Dermatology Practical & Conceptual

Congenital Multiple Nevoid Hypertrichosis

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Case Presentation

A 3-month-old male infant of Georgian origin was brought for evaluation of three congenital, well-demarcated patches characterized by dark, terminal hairs on the sacral-lumbar region and right lower limb (Figure 1, A and B). On physical examination, the patches had coarse hair of abnormal length. No changes in the underlying skin were found. Dermoscopy did not show brown pigmentation or pigment network. The pseudo-Darier sign was not detected on palpation. There was no relevant family history. The child had followed normal developmental milestones. A multidisciplinary pediatric consultation did not disclose abnormalities. Histology performed on the patient's back showed only terminal hair without additional relevant epidermal and dermal changes. A diagnosis of nevoid hypertrichosis was made. A magnetic resonance imaging was considered but postponed in absence of clinical issues. At eighteen-month follow-up no additional clinical problems were discovered.

Teaching Point

Nevoid hypertrichosis is a rare disorder consisting of a solitary, circumscribed area characterized by terminal hair growth on otherwise appearing skin [1]. Multiple patches may be rarely observed. It is predominant in females and arises at birth or shortly after. Hypopigmentation of the underlying skin has rarely been reported. Histopathologically, hair follicles are increased in number, but structurally normal, without characteristic or diagnostic features.

Nevoid hypertrichosis, in particular when it occurs with multiple patches, may be associated with a series of ocular, cerebral, mental, musculoskeletal, neurological, gastrointestinal, pulmonary, cardiac abnormalities and with facial dysmorphia [1,2]. Due to heterogeneity in extracutaneous manifestations, specific diagnostic procedures should follow a clinical suspicion.

The differential diagnosis includes hypertrichosis cubiti [1]. Patches of unusually long pigmented hair or focal

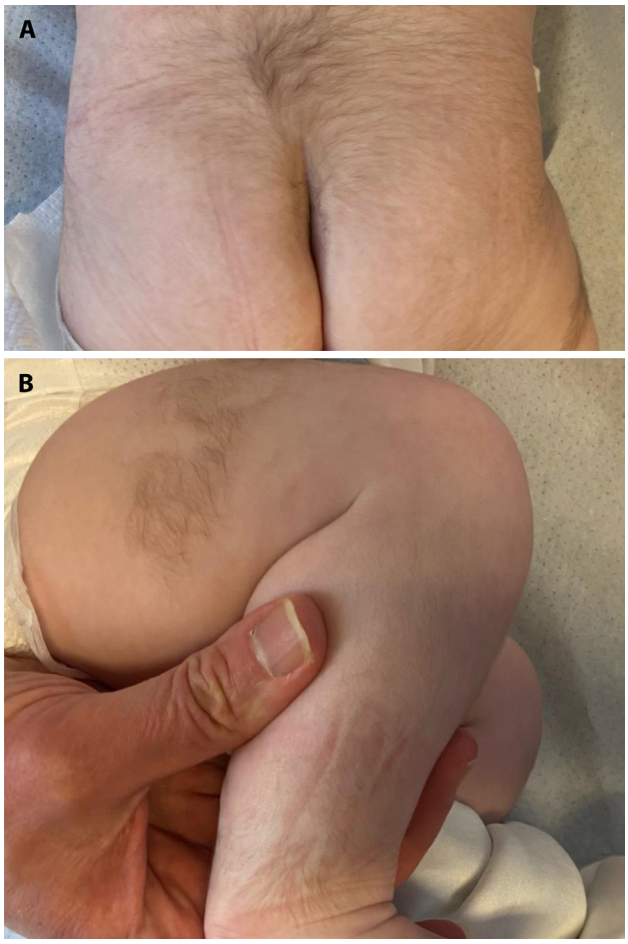

Figure 1. Coarse, black hair on the lumbosacral region (A) and right lower limb (B) (lateral aspect of the right thigh and right ankle).

hypertrichosis may be observed in individuals with Gorlin syndrome and Aicardi syndrome, suggesting a role of the PTCH protein, a negative regulator of pathways involved in hair follicle formation and growth cycle and subjects with Proteus syndrome, due to a possible role of the AKT1 kinase in the hair follicle development [1]. Smooth muscle hamartoma, Becker nevus, and melanocytic nevi may present an overlying focal hypertrichosis, but epidermal changes are helpful for the diagnosis.

References

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