

## Addisonian pigmentation of tubercular etiology-A case report

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### Abstract

Addisonian pigmentation is defined as hyperpigmentation of both exposed and unexposed parts of the body and it typically occurs in primary adrenal insufficiency.

We hereby report a case of a 15 year old boy who presented with progressive hyperpigmentation of the face, trunk, chest, abdomen and extremities which was more accentuated over the sun exposed areas. He also had pigmentation of nails, oral and genital mucosa, palms and soles with accentuation over palmar creases. Routine blood investigations were found to be normal. His serum electrolytes showed hyponatremia and hyperkalemia with evidence of hypocortisolemia and increased serum ACTH. Chest x-ray was suggestive of pulmonary tuberculosis, sputum AFB was positive and Mantoux reaction was strongly positive. CT scan showed bilateral adrenal calcification. All these investigations confirmed primary adrenal insufficiency due to tuberculosis and ATT was initiated along with steroid replacement therapy.

**Keywords:** Addisonian pigmentation, ACTH, hyperpigmentation.

### Introduction

Generalised skin and mucosal hyperpigmentation is a tell-tale sign of Primary adrenal insufficiency which occurs in 95% of cases, thus differentiating it from secondary and tertiary hypoadrenalism.

It often precedes other manifestations by months to years and it occurs when 90% of the adrenal gland is destroyed.<sup>1</sup> Most common cause of adrenal insufficiency in developed countries is auto immune adrenalitis whereas in developing countries it is Mycobacterium tuberculosis. Patient needs lifelong steroid supplementation as ATT does not improve adrenal function.

### Case Report

A 15 year old boy presented with diffuse pigmentation all over the body for the past 1 year. Pigmentation initially started over the face and it gradually spread to involve extremities, back, chest and abdomen with 2 months history of easy fatigability, giddiness, loss of weight, loss of appetite, cough with expectoration and fever. There was no significant medical or surgical illness in the past.

On systemic examination, he was afebrile with significant pallor.

On dermatological examination diffuse hyperpigmentation was present all over the body with accentuation on all the finger and toe nails, palms and soles, oral and genital mucosa and intense pigmentation noticed over the sun exposed areas, palmar creases and genital skin. (Fig. 1-6).

All the routine basic blood investigations were done. His Hemoglobin was 8.9 gms/dl, peripheral smear showed microcytic, hypochromic anaemia, ESR was 60 mm/hr, Serum electrolytes showed hyponatremia and hyperkalemia. Chest X ray PA view showed right lower zone consolidation. USG abdomen

and pelvis was normal. Serum cortisol at 8 am - 0.87 mcg/dl, significantly reduced (normal range 4.3-22.4 mcg/dl). Basal serum ACTH was raised to 1250 pg/ml (normal range 7.2-63.3 pg/ml). Serum aldosterone - 1.44 ng/ml, was low (normal range-2.52-39.2 ng/ml). CT chest revealed right middle lobe consolidation with multiple calcified mediastinal lymph nodes, suggesting the possibility of pulmonary tuberculosis. Contrast enhanced CT abdomen and pelvis showed few tiny calcifications in bilateral adrenal glands. Sputum AFB 1 & 2-positive and Mantoux test-positive (>15mm).

Based on clinical, biochemical and imaging studies, final diagnosis of Primary adrenocortical failure due to adrenal tuberculosis along with pulmonary tuberculosis was made. He was started on Category 1 ATT along with steroids.



**Fig. 1**



Fig. 2



Fig. 5



Fig. 3



Fig. 6



Fig. 4

### Discussion

Primary adrenocortical failure or Addison's disease is a rare endocrine disorder, named after Thomas Addison, the British physician who first described the condition in 1855. The term "Addison's disease" does not imply an underlying disease process, although all of Addison's six patients had tuberculosis of the adrenal gland.<sup>2</sup>

Addison's disease manifests as hyperpigmentation of skin and mucosa, weakness, behavioural change, fatigue, anorexia, nausea, vomiting and abdominal pain.

Addison's disease is caused by congenital adrenal hypoplasia, infections (mycobacterium tuberculosis, HIV, CMV, Cryptococcus neoformans, Histoplasma capsulatum, Toxoplasma gondii and Pneumocystis jirovecii), autoimmune, traumatic, iatrogenic (surgery, medication), vascular (Hemorrhage, emboli), metabolic (amyloidosis), sarcoidosis and neoplastic (primary, metastatic).

With destruction of adrenal cortex, feedback inhibition of the hypothalamus and anterior pituitary is interrupted and ACTH is secreted continuously. ACTH and melanocyte stimulating hormone both are cleaved from the same progenitor hormone, Pro opio melanocortin. When ACTH is cleaved from the prohormone, MSH is concurrently released and it stimulates the enzyme tyrosinase in melanocytes resulting in pigmentation.

Generalised hyperpigmentation is a hallmark and it is most pronounced on the sun exposed areas, areas subjected to repeated frictions, palmar and plantar creases, the normally pigmented sites (nipples and genitalia), recently acquired scars and nevi, mucous membranes<sup>3</sup> (buccal,conjunctival,glans and vaginal). New pigmented nevi may appear. The hairs become more pigmented. Nails may show diffuse pigmentation or longitudinal melanonychia.This pattern of pigmentation often referred as "Addisonian pigmentation". The onset of pigmentation may be insidious and it is epidermal melanotic hyperpigmentation.

Similar pattern of pigmentation has been reported in Nelson syndrome, acromegaly, cushing's syndrome, thyrotoxicosis, pheochromocytoma, ectopic ACTH producing tumours and exogenous administration of ACTH.

Among infections, tuberculosis accounts for about 20-30% of cases of addison's disease and it is associated with bilateral adrenal gland involvement. Adrenal gland is enlarged in early tuberculosis whereas calcification is noted in remote infection. Adrenal tuberculosis is seen in 6% of patients with active pulmonary tuberculosis.

Since the presentation was strongly suggestive of adrenal tuberculosis with active extra adrenal tuberculosis, adrenal biopsy was not performed.

Patient should be started on steroid replacement therapy despite exacerbation of pulmonary tuberculosis to prevent addisonian crisis as ATT does not improve adrenal function.<sup>5</sup> Since rifampicin is a potent enzyme inducer, steroid dose needs to be increased in order to maintain adequate steroid replacement therapy. Steroid replacement therapy gradually improves the skin hyperpigmentation but the mucosal pigmentation may persist.

### Conclusion

Whenever patients with addisonian pigmentation is encountered, tuberculosis must be ruled out in developing countries like India.

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