Images of Hypertrichosis

January 02, 2009

Hypertrichosis refers to the increased growth of vellus or other hair at inappropriate locations beyond the normal variation for a patient’s reference group. The affected areas have a greater number of hair follicles than is normal for the body site. The condition is unrelated to androgen excess and unaccompanied by virilism or menstrual abnormalities.

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Congenital generalized hypertrichosis may result from maternal ingestion of medications (such as minoxidil, phenytoin, and diazoxide) or alcohol, or it may be inherited in an autosomal dominant pattern (eg, hypertrichosis lanuginosa, universal hypertrichosis, or hypertrichosis with gingival hyperplasia) or an X-linked dominant pattern. Generalized hypertrichosis is also a feature of Brachmann-de Lange syndrome (also known as Cornelia de Lange syndrome) (Figure 1), as well as many other syndromes: Ambras, Rubinstein-Taybi, Coffin-Siris, Laband, Hunter, Hurler, Sanfilippo, Bloom, Seckel, Gorlin, Cowden, Seip-Berardinelli, Donohue, Barber-Say, stiff skin, Winchester, trisomy 18, trisomy 3q, and Schinzel-Giedion. Congenital generalized hypertrichosis may also be familial, with a multifactorial mode of inheritance (Figure 2).
Figure 2 – A 7-year-old Spanish girl with congenital generalized hypertrichosis is shown. Note the hairy forearms. Her 37-year-old father also has hypertrichosis.

Congenital localized hypertrichosis is a notable feature of congenital melanocytic nevi (Figure 3), congenital Becker nevi, nevoid hypertrichosis, nevus pilosus, smooth muscle hamartomas, plexiform neurofibromas, and linear epidermal nevi. This localized type may be associated with an underlying spina bifida occulta, diastematomyelia, or kyphoscoliosis. Hypertrichosis of the pinnae is most commonly seen in patients with XYY syndrome and in infants of mothers who have diabetes mellitus (Figure 4). Hypertrichosis cubiti (hairy elbows) and anterior cervical hypertrichosis are associated with both autosomal dominant and recessive inheritance patterns, although they may be idiopathic.

Figure 3 – An 18-year-old boy has a congenital melanocytic nevus with characteristic hypertrichosis on the upper right arm. (Courtesy of Robert P. Benteau, MD.)

Acquired generalized hypertrichosis is most frequently caused by medications, such as phenytoin (in 5% to 10% of patients), cyclosporine, danazol, minoxidil, penicillamine, diazoxide, psoralens, anabolic agents, corticosteroids, acetazolamide, hexachlorobenzene, and streptomycin. Typically, phenytoin-induced hypertrichosis occurs to a greater extent on the extremities than on the face and trunk (Figure 5). In contrast, minoxidilinduced hypertrichosis characteristically involves the face, shoulders, and extremities. Drug-induced hypertrichosis usually resolves within months after the offending agent is discontinued. Acquired generalized hypertrichosis may also result from starvation, neoplasm, encephalitis, multiple sclerosis, acrodynia, porphyria, dermatomyositis, and hypothyroidism.
Acquired localized hypertrichosis arises after chronic irritation, friction, or inflammation and may develop around chickenpox scars, the sites of insect bites, at the periphery of burned skin, and on the legs after radical inguinal lymphadenectomy. The condition has also been noted after topical use of hydrocortisone, after the application of a plaster of Paris cast or fiberglass cast, after x-ray or UV irradiation, and in patients with mental illness who repeatedly bite or scratch their hands and arms. The condition has also been noted after topical use of hydrocortisone, after the application of a plaster of Paris cast or fiberglass cast, after x-ray or UV irradiation, and in patients with mental illness who repeatedly bite or scratch their hands and arms.2,15-17

CLINICAL EVALUATION
Hypertrichosis must be distinguished from hirsutism, or excessive male-pattern hair growth that results from an excess of androgens. Hirsutism is characterized by excessive coarse hair on areas of the body that are sensitive to androgens and where there is normally very little hair, especially in females. These androgen-dependent areas include the upper lip, chin, cheeks, chest, lower abdomen, and inner aspects of the thighs. Other signs and symptoms of androgen excess (or virilism) include clitoromegaly, acne, frontal balding, increased muscularity, loss of female body contour, deepening of the voice, increased sebum output, and changes in libido.

The history should include the age at onset and the site and progression of the increased hair growth. Ask about the use of medications, onset of puberty, menstrual irregularities, concomitant illnesses, past health, family history of increased hair growth, and ethnic background. Document the area of increased hair growth. If it occurs in androgen-dependent areas, abnormalities of the pituitary gland, adrenal glands, and gonads must be ruled out—particularly in patients with signs of virilism. No laboratory testing is necessary for patients with hypertrichosis. However, if hirsutism is suspected, serum levels of dehydroepiandrosterone, dehydroepiandrosterone sulfate, and testosterone and urinary 17-ketosteroids should be measured.18,19

MANAGEMENT
Treat the underlying cause whenever possible. Offending pharmacological agents should be discontinued.
Patients with hypertrichosis can conceal the hair with makeup or lighten it with over-the-counter bleaching cream. Mechanical methods for removing unwanted hair include cutting with scissors, shaving with a razor or electrical shaver, plucking, chemical or wax epilation, electrolysis, intense light therapy, and laser hair removal.18,20
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Published on Psychiatric Times
(http://www.psychiatrictimes.com)

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