Journal of the Turkish Academy of Dermatology dISSN 1307-394X

Persistent Hypertrichosis Universalis: A Distinct Entity of Congenital Hypertrichosis

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Sir,

Hypertrichosis implies excessive growth of hair on an individual, compared with persons of same age, sex and race which are either vellus, or lanugo as against hirsutism which indicates abundance of hair of the nature of terminal hairs and which is androgen dependent [**1**]. Congenital hypertrichosis universalis [CHU] is a rare disturbance characterized by persistence of lanugo like hairs over the whole body except palms, soles and mucous membranes, since birth. These hairs may remain constant, increase with age or even decrease as the child grows older [**2**].

We report a case of a girl aged 8yrs suggestive of CHU. Soft downy hairs progressively grew since birth and covered her upper extremities, lower extremities as also her back. Those over the back had converged into whorled pattern at the time of presentation. Palms and soles were spared. Her face and external ears were normal on inspection except for long eyelashes and thick eyebrows and she did not have any other physical abnormality. Inspection of the oral cavity and radiological investigation of the denture was normal. Her eyes were declared normal by the ophthalmologist and a detailed systemic examination did not disclose any pathology. There was no offending drug history. Parents of the child were not affected but she was accompanied by her 3yr old sister who showed slightly increased hair growth over her upper extremities which had also been growing since birth [**Figure 3**]. This younger sibling too was free from any additional congenital anomaly. The mother of the children could not recall being prescribed any drug which could explain the hypertrichosis in her offspring and she was not used to consuming alcohol. Both children had normal mental development.

Baumeister [**3**] et al has classified hypertrichosis as localized or generalized (universal) and congenital or acquired. Congenital hypertrichosis universalis and congenital hypertrichosis lanuginosa have been used synonymously by others [**4, 5**]. CHU has been differentiated from congenital hypertrichosis lanuginosa by the pattern of hypertrichosis and by associated anomalies. Congenital hypertrichosis lanuginosa is an extremely rare condition where the child usually presents with soft downy hair with occasional cases of teeth deformity.

CHU has been subdivided into Transient congenital hypertrichosis universalis [TCHU] and Persistent hypertrichosis universalis [PHU] [**3**]. In another form of CHU known as the *Ambras*' syndrome the subject is covered with thick hair from birth with prominence over entire face, ears and shoulders linked with facial dysmorphism [**2**].

TCHU usually presents since birth but disappears in infancy. It is not accompanied by any other congenital malformation except for occasional report of tooth deformity [**6**] and one report with associated glaucoma [**7**]. PCHU is characterized by increased hair growth as the child grows older [**8**] and there may be frontal, preauricular and temporal abundance of hairs. Accumulation of hairs in swirls is often seen on the back. Other birth defects are not present.

After careful evaluation we could classify our cases as belonging to persistent hypertrichosis universalis, though forehead increase of hair was not marked in our cases.

Published: *J Turk Acad Dermatol* 2010; **4 (1)**: 04101L; This article is available from: http://www.jotad.org/2010/1/jtad04101L.pdf
Key Words: congenital hypertrichosis, whorled pattern

Congenital hypertrichosis universalis is an important entity as it is cosmetically distressing and can lead to severe depression. It has to be clinically differentiated from other similar disorders which are not so benign in nature and from childhood hirsutism which is often caused by endocrine disease or by drugs. Further, an understanding of the different subtypes of CHU is useful to diagnose, plan therapy and predict the prognosis in an individual afflicted with this disorder. Reassurance, regular monitoring, treatment with the minimally pain producing Q-switched Nd: YAG laser [**9**] should form the basis of management of this complex condition.

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