ejpmr, 2022,9(3), 333-335

SJIF Impact Factor 6.222

EUROPEAN JOURNAL OF PHARMACEUTICAL AND MEDICAL RESEARCH

www.ejpmr.com

Case Study

ISSN 2394-3211

EJPMR

A CASE OF FAMILIAL CONGENITAL HYPERTRICHOSIS LANUGINOSA

[1]Dr. M. Gowri, [2]Dr. S. Amudhadevi, [3]Dr. K. Selvaraju, [4]*Dr. K. S. Kumaravel and [5]Dr. G. Balaji

[1]Junior Resident in Pediatrics Govt Mohan KumaramangalamMedical College, Salem, Tamilnadu, India.
[2,3]Assistant Professor of Pediatrics Govt Mohan KumaramangalamMedical College, Salem, Tamilnadu, India.
[4]Professor of Pediatrics Govt Mohan KumaramangalamMedical College, Salem, Tamilnadu, India.
[5]Associate Professor of Dermatology Govt Mohan KumaramangalamMedical College, Salem, Tamilnadu, India.

***Corresponding Author: Dr. K. S. Kumaravel**

Professor of Pediatrics Govt Mohan KumaramangalamMedical College, Salem, Tamilnadu, India.

Article Received on 22/01/2022 **Article Revised on 12/02/2022** **Article Accepted on 02/03/2022**

CASE REPORT

A female baby born by preterm delivery (30-32 weeks), born of a non-consanguinous marriage presented with increased presence of hair over her whole body since birth. Newborn care was given for prematurity and low birth weight at the neonatal intensive care unit for three weeks. The baby had a thick covering of smooth and fine hair at the time of birth, which began to shed after two weeks. Mother had gestational diabetes mellitus and was on insulin and oral hypoglycemic drugs. A similar clinical picture of increased hair growth was present in the mother and also in the previous sibling. In both of them the hair shedding started few weeks after birth.

Cutaneous examination of the newborn baby revealed soft, fine, black hair distributed over the face, eyebrows, chest, abdomen, back, extensor aspect of both upper and lower limbs (Fig: 1). The forehead was relatively hairy and the growth was more abundant on the eyebrows and eye lashes producing peculiar monkey facies (Fig 2). The back showed more hair towards the midline, along the spine. There was sparing of the palms and soles. Mucosa and nails were normal. Oral examination showed gingival hyperplasia. There were no signs of virilisation. The picture was consistent with congenital hypertrichosis lanuginosa. The routine haematological parameters and 17-hydroxy progesterone levels were normal. The skeletal survey on x-ray was unremarkable and no bony deformity was detected. Echocardiography was normal. Ophthalmological examination was normal. Ear examination was also normal. On follow up at 4 months of age, the hypertrichosis in the back, face and extensor aspect have resolved to a larger extent (Fig: 3).

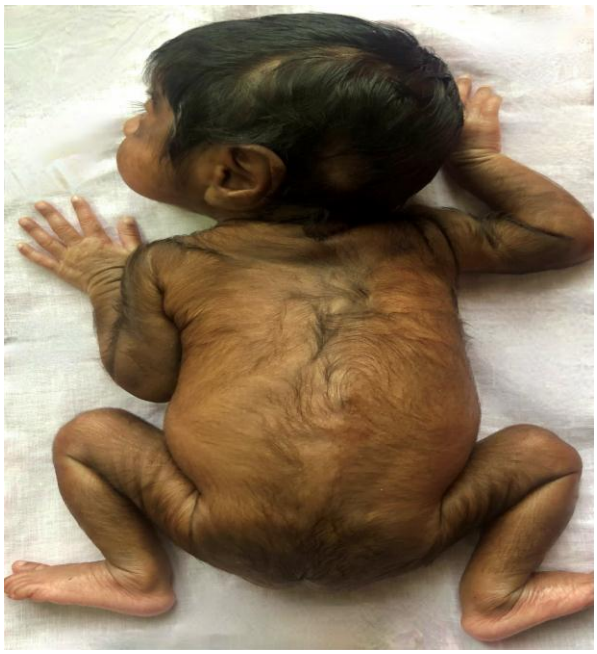

Fig. 1: Picture of neonate with profuse hair distributed over the entire back more towards the midline and extensor aspects of both upper and lower limbs.

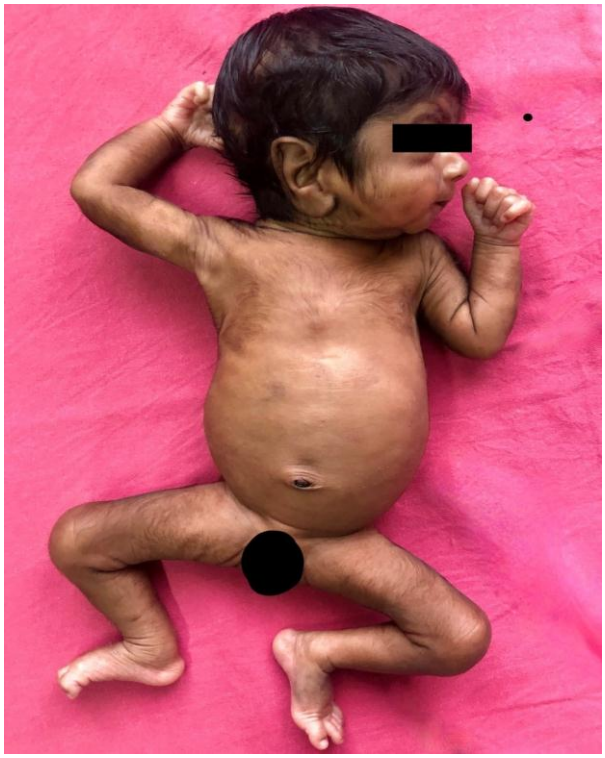

Fig. 2: Front picture of the same neonate, hair distributed over the face, eyebrows, chest and abdomen.

