

Familial anterior cervical hypertrichosis

Sir,

Hypertrichosis is a condition of excessive hair growth independent of associated underlying endocrine disorder. Primary hypertrichosis has been classified by the extent of distribution in localized or generalized forms. Primary localized hypertrichosis may occur as hypertrichosis cubiti (elbows), posterior cervical

There is little evidence of any increased morbidity or

hypertrichosis, faun tail deformity, or anterior cervical hypertrichosis (ACH). ACH is a very rare form of primary localized hypertrichosis. It consists of a tuft of terminal hair on the anterior neck just above the laryngeal prominence.^[1,2] Familial ACH (FACH) is a variant of ACH and, as far as we know, only 18 cases of five families have been previously reported.^[1] This condition has been described in association with other systemic disorders, but the concomitance with cutaneous disease has not reported.^[3,4] In this article we present the case of a family with three members affected and report the concomitance with new systemic diseases and morphea.

We report the case of a 56-year-old woman with history of diabetes mellitus, hypertension, and thrombocytopenia who was evaluated in our dermatology unit for pruritic hyperpigmented plaques with skin thickening on abdominal and lumbar areas [Figure 1] and a congenital tuft of terminal hair on the anterior aspect of the neck. A skin biopsy from the abdomen and lumbar area was compatible with morphea. Laboratory tests showed liver function abnormalities and abdominal computed tomography scan showed portal hypertension and splenomegaly. These findings were compatible with chronic liver disease. Immuno-serologic tests were normal. Two of her three daughters also had the congenital patch of terminal hair on the anterior neck [Figure 2]. One of them, a 27-year-old female with Down's syndrome, presented dysesthesia of the hands and physical examination revealed overlapping of fourth over third toe of both feet [Figure 3]. Another daughter, a 35-year-old female, with history of great vessel transposition, corrected when she was a new born, had a normal physical examination. No other abnormalities were detected. There was no history of consanguineous marriages in the family.

FACH can present as an isolated defect or in association with various abnormalities of other organ systems.^[2-5] Most of the families reported had not concomitant diseases.^[2,5] Trattner *et al.* described an Arab family with three affected relatives, all of them with peripheral neuropathy and hallux valgus, and one with atrophy of optic nerve and macular dysfunction.^[3] Tsukahara *et al.* reported a Japanese family with seven members in three generations with ACH, one of them with Turner's syndrome.^[4] In our case, only dysesthesia was observed in the patient with Down's syndrome.



Figure 1: The mother with hyperpigmented plaques with skin thickening on the abdominal area



Figure 2: The mother and her two daughters with congenital patches of terminal hair on the anterior neck



Figure 3: Association with overlapping of fourth over third toe of both feet in one of our patients

This patient also presented toes overlapping which was previously described in a child with sporadic ACH.^[1] The concomitance of FACH with great vessel transposition, chronic liver disease, and morphea has never been described before. They are incidental associations, but we suggest a careful evaluation of ACH patients to exclude any systemic disease.

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