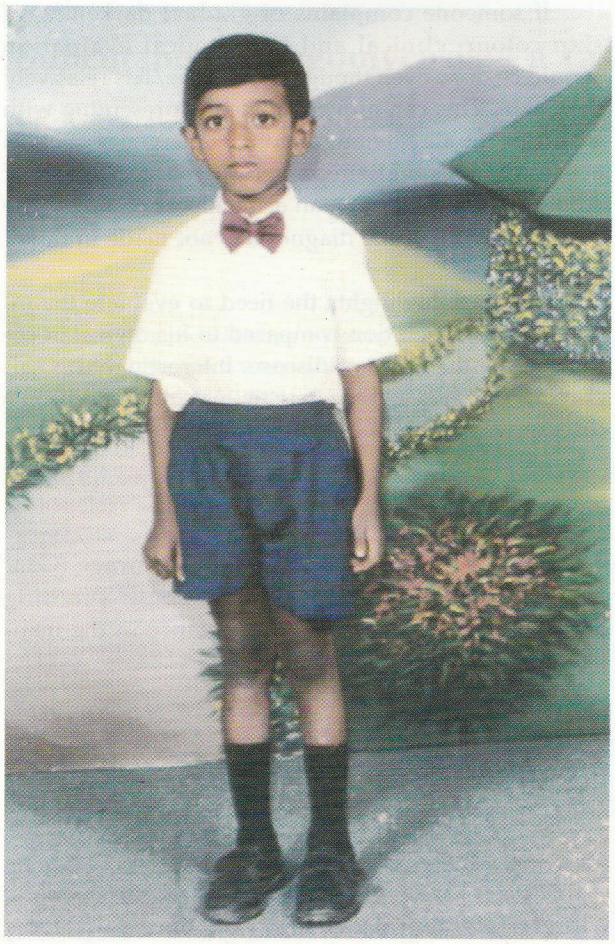
Routine biochemical investigations revealed, serum sodium 140 m.mols/L, serum potassium 4.1 m.mols/L, Hb 11.3 g/dL white blood count was 5400 mm^3 with 28% neutrophils, 55% lymphocytes and 7% eosinophils. Blood urea, fasting blood sugar, serum transaminase level, serum iron studies, serum ferritin, serum calcium, erythrocyte sedimentation rate (10 mm 1st hour) and mantuox test were normal. The telechest was normal. There was no suprarenal calcification on X-ray of erect abdomen. Ultra sound scan of abdomen revealed no supra renal mass, calcification or spleenomegaly.

Comparison of a photograph taken at 10 years of age clearly showed a marked increase of pigmentation from that time (Figure 2).

A short corticotrophin test (IV tetracosactrin 250 micrograms) showed a flat serum cortisol response (three samples of blood cortisol level at 8 am, 30 min after and 60 min after tetracosactrin were less than 1 micrograms/dl (normal value 5-25 micrograms/dl).

The patient was not in adrenal crisis but had symptoms of primary adrenal failure with a very low serum cortisol level. With the clinical history of above, examination findings and very low levels of serum cortisol, a diagnosis of primary adrenal failure, most probably due to auto immune adrenal destruction was made.

[1]*Registrar*, [2]*Consultant Dermatologist, North Colombo Teaching Hospital, Ragama.*



