VOHWINKEL SYNDROME (KERATOMA HEREDITARIA MUTILANS) (A case report)

T. R. KAPUR* AND O. P. SINGH†

Summary

Keratoma hereditaria mutilans (KHM) in a young Gurkha girl presented the following clinical features of this rare syndrome; palmo-plantar hyperkeratosis beginning early in life, marked keratosis of dorsa of the hands and feet and constriction of the digit of left 4th toe. Clinical variants of KHM are discussed.

In 1929, Vohwinkel described a syndrome with diffuse hyperkeratosis of the palms and soles which may extend to dorsa of the hands and feet, as well as knees and elbows. The lesions have a peculiar, linear and starfish shaped configuration[1]. In addition, constriction of digits is a prominent feature[2]. Rarity of this syndrome in world literature prompted us to report this case.

Case Report

A 16 years old Indian domiciled Gurkha girl noticed marked thickening of the skin of the hands and feet and a constricting band over the left 4th toe at the age of 10 years, which was progressing gradually. Family history of similar disorder was absent.

On physical examination, hyperkeratosis of the palms and soles was observed, which was much more marked on palms. Hyperkeratotic firm warty papules resembling starfish shaped keratosis over distal half of dorsa of both the hands involving thumb, knuckles of all proximal phalanges and first interphalanges joints, were seen. These lesions were also present on dorsa of all the toes, but were less marked. The skin over proximal half of hands, wrists adjoining 1/3 of dorsa of forearms and dorsa of both feet was rough and hyperkeratotic. There was a constricting band in the middle of 4th left toe (Fig. Page No. 221).

Comments

Vohwinkel's original patient was a 24 years old white woman who had diffuse, honeycombed, palmar and plantar hyperkeratosis and constriction of fingers which appeared in her teens ten years after the onset of the hyperkeratosis[3]. In the present case the constriction started almost simultaneously with the hyperkeratosis. Since Vohwinkel's report in 1929, several articles have appeared in medical literature regarding this keratotic dermatosis[4,5]. These reports emphasize the following clinical features.

* Classified specialist (Derm and Vener), Military Hospital, Pathankot.

† Lecturer, Dermatology and Venereology, All India Institute of Medical Sciences, New Delhi-16.

Received for publication on 8—11—1976

1. diffuse hyperkeratosis of palms and soles.
2. constricted digits
3. typical starfish keratosis of dorsa of the hands and feet and linear keratotic lesions on the elbows and knees.
4. usual affliction of white European females.
5. invariable appearance of the disease in early life.
6. familial incidence (too few patients with KHM have been described to define the nature of the possible hereditary pattern). Other clinical features reported in association with this syndrome are
7. high frequency hearing loss.
8. mutism.
9. diffuse alopecia of the scalp of the pseudopelade type and
10. a transient episode of spontaneous plantar bullae[1].

A variant of keratosis herediataria multilans has been described in Negroes[6]. The cutaneous keratotic changes seen on the elbows and knees in the classic form were absent in these patients and there was no family history of similar disease. In view of this, the present case may be considered a forme-fruste of vohwinkel syndrome. Presence of hyperkeratotic lesions seen on the palms and soles, extending to dorsa of the hands, wrists and the forcarms with marked hyperkeratosis on dorsa of the interphalangeal joints and a constriction band in the left 4th toe in a young female with onset in childhood, suggested a clinical diagnosis of keratoma heriditaria mutilans.

REFERENCES

1. Gibbs RC and Frank SB: Keratoma Hereditaria Mutilans (Vohwinkel) Arch Derm 94 : 619, 1966.
2. Rook Wilkinson DS : Textbook of Dermatology, Blackwell Scientific Publications. London, 1972.
3. Vohwinkel KH : Keratoma hereditarium mutilans, Arch Derm Syph, 158 : 354, 1929.
4. Wigley JEM : A case of hyperkeratosis palmaris et plantaris associated with ainlum like constriction of fingers : Brit J Derm : 41 : 188, 1929.
5. Wirz F : Keratoma Hereditarium Mutilans, Arch Derm Syph, 159 : 311, 1930.
6. Spencer G : Ainhum associated with hyperkeratosis palmaris et plantaris, Arch Derm Syph, 45 : 574, 1942.

The Sixth Annual Conference of
Indian Association of Dermatologists, Venereologists and Leprologists
will be held at Calcutta
on February 4 – 5, 1978.