

Case Report

Addison's disease may present to a dermatologist as melasma

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Abstract Addison's disease is not a very common medical illness that can present to a dermatologist in different ways. It can start as an obvious pigmentary disorder or it may never involve skin. We present a case of a young male who reported to dermatology outdoor with a typical melasma-like eruption over his face. Shortly after this pigmentation, he started having generalized weakness, repeated vomiting and weight loss. He was evaluated by physician and turned out to be a case of Addison's disease with melasma-like pigmentation. He was then managed accordingly.

Key words

Addison's disease, melasma, pigmentary disorders.

Introduction

Addison's disease was first described in 1855 by Thomas Addison as a result of adrenal insufficiency.¹ The basis of Addison's disease has dramatically changed from an infectious cause to autoimmune pathology since its initial description. However, tuberculosis is still the predominant cause of Addison's disease in developing countries. It can occur in persons of any age; however, it is most common in people aged 30-50 years and more so in females. Symptoms are often nonspecific and include fatigue, weakness, anorexia, nausea, abdominal pain, gastroenteritis, diarrhea, and mood lability. Weakness and weight loss of 1-15 kg are universal features of Addison's disease in the adults.^{1,2} Dermatological findings include hyperpigmentation of the skin and mucous membranes, decreased pubic and

axillary hair in women and vitiligo. Pigmentary changes are supposed to be caused by the stimulant effect of excess adrenocorticotrophic hormone (ACTH) on the melanocytes to produce melanin. Hyperpigmentation of the skin and mucous membranes often precedes all other symptoms by months to years. The presence of normal-appearing skin does not exclude the diagnosis. Vitiligo may also be seen in association with hyperpigmentation in idiopathic autoimmune Addison's disease due to the autoimmune destruction of melanocytes.¹⁻³ Hyponatremia, hyperkalemia and hypotension are commonly seen while hypercalcemia is an uncommon finding. The preliminary test for adrenal insufficiency is the measurement of serum cortisol levels from a sample of blood obtained in the morning between 6:00 and 8:00 am (when the levels are highest). The hypothalamic-pituitary axis can be evaluated by using 3 tests: the ACTH (Cortrosyn®) stimulation test, the insulin tolerance test, and the metyrapone test. Once the diagnosis of

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adrenal insufficiency is confirmed, the site of the defect in the hypothalamic-pituitary axis should be determined by using ACTH sampling, ACTH provocative testing, or corticotrophin-releasing hormone (CRH) provocative testing. Both computed tomography (CT) and magnetic resonance imaging (MRI) demonstrate a diminished adrenal gland in patients with autoimmune destruction and an enlarged adrenal gland in patients with infection.^{2,4,5} Tissue cultures in patients with tuberculosis reveal acid-fast bacilli. The adequate replacement of glucocorticoids and mineralocorticoids is the primary goal of treatment.¹ Recent studies show that dehydroepiandrosterone therapy improves the patient's quality of life.⁶ With optimum dosing the glucocorticoids are adequately replaced with minimal adverse effects. The aim of presenting this case is to emphasize the dermatological manifestations of the disease and to refresh the knowledge of the dermatologists about this potentially grave medical illness.

Case history

A 23-year-old young man presented to medical outdoor of PAF Hospital, Sargodha with 5 months history of progressive weight loss, poor appetite, nausea, occasional vomiting and facial pigmentation. He noted a weight loss of 12kg during this period and used to vomit out occasionally, specially when under stress. His symptoms had aggravated during the last seven days and he had become drowsy at the time of examination. There was no history of fever, cough or night sweats. A couple of months before these symptoms, he started developing pigmentation over his nose and both cheeks that became prominent in about two weeks time (**Figure 1**). He reported to dermatologist