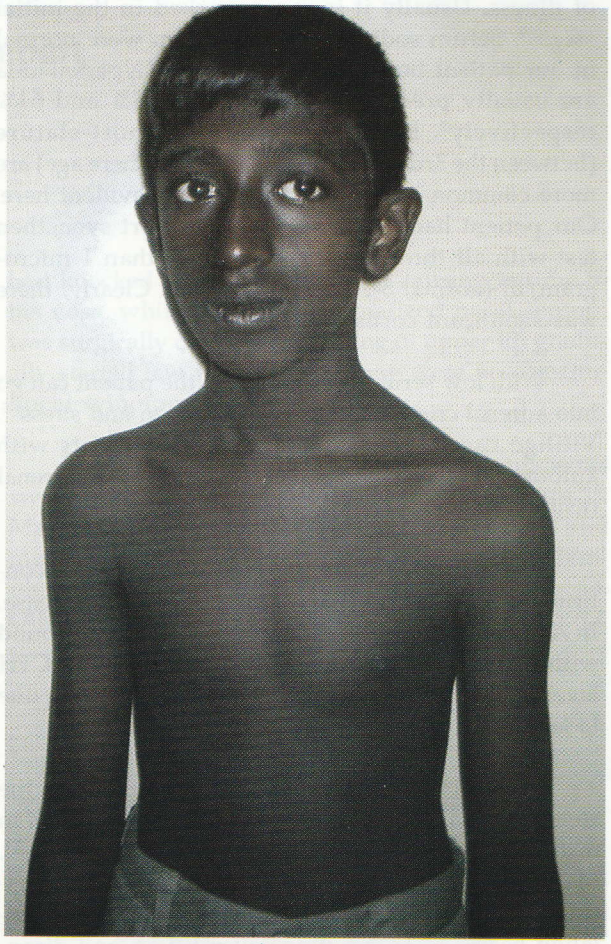


Figure 2 (a) and 2 (b). *The colour of skin at 10 years and at 13 years (after developing Addison's disease).*

Oral hydrocortisone 15 mg mane, 5 mg lunchtime, 5 mg nocte and fludrocortisone 0.05 mg daily were recommended by the endocrinologist. One week after the treatment, the symptoms such as salt and sour craving, weakness, fatigability and nausea improved. The patient and his parents were counselled regarding, the disease, how it is going to affect his life and further management, including emergency precautions.

Discussion

Chronic primary adrenal insufficiency has various types of clinical presentations in which hyperpigmentation is a characteristic physical finding (94%-98%)[1,2]. But it is difficult to assess darkening of skin in dark skinned races[1,2]. A high degree of clinical suspicion and relevant investigations become necessary for early diagnosis. Generally children's clinical presentations are similar to adults[1].

Hyperpigmentation is the most specific sign in primary adrenal insufficiency[1]. It occurs due to increased content of melanin in skin due to increased activity of melanocyte stimulation due to increased production of ACTH[2]. Generalized, buccal, anal and the mucous membrane hyperpigmentation is obviously seen in the lightly pigmented races[2]. Nail darkening is also a characteristic feature, especially longitudinal dark bands[2]. This patient had whole nail plate darkening with more marked longitudinal darkening in all the nails.

Craving of salt is also a specific symptom of primary adrenal insufficiency (16%)[1,2]. This patient had craving for salty and sour foods. In primary adrenal failure, weakness, fatigability, anorexia and weight loss can be seen in all patients (100%), nausea (86%), abdominal pain (31%)[1-3]. Weight loss is generally less marked in children[2]. Although postural hypotension is a more common presentation (80%)[1-3], it was not seen in our patient probably due to the long duration

of illness. Usually it is more marked in the initial stages[2]. Serum sodium and potassium were normal in our patient but hyponatremia and hyperkalemia are usually present at diagnosis in 88% and 64% respectively[2]. Hypoglycaemia and short stature (between the 3rd and 25th percentile for there age) are more common in children[2]. It was not evident here. Our patient had a flat response to short synacthen test with all three values being less than 1 microgram/dl (normal 5-25 microgram/dl). Clearly, there was significant cortisol deficiency.

With low serum cortisol levels the patient can go into adrenal crisis when there is infection and stress[1-5]. Vitiligo can be seen in 10%-20%[2] of patients with autoimmune types but not in other types of adrenal insufficiency.

ACTH level (> 80 ng/l)[5,4] is recommended to confirm the biochemical diagnosis of Addison's disease. In Addison's disease adrenal antibodies may be found with many cases of autoimmune adrenalitis[5]. ACTH level and anti adrenal antibodies were not done due to lack of facilities.

In the treatment, the goal should be to use the smallest dose that relieves the patient's symptoms in order to prevent weight gain and osteoporosis[1,2,5]. Morning and evening doses need to act as physiological surges of cortisol[1,2,3,5]. The cutaneous hyperpigmentation begins to fade within several days and disappears after weeks to months of adequate adrenal hormone replacement[2]. It may take more time than mentioned above, because our patient had relatively dark skin. This patient can be expected to have a normal life expectancy and can be expected to lead a fully active life, including vigorous exercise, with proper treatment[2]. During the management of this patient, we started hydrocortisone and fludrocortisone together because the latter acts as a substitute for aldosterone[1,2,4]. Glucocorticoid doses should be increased during any illness or surgery[1,2,4].

If someone complains of gradual darkening of skin colour, clinical and biochemical evaluation should be done even though he/or/she/has relatively dark skin. This can prevent someone presenting with a serious life threatening medical emergency such as acute hypoadrenatism as a first manifestation of Addison's disease during an acute illness or stress. The patient may die if the diagnosis is not made in time[1].

This case highlights the need to evaluate the increase of pigmentation compared to his own skin colour before the onset of disease. Interestingly his salt craving which is a characteristic feature of Addison's disease was associated with sour craving as well.

Acknowledgement

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