

A Bearded Indian Female: A Rare Presentation of Cushing's Syndrome

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A 16-year-old Indian girl presented with increased facial hair growth, weight gain, amenorrhea and generalized weakness for the last 3 months. On examination she was found to have severe hirsutism, her modified Ferriman-Gallwey score was 24/36, she had broad purple striae on abdomen, hypertension and proximal myopathy. On investigations, the patient was found to have ACTH dependent Cushing's syndrome. She had non-suppressed overnight and low dose dexamethasone suppression tests

with ACTH (Adrenocorticotrophic Hormone) level of 135pg/ml. In the high dose dexamethasone suppression test, the serum cortisol level did not suppress to > 50% of the basal value. Imaging of the pituitary revealed pituitary hyperplasia without any tumor and CT scan of the adrenals revealed bilateral adrenal hyperplasia. CT scans of the neck, chest and abdomen were performed to determine the ectopic source but the source could not be found. Based on the above investigations a diagnosis of Cushing's syndrome due to a probable ectopic ACTH source was maintained. There are no facilities for PET scan



Figure 1. Figure showing marked hirsutism in a case of Cushing's syndrome

and other functional imaging procedures and hence, these were not performed.

Hirsutism in Cushing's syndrome has a prevalence of 81% and discriminant index of 2.8.¹ Features with high discriminant index of Cushing's syndrome include bruising, myopathy, hypertension and plethora. Most common form of hirsutism is vellus hypertrichosis on the face which should be distinguished from darker, terminal differentiated hirsutism. Our patient had terminally differentiated hirsutism. In approximately 10% of cases, Cushing's syndrome is associated with ectopic ACTH secreting tumors.² In one of the studies, hirsutism was found to be present in 20% cases of ectopic Cushing's syndrome.³ Hirsutism is postulated to be due to excessive production of adrenal androgens by the excess ACTH production. Unlike Cushing's disease, with female preponderance, this syndrome is more common in men. We present a case with extensive hirsutism rarely seen in a patient with Cushing's syndrome.

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