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ADDISONIAN PIGMENTATION: A CASE STUDY

Dermatology

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ABSTRACT

Addisonian pigmentation is characterised by diffuse pigmentation of skin and mucosae with accentuation over the sun exposed, flexural and periungual areas. We report "Addisonian pigmentation" in a 4-year-old boy secondary to non-classical adrenal hyperplasia and possibly ectopic ACTH secretion. This case underscores the significance of hormonal evaluation in cases of diffuse hyperpigmentation in childhood.

KEYWORDS

INTRODUCTION

Addisonian pigmentation is any form of diffuse skin hyperpigmentation of skin mimicking Addison's disease more pronounced on sun-exposed areas, flexural folds, periungual areas and skin creases including palmar creases[1] The exact prevalence in childhood is not known as it is a rare entity. There are various causes of Addisonian pigmentation like endocrinal causes of Addison's disease, cushing's syndrome, thyrotoxicosis, nutritional deficiencies like vitamin B12, folic acid deficiency. Some other causes include drugs like phenytoin, cytotoxic drugs, amiodarone, exposure to chemicals like arsenic, bismuth and other rare syndrome like Lauzier hungiker syndrome, POEMS syndrome.[1] We are describing here a classical case of addisonian pigmentation probably due to non-classical CAH with ectopic ACTH secretion still under evaluation.

CASE REPORT

A 4-year-old male child presented with complaints of generalised darkening of the skin, oral mucosa and nails since $1^{1/2}$ years of age. Pigmentation first started over face and upper limbs and gradually progressed to involve the rest of body, all nails and oral mucosa in next 5-6 months. Patient previously had history of 3 episodes of febrile seizures in last 2 years. Physical examination revealed diffuse dark greyish brown pigmentation of skin all over the body, more marked over extensors with less pigmentation over axillae and popliteal fossa. (Fig1) There was darkening of palmar creases, blackish discolouration of both finger and toe nails, brownish black discolouration of sclera, hard palate, gums and tongue. (Fig 2, 3, 4 and 5). Height for age was in 92th centile and testis size was normal for age. Investigations like complete haematological, biochemical and radiological including serum vitamin B12, thyroid function, skeletal survey, USG abdomen, CECT abdomen were within normal limits. His serum cortisol level was low (< 0.50 microIU/ml), ACTH levels was high (145.80 pg/ml) and ACTH stimulation test (basal - 3.54 µg/dl, 30 mins - 4.00microg/dl) was negative suggesting primary adrenal insufficiency. Patient's mantoux test was positive (16x10mm). Serum testosterone and 17-OH progesterone was normal. MRI brain revealed some small foci of hyperintensities in frontoparietal and periventricular areas with normal pituitary gland. Skin biopsy taken from abdominal skin showed increase in melanisation in basal cell layer. Superficial dermis showed scanty perivascular inflammatory infiltrate. Patient was started on hormonal therapy (oral hydrocortisone) 2.5 mg tds and is on regular follow up. His pigmentation has decreased on both cutaneous, nail and oral mucosal and he is continuing with the medication.

DISCUSSION

Addisonian pigmentation is usually an early manifestation seen in primary adrenal insufficiency which may precede other manifestation by 10 years.[2] Hyperpigmentation of the skin and mucosal surface, the most specific sign of primary adrenal insufficiency occurs in upto 92% of the patients.[3] Along with the diffuse hyperpigmentation, there occurs increase in pigmentation commonly at the flexures, sites of pressure and friction, palmar and plantar creases, scars, sun exposed areas, genitilia and areola. Pigmentation of oral mucosa is classical, blue black or brown hyperpigmented macules present in spotty or streaked configuration over tongue, gingival, buccal mucosa and hard palate. After ruling out various other causes of addisonian pigmentation like cushing disease, hyperthyroidism, vitamin B 12 deficiency, drug intake and after the investigations we came to a conclusion of primary adrenal insufficiency as the cause of addisonian pigmentation. Classical congenital adrenal hyperplasia is the most common cause of primary adrenal insufficiency,[4]but in our case 17-OH progesterone, skeletal survey, normal genitalia ruled out classical CAH, so the diagnosis of non-classical CAH with ectopic ACTH production was kept, since ACTH was high although pituitary size was normal.[5,6] Hyperpigmentation occurs secondary to overproduction of pro-opiomelanocortin by-product beta lipoprotein, which is secreted in excess amount concomitantly with corticotrophin from the pituitary gland because of lack of feedback inhibition seen in adrenal insufficiency states. The pigmentation usually improves with hormonal therapy.[4]Our case typically had all the features of addisonian pigmentation except flexures and genitalia didn't show accentuation. Patient was started on hormonal therapy since past six months and 30% improvement was noted in the pigmentation at present.

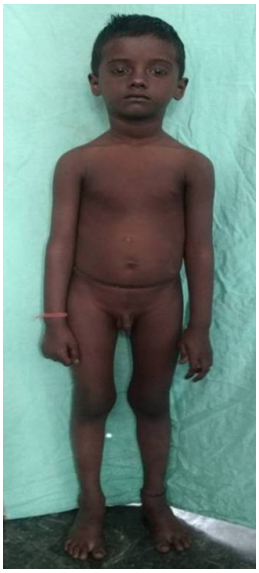

Figure 1: Diffuse hyperpigmentation all over body

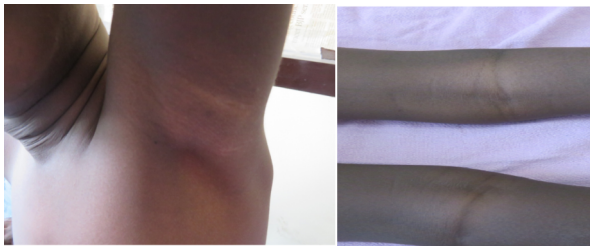

Figure 2: Less pigmentation over axillary folds and popliteal fossa

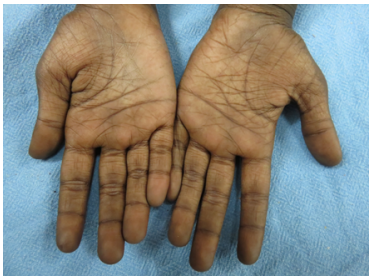

Figure 3: Darkening of palmar creases

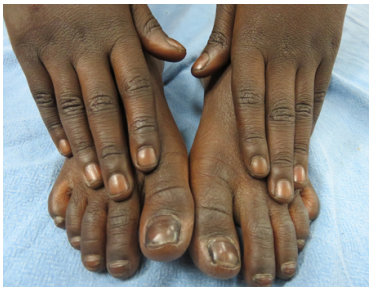

Figure 4: Blackish discolouration of finger and toe nails

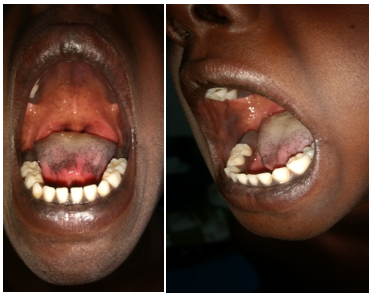

Figure 5: Brownish black discolouration of hard palate, buccal mucosa and tongue.

CONCLUSION

Diffuse pigmentation in childhood can be multi factorial.1 However, systemic causes like endocrinal abnormalities and nutritional deficiencies are common cause, addisonian pigmentation can be an early marker of primary adrenal insufficiency, it is of vital importance to investigate for any underlying hormonal abnormality in order to initiate early treatment with hormonal therapy, which would manage the pigmentation as well as the systemic manifestation effectively and timely.

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