PUB: 03/2008

Camisa disease: A rare variant of Vohwinkel's syndrome

Sir,

Vohwinkel's syndrome is a rare, dominantly inherited keratoderma of palms and soles with a characteristic honeycomb appearance, linear and/or starfish keratoses on the extensor surfaces of the elbows, knees, knuckles and hands with flexion contractures and constricting bands (pseudoainhum) of digits resulting in autoamputation.[1,2] We report a case of Camisa disease, a rare variant of Vohwinkel's syndrome.

A 3-year-old female child born of 2nd degree consanguineous marriage presented with hyperkeratosis of the palms and soles, linear hyperkeratotic plaques over dorsa of hands with loss of left great toe, of 1-year duration [Figure 1]. Constricting fibrous bands were seen over the right great toe and right 5th toe [Figure 2]. Generalized ichthyosis was present, being more prominent over extremities [Figures 1-2]. Ridging and onychodystrophy was seen in toe nails though hair growth was normal.

Figure 1: Linear keratosis over dorsum of hands associated with ichthyosis

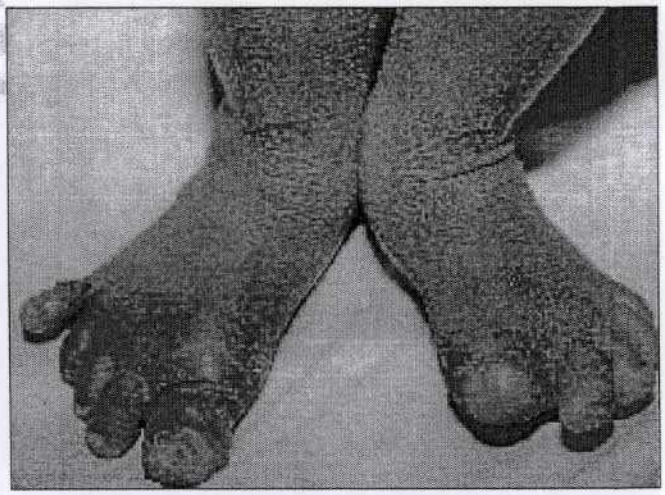

Figure 2: Constricting fibrous bands over right great toe and fifth toe with loss of left great toe

Her audiogram, eyes and dental examination were normal. There was no delay in developmental milestones. Hematological and biochemical investigations were within normal limits and peripheral smear showed microcytic hypochromic anemia with mild eosinophilia. Blood serological investigations (HIV and VDRL) were normal. Abdominal scan showed hepatomegaly. Skin biopsy revealed hyperkeratosis, focal parakeratosis, acanthosis, elongation of rete ridges and sparse dermal lymphocytic infiltrate with normal appendages.

Camisa disease is a rare variant of Vohwinkel's syndrome associated with generalized ichthyosis and without deafness.[1,2] On the basis of recent molecular studies, it is now clear that Vohwinkel's syndrome associated with ichthyosis is caused by mutations in loricrin gene.[3-5] However, a variant of Vohwinkel's syndrome which had all the classical clinical features of Vohwinkel's syndrome but lacking atypical associations like ichthyosis and sensorineural deafness with negative gene mapping for loricrin mutation has been reported recently.[6]

Along with the features of Vohwinkel's syndrome, our patient had generalized ichthyosis, which is similar to the previous case reports of Camisa variant of Vohwinkel's syndrome.[1,4] The other clinical variant of Vohwinkel's syndrome is associated with deafness but no ichthyosis.[3] However, audiogram

How to cite this article: Rajashekar TS, Singh G, Naik CL, Okade R. Camisa disease: A rare variant of Vohwinkel's syndrome. Indian J Dermatol Venereol Leprol 2008;74:81.

revealed no hearing loss in our patient. Histologically the skin lesion showed hyperkeratosis, focal parakeratosis, acanthosis, elongation of rete ridges and sparse dermal lymphocytic infiltrate with normal appendages, which were consistent to earlier case report of an ichthyotic (or Camisa) variant of Vohwinkel's syndrome.[4] Thus our case represents a rare variant of Vohwinkel's syndrome, termed as Camisa disease.

T. S. Rajashekar, Gurcharan Singh, Chandra Naik L. Rajendra Okade

Department of Dermatology,
Sri Devaraj Urs Medical College, Tamaka, Kolar, Karnataka, India

Address for correspondence: Dr. T. S. Rajashekar, Department of Dermatology, R. L. Jalappa Hospital and Research Centre, Tamaka, Kolar - 563 101, Karnataka, India.
E-mail: yeshits@rediffmail.com

REFERENCES

1. Camisa C, Rossano C. Variant of keratoderma hereditaria mutilans (Vohwinkel's syndrome). Treatment with orally administered isotretinoin. Arch Dermatol 1984;120:1323-8.
2. Solis RR, Diven DG, Trizna Z. Vohwinkel's syndrome in three generations. J Am Acad Dermatol 2001;44:376-8.
3. Korge BP, Ishida-Yamamoto A, Pünter C, Dopping-Hepenstal PJ, Iizuka H, Stephenson A, *et al.* Loricrin mutation in Vohwinkel' keratoderma is unique to the variant with ichthyosis. J Invest Dermatol 1997;109:604-10.
4. Takahashi H, Ishida-Yamamoto A, Kishi A, Ohara K, Iizuka H. Loricrin gene mutation in a Japanese patient of Vohwinkel's syndrome. J Dermatol Sci 1999;19:44-7.
5. Maestrini E, Monaco AP, McGrath JA, Ishida-Yamamoto A, Camisa C, Hovnanian A, *et al.* A molecular defect in loricrin, the major component of the cornified cell envelope, underlies Vohwinkel's symdrome. Nat Genet 1996;13:70-7.
6. Ali MM, Upadya GM. Variant of Vohwinkel's syndrome. Indian J Dermatol Venereol Leprol 2006;72:449-51.