INTRODUCTION

Congenital hypertrichosis is a large and complex entity in the disorders of hair growth. Nevoid hypertrichosis (NH) is an uncommon disorder consisting of a solitary, circumscribed area of terminal hair growth present at or soon after birth. The involved hair may be normal or hypo-pigmented, with or without any associated cutaneous and extra-cutaneous abnormalities. We report a 12-year-old boy with localized hypertrichosis but without any underlying pigmentary or structural abnormality.

CASE REPORT

A 12-year-old boy presented with a circumscribed linear patch of terminal hair over the middle of the forehead since birth. Dermatology consultation was sought due to cosmetic reasons. At birth, a patch of hair appeared over the forehead which gradually increased in size over the next few years. The overlying hair became pigmented, coarse and also increased in length. Parents are healthy and unrelated. Pregnancy and labor were uneventful, and the boy had normal developmental milestones. The younger brother did not have any cutaneous or systemic abnormalities.

Cutaneous examination revealed a localized 4×2 cm well-circumscribed patch of terminal hair present over the forehead, 1 cm left of the midline [Figure 1] along Blaschko’s lines. The overlying hairs were black, coarse and measured 3-4 cm in length. The skin over the NH patch was normal without any pigment alteration. The color of the hair corresponded with the scalp hair.

Routine hematological and biochemical investigations were normal. Chest X-ray was normal. Skin biopsy was performed to rule out an underlying nevus. The histological examination revealed an increased number of morphologically normal hair follicles [Figure 2].

DISCUSSION

Primary hypertrichosis has been defined based on the age of onset as congenital or acquired and on the extent of distribution as localized or generalized. Nevoid hypertrichosis is an uncommon congenital disorder consisting of terminal hair growth in a localized distribution. The skin lesions are usually present at or soon after birth and are typically solitary, though cases of multiple patches developing after puberty have been reported. A few cases of multiple NH in which patches are either round, in a check-board or linear pattern, along Blaschko’s lines have also been documented. The color of the terminal hair may be the same as the color of the scalp hair, rarely grey or white. The histopathology usually shows a normal epidermis with increased number of morphologically normal hair follicles in the dermis but is needed to rule out any underlying nevus.
It has not yet been clarified whether NH is a malformation of unknown etiology or is genetically determined. Interestingly, an autosomal dominant mode of inheritance has been proposed in giant NH. Lopez-Barrantes et al., concluded that NH may occur in a mosaic pattern and may be associated with other cutaneous diseases that may show a segmental pattern as a twin spotting phenomenon with severe mental, ocular, or myoskeletal anomalies or without any associated abnormalities. Twin spots consist of two genetically different clones of neighboring cells in a background of normal cells. If the mutations give rise to a clinically

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**Table 1: Cutaneous and systemic associations of Nevoid hypertrichosis**

| Reported publications                          | Nevoid hypertrichosis | Color of hair | Skin character | Associated abnormalities                                                                 |
|-----------------------------------------------|-----------------------|---------------|----------------|-----------------------------------------------------------------------------------------|
| Dudding, Rogers, Roddick et al., Am J Med Genet 1998 | Multiple              | –             | Depigmentation along Blaschko’s lines of unaffected skin | Retinal hyperpigmentation, epidermal nevus of arm and nevus sebaceous over scalp, spontaneous resolution |
| Chang, Hong, Kim et al., J Dermatol 1997      | Multiple, linear      | Depigmented underlying hair | Hypopigmented skin | Nil                                                                                     |
| López-Barrantes, Torrelo, Mediero et al., Eur J Dermatol 2002 | Multiple lesions   | No hypopigmentation | Linear hypopigmentation along Blaschko’s lines | No extra-cutaneous anomalies                                                             |
| Cox NH, McClure JP, Hardie RA. Clin Exp Dermatol 1989 | Multiple patches  | Normal         | Nil            | Lipodystrophy                                                                          |
| Rupert LS, Bechtel M, Pellegrini A. Pediatr Dermatol 1994 | Multiple            | Premature greying of underlying hair | –             | Nil                                                                                     |
| Schauder, Hanefeld, Noske et al., Br J Dermatol 2000 | Linear               | Depigmented underlying hair | Normal         | Cerebral and ocular malformation                                                        |
| Rogers M. Clin Exp Dermatol 1991              | Linear               | Hypopigmented hair | Hypopigmented skin | Nail, neuro, gastrointestinal, ocular, pulmonary and skeletal abnormalities               |
| Vergani, Betti, Martino et al., Pediatr Dermatol 2002 | Giant (whole back and arms) | –             | –             | Family history present                                                                  |
| Suchter MF, Khan M, Milgram S., Pediatr Dermatol 2008 | Linear               | Normal         | Normal         | No systemic involvement                                                                 |
| Sotiriadis, Patsatsi, Lazaridou et al., Pediatr Dermatol 2009 | Multiple, nevoid   | –             | –             | Nil                                                                                     |
| Ballmer-Weber, Inaebnitt, Brand et al., Dermatology 1996 | Multiple           | –             | Hypomelanosis of Ito | Facial dysmorphia, musculoskeletal abnormalities, dental, hernia, mental abnormalities |
| Taşkapan, Dogan, Cekmen et al., J Am Acad Dermatol 1998 | Single              | –             | –             | Duplication of right thumb                                                              |
visible phenotype, the twin spot is evident as paired nevoid skin abnormalities.

Congenital disorders in which localized hypertrichosis occur are Becker’s nevus, congenital melanocytic nevi and smooth muscle hamartoma.[5] Becker’s nevus presents at puberty. These conditions can be differentiated by clinical features and skin biopsy. In our case, absence of pigmented alteration of the underlying skin, absence of induration or pseudo Darier’s sign rules out the other congenital disorders mentioned above. Lumbo-sacral hypertrichosis or faun tail deformity is localized to the sacral midline and is associated with a variety of concurrent cutaneous malformations such as sacral dimple, lipoma, port-wine stain, or dermoid cyst. This developmental defect often overlies bone and spinal cord defects, most commonly spina bifida occulta and diastematomyelia.

The cutaneous lesions may persist for life, however, spontaneous resolution has also been reported.[5] Hair reduction lasers can be used to reduce the lesional hair growth. We started Long pulse Nd: YAG laser for hypertrichosis in our patient. After five sessions there was 50-60% reduction in hair growth. Patient is under follow-up and further sessions are planned.

The other associations which have been reported are Hypertrichosis cubiti, an uncommon variety of congenital circumscribed hypertrichosis in which unexplained long vellus hair are present over the extensor surface of the upper extremities along with unexplained short stature in some cases.[5] Sotiriadis et al., documented that NH is not often associated with systemic abnormalities.[8] However, they mentioned partial lipoatrophy or bony abnormalities as reported in the literature.[9,10] Further, a review of the literature by us revealed that NH may be associated with ocular, cerebral, mental, myoskeletal, neurological, gastrointestinal, pulmonary and facial dysmorphia [Table 1]. Patients with NH must be investigated to rule out any systemic involvement associated with the skin lesion(s).

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