Read Item - Circumscribed Hypertrichosis

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Abstract: Doctor's resource on various forms of limited or circuscribed areas of hairiness.

Definition

Hypertrichosis occurs in both sexes. It is defined as excessive growth of hair on sites not normally hairy. The hairs may be of the lanugo or vellus type and some of these hairs evolve into terminal hairs. Hair may be excessive in density or length and the condition may be localised or generalised. The process is not androgen dependant.

The normal hair patterns have previously been discussed. Localised hypertrichosis occurs as an isolated finding or in association with a number of neurological, endocrinological and naevoid disorders.

Hairy Ears

Hereditary development of coarse hairs on the pinna of the ear occurs most frequently and with the longest and densest hairs in Indians. It first appears around 20 years and thereafter increases in incidence and severity such that by the age of 70 years, 70% of men in Madras are affected. Normally only males are affected and hypertrichosis of the pinna is believed to be a sex linked recessive trait. The combination of hairy ears and azospermia has been proposed as the only known Y chromosome linked disorder.

Congenital hairy ears occur in babies born to diabetic mothers (unrelated to maternal diabetic control), babies with XYY syndrome and in some normal infants. Acquired hairy ears have been observed in AIDS, and often accompany trichomegaly of the eyelashes and eyebrows.

Hairy Eyebrows

Extremely long eyebrows or eyelash hair or eyebrows that join in the centre or synophrys are usually familial. In infants it may occur as a localised problem or rarely as part of a syndrome such as Cornelia de Lange, Rubinstein-Taybi, congenital hypertrichosis lanuginosa or congenital trichomegaly with dwarfism, mental retardation and pigmentary degeneration of the retina.

Long eyebrows are commonly the most pronounced aspect of the drug-induced acquired hypertrichosis as occuring with cyclosporin, minoxidil and diazoxide. AIDS can also induce eyebrow growth.

Hairy Palms

Localised patches of hair on the palms and soles have been reported in two families as an autosomal dominant trait. The hypertrichosis was associated with increased pigmentation and textural changes in the skin. This could represent hypertrichosis in association with a connective tissue hamartoma as histology shows an increased amount of disorganised elastic tissue.

Hairy Elbows

In familial hypertrichosis cubiti, excessive hair growth develops in infancy over the lower third of the upper arm and the upper third of the forearms. The hypertrichosis is progressive for a few years before partially regressing around puberty. The mode of inheritance is uncertain, and both autosomal recessive and autosomal dominant patterns have been suggested. An association with short stature is seen in 10%. An increased proportion of anagen hairs (>90%)are seen histologically and suggests this condition is caused by prolongation of anagen.

Becker's naevus

This condition is an androgen-mediated hyperplasia that first appears at puberty as a

unilateral localised area of hyperpigmentation. In 50% of cases it is followed by hypertrichosis. The hairs are predominantly terminal hairs. Acne occasionally develops within a BeckerÕs naevus. The incidence is about 0.5% and the male to female ratio is 10:1. About half occur on the shoulder and the rest on the trunk and a few on the limbs or face. Multiple lesions are rare as are familial cases. The term naevus requires qualification, as there is no increase in the number of melanocytes and histology simply shows acanthosis with basal hyperpigmentation. An increase in smooth muscle bundles in the dermis is sometimes noticed.

Uncommon associated abnormalities include ipsilateral breast hypoplasia, spina bifida, pectus carinatum, accessory scrotum, morphoea and ipsilateral leg hyperplasia. Electrolysis has been used with mixed success to treat the hypertrichosis, but the pigmentation is very difficult to treat.

Congenital melanocytic naevus
Both congenital and acquired naevi can develop hypertrichosis, usually consisting of terminal hairs. The presence of hairs does not help to determine if a pigmented lesion is benign or malignant as a melanoma may arise at the edge of a hairy naevus and in addition loss of hairs tends to be a late feature in the development of melanoma. Congenital pigmented hairy naevi are probable the commonest cause of localised hypertrichosis in infancy. The German term 'Tierfellnevus' (animal skin naevus) has been used to describe a particularly hairy variant and is an apt morphological description.

Fauntail
A congenital midline dorsal patch of hypertrichosis or fauntail is an important clue to a possible occult spinal abnormality that may in time produce neurological deficits. It consists of a tuft of long silky hair in the sacral area usually overlying normal skin.

Occasionally there will be a sinus or a dimple within the fauntail and rarely a lipoma or a capillary naevus will be found beneath it. Sinuses are usually 1 to 2 mm wide and can connect directly with the spinal cord. They are a potential portal of entry for infection and they can be associated with a cystic dilatation within the spinal cord that produces neurological deficits.

Spinal dysraphism (failure of spinal fusion) occurs beneath the fauntail four times more frequently in girls than boys. In association with this the cord can be transfixed by a bony spicule (diastematomyelia), or the cauda equina may be tethered by fibrous cords. With time this produces traction damage to the cord as a result of differential growth of the vertebra and the cord. Neurological deficits are not present at birth, but gradually develop over the first five years. If identified early permanent damage can be prevented by prophylactic surgery.

Midline hypertrichosis can occur anywhere along the back associated with cervical, thoracic or lumbar dysraphism. The stimulant for hair growth is presumably a growth factor secreted by neural tissue, however none has been identified.

Hair Collar Sign
In 1989 Commen described a distinctive collar of hypertrophic hairs that palisaded around a bald nodule containing heterotopic brain tissue. The nodule usually overlies an embryonic fusion line. The collar of long terminal hairs grows faster, earlier and longer than the rest of the infants scalp hair, has also been described in a number of other neuroectodermal defects including dermoid cysts, dermal sinus tumours, encephalocoele, sequestrated meningocoele, and leptomeningocoele. If the cyst fills when the baby cries this suggests an intracerebral communication.

The hair collar sign is an important clue alerting the clinician to the possibility that the lesions extends intracranially and that a CT or MRI scan is required, particularly if a biopsy is being considered. Ultimately the central nodule will flatten to form a circular smooth patch of rubbery hairless skin and the collar of hypertrophic hair will blend in with the adjacent scalp hair as it grows. The palisading hairs are then discernible only on close inspection.

Naevoid hypertrichosis
Hypertrichosis of a limited extent occurs as an isolated developmental defect. It can also occur over a patch of thickened parchment-like skin or complicate a benign tumour such as a neurofibroma or a smooth muscle hamartoma. Smooth muscle hamartomas show transient piloerection when stroked.

Congenital hemihypertrophy with hypertrichosis
This rare syndrome present at birth becomes more pronounced at puberty. Other possible associations include mental retardation, multiple naevi, telangiectases and internal malignancies.