for this problem and was given some local cream to apply along with sunscreen. On examination he was found to be weak and lethargic with marked pallor. His blood pressure was 90/60 mm of Hg and there was no postural drop. Systemic examination was unremarkable except he was drowsy but arousable. Hemoglobin was 13.3 gm/dl, ESR 20 mm fall after first hour, bilateral cardiophrenic angles were blunt on PA view of the chest X-ray. His metabolic profile including serum urea, creatinine, electrolytes and blood sugar fasting, all were within normal limits. Early morning serum cortisol level was 2.5 microgram/dl (well below the normal range of 9-25). Short synacthen test was consistent with hypocortisolism. CT scan of the abdomen showed bilateral adrenal enlargement and hepatomegaly (**Figures 2 and 3**). CT scan and MRI of the brain were normal. HBsAg, Anti HCV, HIV serology, ANA and RA factor were negative. On the basis of history, clinical findings and lab investigations he was diagnosed to be suffering from Addison's disease secondary to disseminated tuberculosis involving adrenals. Dermatological examination revealed typical melasma-like pigmentation over bridge of the nose and prominence of the cheeks (**Figure 1**). There was no pigmentation in palmar creases, flexures, nipples, knuckles, genitalia or any body scars. Oral mucosa was also normal looking. No dyspigmentation was found anywhere else over the body. Hair distribution and density was normal. He was started on antituberculosis treatment including rifampicin, isoniazid, pyrazinamide, myambutal and amikacin along with cortisol replacement in the form of methyl prednisolone 10 mg in the morning and 5mg in the evening. His symptoms subsided within

days. Appetite improved, nausea and vomiting relieved and he started gaining weight. Later



Figure 1 Typical (centrofacial and malar) melasma-like pigmentation.

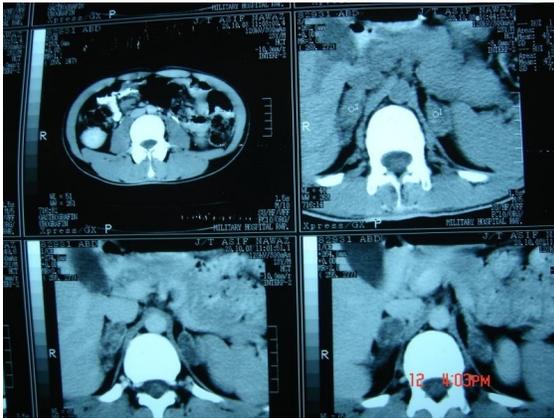


Figure 2 Abdominal CT scan sections showing bilateral adrenal enlargement.



Figure 3 Close up of a single section showing bilateral adrenal enlargement.

the dose of steroid was adjusted to 7.5 mg per day. Local treatment for facial pigmentation was also affective in relieving the problem. Antituberculosis treatment was continued for nine months. He was instructed to increase the dose of steroid during stress and wear a bracelet or locket with his diagnosis and treatment engraved on it.

Discussion

Generally, it is the combination of skin and mucosal pigmentation with gut disturbances and weight loss that carries high predictive value in diagnosis of Addison's disease, while individual sign and symptom has poor differentiating value. However, the disease can manifest without any skin changes at all or skin pigmentation may be the only presenting feature in an individual with Addison's disease.^{2,7-9} Hyperpigmentation of the skin is considered a hallmark of Addison's disease and is present in 95% of patients with chronic primary adrenal insufficiency and this can occur in flexural areas, skin creases, nipples, pigmentation, scars, pressure points, perineum, and buccal mucosa. It can even precede other manifestations of the disease, such as fatigue, weakness, progressive asthenia, and orthostatic hypotension.^{3,7-9} Occasionally, pigmentation may be generalized.¹⁰ Melasma-like pigmentation without involving any other area of the body is an uncommon finding in Addison's disease.³ Our patient initially presented with only melasma like hyperpigmentation and Addison's disease were never thought of at that time, but when it was soon followed by other suggestive manifestations we correlated it and considered it to be one of dematological presentation of the disease. In our part of the world, tuberculosis is still the common cause of

Addison's disease, and bilateral enlargement of the adrenals on CT scan pointed out towards the infectious (tuberculous) etiology.

Conclusion

Every melasma-like pigmentation is not always melasma. We, dermatologists, should have a high index of suspicion for a rare and potentially dangerous medical illness i.e. Addison's disease.

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