HYPERTRICHOSIS UNIVERSALIS CONGENITA
(Case report and Review)

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Summary

A rare disease, hypertrichosis universalis congenita was seen in a 7 month old male child. Father also had a similar disorder. There was long black hair all over the body of the child except on the palms, soles and genitalia. Patient has been advised electrolysis when he is about 8 years of age. A brief review of literature is included.

Hypertrichosis universalis congenita, also known as a congenital hypertrichosis languinosa is a rare but interesting condition which was probably first described by Bartholin in 1654. During the last century it attracted a lot of scientific curiosity as many of the affected individuals were displayed at exhibitions and in country fairs; the most famous of these being 'Jo-jo, the dog-faced boy', also billed as 'The Human Skye Terrier'. We present an infant with this disorder whose father was similarly afflicted.

Case Report

A seven month old male baby was admitted to the Paediatric service of Postgraduate Institute of Medical Education and Research (PGI), Chandigarh, on 18th June 1979 with the complaints of excessive hair all over the body from birth. His birth and development had been normal. Family history revealed that he was the only child. His father also had excessive hair growth all over the body since birth. The father developed massive hyperplasia of the gums in 1971 (at 20 yrs. of age) for which he was operated at P.G.I.

Physical examination revealed excessive hair all over the body except the palms, soles and genitalia. The hair was long, black and coarse (Fig. 1). He had thick eyebrows and the hairline could not be delineated over the forehead. The nose was flat and depressd and lips were thick. His face resembled a dog's facies (Fig. 2). Systemic examination including examination of gums and genitalia were normal. Developmental milestones were also normal. Skull X-ray including the jaws showed normal tooth buds. Patient was discharged with the advice to attend Dermatology clinic for electrolysis at the age of 8 years. Genetic counselling was given.

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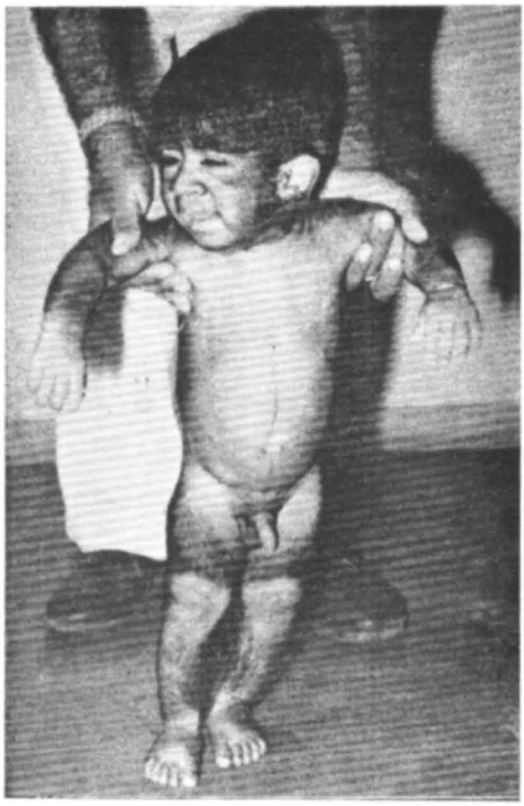

Fig. 1
Showng generalised hypertrichosis.

Discussion and Review of Literature :

Hypertrichosis has been defined as growth of hair on any part of the body in excess of the amount usually present in persons of the same age, sex, and race. Hypertrichosis universalis congenita first described in 1654[1] is usually inherited as an autosomal dominant trait, where excess hair all over the body is present from birth. However, Gates in 1946[2] gave examples of several families to show that the condition had arisen independently as mutation.

An incidence of one in thousand million was cited by Von Luschan in 1907[3], while Danforth in 1925[4] doubted if more than 30 unrelated families had been recorded. After an extensive review of literature Felgenjauer[5] in 1969, was able to find only 32 cases, adding a kindred with several affected members. Pathogenesis is unknown. It is postulated that the gene which regulates the development of hair and teeth is at fault. It is also possible that a biochemical defect exists in one or more of the hormones controlling non-sexual hair growth.

Two forms of the condition are described ; the dog-faced form and the monkey-faced form. The facies, distribution pattern of hair, dominant mode of inheritance and systemic involvement suggest that the child reported here has the dog-faced form of the disease.

The second type, called the monkey-faced form[6], is the rarer of the two and the pattern of its inheritance is uncertain. The life expectancy is poor and death may occur early in infancy without any obvious cause. If

the child survives infancy, he acquires a distinctly similar facies with a broad flat nose, thick drooping lips and prognathism. Hence the name.

A disorder which needs to be differentiated from this disease is hypertrichosis with hereditary gingival hyperplasia[7,8]. This is also a rare disorder where hypertrichosis is usually noted at puberty although 6 cases have been reported where it was present at birth or shortly thereafter. Here, the excess hair are fine in childhood but become coarse and black during adulthood. The sites of excessive hair are eyebrows, face, limbs, and middle of back. It is stated that congenital hypertrichosis languinosa is not associated with gum hyperplasia, though adontia and hypodontia have been recorded. The father of our patient developed hyperplasia in 1971 at 20 years of age. Therefore, it seems that these disorders may overlap in their manifestations and may be part of a common genetic defect.

Management of this rare entity is difficult. Removal of unsightly hair by electrolysis appears to be the only hope at present. This is laborious and difficult. Other method such as shaving and bleaching can offer only transient relief cosmetically.

The appearance creates serious psychological problems. Emotional support to the child and parents, help from social agencies, school and job placement are of particular importance. The dental anomalies can be corrected surgicaly with good results. Last but not the least, the role of genetic counselling in this autosomal dominant disorder has to be emphasized.

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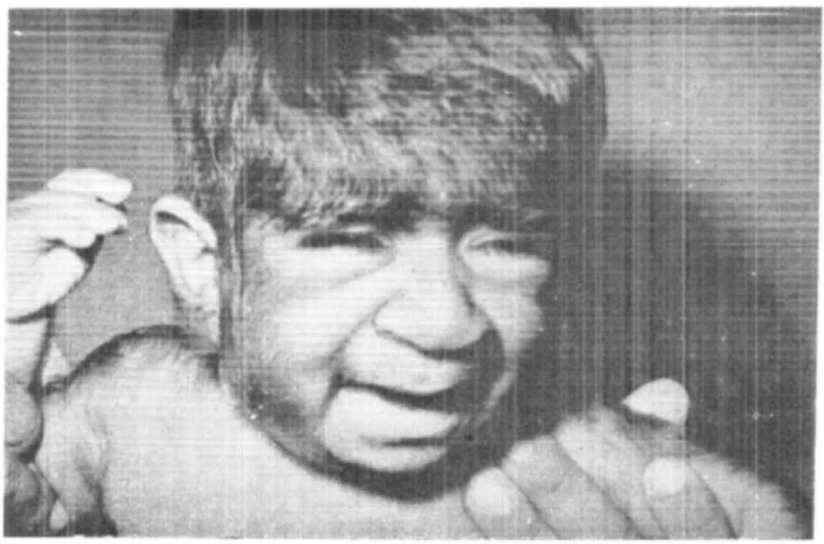

Fig 2 Close up of face showing coarse dog like facies and marked hypertrichosis with long coarse scalp and facial hair.

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