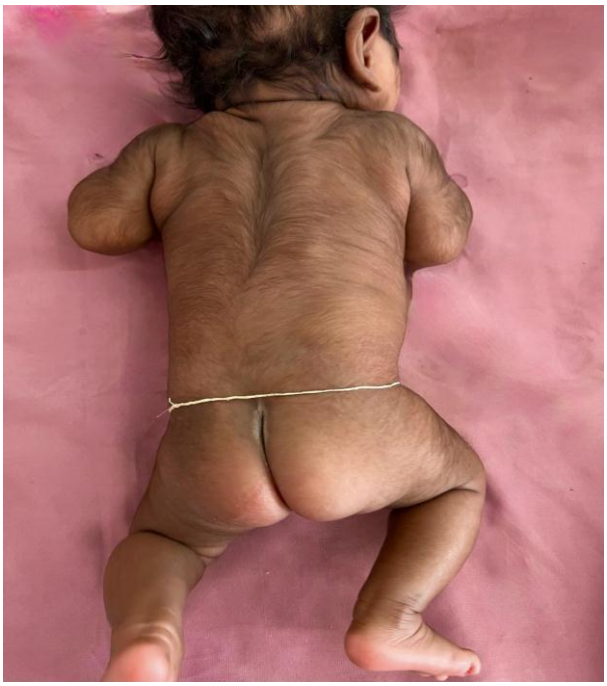

Fig. 3: Four months old baby picture of the same case during follow-up, hair distribution over the back, face, extensor aspects resolved to a greater extent.

DISCUSSION

Hypertrichosis is a very rare disorder where hair develops on any part of the body in an amount that is more than present on another individual of the same age, sex and race, excluding the androgen-dependent areas of hair growth.[1] It can be congenital or acquired.[2,3] Among the congenital types, one type is Congenital Hypertrichosis Lanuginosa (CHL), in which the patient presents with soft, non-pigmented and non-medullated lanugo hair, distributed all over the body except palms and soles, as lanugo hair has not been replaced by terminal hairs. This form has been reported in patients with abnormalities of chromosome 8q and as autosomal dominant trait inheritance. Another variant is Congenital Hypertrichosis Universalis, where the clinical presentation is the same but terminal hair is present from birth instead of lanugo hair. Familial cases with autosomal dominant inheritance and x-linked inheritance have been documented. Generalised Hypertrichosis can be associated with other anomalies such as gingival fibromatosis, amaurosis congenita cone-rod type, congenital lamellar cataracts, pigmentary retinopathy, coarse face, obesity, short stature, brachydactyly, hypo/aplastic nails and intellectual disability.[4] CHL can also be a component feature of complex syndromes - Cornelia de Lange syndrome (cutis marmorata, arched eyebrows, synophyrs), Coffin-Siris syndrome (bushy eyebrows, abnormal dentition, ear anomalies), Barber-Say syndrome (atrophic lax skin, a hairy bullous nasal tip), Wiedmann-Steiner syndrome (hypotonia, short stature, hypoplastic 12th ribs and a dysplastic hip, hypoplastic middle phalanx of 5th finger), Cantu syndrome (wide posterior fossa in the skull, distinctive osteochondro-dysplasia), Donohue syndrome (congenital insulin resistance, dwarfism, elfin-like facies), Rubinstein-Taybi syndrome (bird-like facies, hypertelorism, microcephaly), Schinzel-Giedion syndrome (craniofacialdysmorphism, cobblestone lissencephaly), Gorlin-Chaudry-Moss syndrome (midfacial flattening, underdeveloped genitals, PDA).

The diagnosis of CHL should be based on a detailed history, with or without the presence of other anomalies - particularly of the face, teeth and kidneys. CHL may cause significant emotional distress, not only in the affected patient but in the family as well. There are different approaches to the treatment including cosmetic procedures such as electro surgical epilation, pulsed light source, treatment with ruby, alexandrite, diode and especially ND:YAG laser and pharmacological treatment.[5,6,7] Topical eflornithine, a selective and irreversible inhibitor of the enzyme ornithine decarboxylase, which is found within the hair follicle, is used as pharmacological therapy.

Although this clinical occurrence is uncommon, it can encompass a wide range of disorders that might cause genetic and prognostic issues not only in the patient but in the subsequent progeny.

REFERENCES

1. Bergfeld W. Hair disorders. In: Solomon LM, Esterly NP, Dorinda Loeffel E, editors. Major problems in clinical pediatrics, Volume XIX:

Adolescent Dermatology. Philadelphia: WB Saunders editors, 1978; 361–6.
2. Beighton P. Congenital hypertrichosis lanuginosa. Arch Dermatol, 1970; 101(6): 669–72.
3. Solomon LM, Esterly NP. Structure of fetal and neonatal skin. In: Solomon LM, Esterly NP, editors. Major problems in clinical pediatrics, Volume IX: Neonatal Dermatology. Philadelphia: WB Saunders editors, 1978; 1–19.
4. Goldstein GD, Dunn M. Hirsutism: "assessing hair-raising topics". Chest, 1985; 88: 915–6.
5. Stenn KS, Paus R. Controls of hair follicle cycling. Physiol Rev., 2001; 81: 449–94.
6. Trüeb RM. Causes and management of hypertrichosis. Am J ClinDermatol, 002; 3: 617–27.
7. Wendelin DS, Pope DN, Mallory SB. Hypertrichosis. J Am AcadDermatol, 2003; 48: 161–7.