

Addison's Disease: A rare case report

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ABSTRACT

A female patient presented with progressive weakness, asthenia and generalized hyperpigmentation. The characteristic hyperpigmentation pointed towards possibility of Addison's disease which was proved by markedly decreased plasma cortisol levels, hyponatremia and hyperkalemia. This could be one of the very few cases of Addison's Disease reported.

Keywords: Characteristic hyperpigmentation, Addison's disease.

INTRODUCTION

Addison's disease (AD) is a rare primary adrenocortical deficiency mostly caused by autoimmune idiopathic atrophy or tuberculous infiltration of the adrenal gland [1]. The other causes are surgical removal, hemorrhage, metastatic invasion and fungal infection of the gland. Weakness, characteristic pigmentation of skin and mucous membrane, weight loss, anorexia and hypotension are the most common features of AD [2]. Primary adrenal insufficiency can be a life threatening disorder particularly in stressful situation, since cortisol secretion cannot be increased on demand at all. The prevalence of primary AD has been reported to be 39 to 60 per million population. Secondary adrenal insufficiency is relatively more common due to increasing therapeutic use of exogenous steroids, but this characteristically lacks pigmentary changes of AD.

CASE REPORT

A 50 years old female patient presented with progressive weakness, fatigability, anorexia and hyperpigmentation of skin since last one year. The pigmentation first started and was more on exposed parts like face, back of the hands, elbows and then it involved some covered parts, oral mucous membrane and nails as well. There was no history of tuberculosis or any other systemic major illness in past. There was no history

of any drug therapy preceding these complaints. For her general complaints she was taking treatment from physician and was hospitalized twice for profound hypotension, shock and weakness. Her blood pressure was 90/60 mmHg. All other vital parameters were within normal limits. On dermatological examination she had bluish-black hyperpigmentation of face, more on malar and forehead area, along with hyperpigmentation of the hands, forearms and palms (Figs 1 and 2). The palmar creases and nail bed also showed hyperpigmentation. Tongue and buccal mucosa also showed patchy hyperpigmentation (Fig. 3) The characteristic hyperpigmentation struck a possibility of AD which was confirmed by lowered 8 A.M. plasma cortisol level of 35.48ng/ml (normal 60-285ng/ml), hyponatremia with serum sodium level of 120 meq/litre (normal 135-155 meq/litre), hyperkalemia with serum potassium level of 5.5 meq/litre (normal 3.5-5.2 meq/litre). Other investigations like complete blood count, RBC indices were within normal limits. Peripheral smear and vitamin B12 levels were not done. An ultrasonography was done which revealed partly atrophied adrenal glands. Tuberculosis was ruled out clinically as well as immunologically. Patient was not willing for admission for the treatment with injectable hydrocortisone so, patient was started with tablet prednisolone 5 mg in morning 7 A.M. and 2.5 mg in evening with an advice to take more salts and fruits. The patient in first week started feeling better with significant improvement in general complaints and pigmentation.

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Figure 1: Bluish- black hyperpigmentation of face, more on malar and forehead area.



Figure 2: Hyperpigmentation of the hands, forearms and palms.



Figure 3: Patchy hyperpigmentation over buccal mucosa and tongue.

DISCUSSION

AD was first described by Thomas Addison in 1855 [3]. In AD, plasma Adrenocorticotropic

hormone (ACTH), Melanocyte Stimulating Hormone (MSH) and associated peptides are elevated because loss of the cortisol- hypothalamic-pituitary feedback relationship resulting in characteristic hyperpigmentation which is seen in more than 90% of patients. However, the disease can manifest without any skin changes at all or skin pigmentation may be the only presenting feature in an individual with AD [4,5]. In the secondary adrenal insufficiency which is more common and is generally seen following prolonged administration of excess glucocorticoids, ACTH and MSH level are low or normal and hence no such pigmentary changes are present. Thus, pigmentary changes are one of the differentiating features between primary and secondary adrenal insufficiency. The hyperpigmentation is one of the most striking and commonest feature of AD [1]. It is because of compensatory over secretion of MSH from pituitary in response to poor adrenal secretions. This pigmentation is initially more on sun exposed parts, scars, folds, palmar creases and mucous membranes but may become generalized. The pigmentation may take a form of dark-tan in light colored individuals to bluish-black pigmentation in dark colored patients. 8 AM serum cortisol levels and/or ACTH stimulation either by 8 hour infusion or injecting synthetic ACTH are the definitive mode of investigations. Besides glucocorticoids there is also deficiency of mineralocorticoid and aldosterone in some patients which results in sodium depletion, hyperkalemia, hypotension and sometimes acidosis. Some patients may have associated severe gastrointestinal complaints. Drug of choice in AD is hydrocortisone. But management also consists of a physiological replacement of steroid in the form of equivalent doses of prednisolone 7.5 mg, which may be subdivided into 5 mg in morning and 2.5 mg in evening. If available fludrocortisone, a mineralocorticoid may be added to this regimen, besides this patient may be educated regarding the nature of the disease, risk and care to be taken during situations like infections

This could be one of the very few cases of AD reported.

CONSENT

The examination of the patient was conducted according to the Declaration of Helsinki principles. Written informed consent was obtained from the patient for publication of this article.

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