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SUMMER VARIANT OF AUTOSOMAL DOMINANT ICHTHYOSIS VULGARIS

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Eleven cases of mild autosomal dominant ichthyosis vulgaris (ADIV) were seen who presented with asymptomatic, brownish-black hyperkeratotic lesions over the flexure surfaces of wrists, dorsum of hands, knees and ankles for 1-2 months during peak summer months. The lesions disappeared with the onset of monsoon. All these cases had typical lesions of ADIV during winter months. This paradoxical phenomenon of ichthyosis alternating with hyperkeratotic pigmented lesions in the same individual during different seasons remains unexplained. Treatment with 20% urea cream and oral vitamin A had no effect on the course of the disease.

Key words : Dominant ichthyosis vulgaris, Summer variant.

Autosomal dominant ichthyosis vulgaris (ADIV) is a common keratinizing disorder. Classically, it presents with small, fine brown scales over the extensor surfaces of extremities and trunk, and usually spares the flexural folds. It usually exacerbates in winter and clears in summer.[1,2] In a milder form, dryness and roughness are the only manifestations.[3] In the past three summers, we have seen cases with hyperkeratotic, hyperpigmented, localized lesions over the wrists, knees and ankles, appearing in the peak summer and spontaneously disappearing with the onset of monsoon. All these cases exhibited typical lesions of autosomal dominant ichthyosis vulgaris in winters.

Case Reports

Eleven patients (7 males, 4 females), aged 7-30 years in 9 families attended with asymptomatic, brownish-black, thickened lesions on the flexure surfaces of wrists, dorsum of hands, knees and ankles for 1-2 months (Figs. 1 and 2) during peak summer months. Lesions gradually increased in size and thickness. After immersion in water, the skin appeared sodden, slate-gray in colour and wrinkled. In all cases with the onset of monsoon, the lesions would clear spontaneously within 4-6 weeks. In winters, these patients presented with mild scaling on trunk. In none of the cases, history of atopy, local or systemic drug intake, bullous eruptions or chronic irritation could be elicited. In 7 out of 9 families, dominant ichthyosis vulgaris was seen in other members also, and in one family 3 members had such lesions during summer. It was interesting that this phenomenon was not seen in the parents though 45% had ADIV. Uniform mild thickening was observed on the palms and soles throughout the year. Skin on the extensor surfaces of the extremities was rough and dry. Hair, teeth and nails were normal and there was no associated systemic illness. Histopathology revealed typical features of dominant ichthyosis,[4] moderate hyperkeratosis, thinning or absence of granular cell layer and slight acanthosis, while dermis was essentially normal. Considering these cases to be a variant of ADIV they were treated with 20% urea cream and oral vitamin A (50,000 IU twice a day) without any improvement. The remission was spontaneous with the onset of monsoon.

Comments

Keratosis pilaris, hyperkeratosis of the palms and soles and localized shiny hyperkeratosis on the elbows, knees and ankles are

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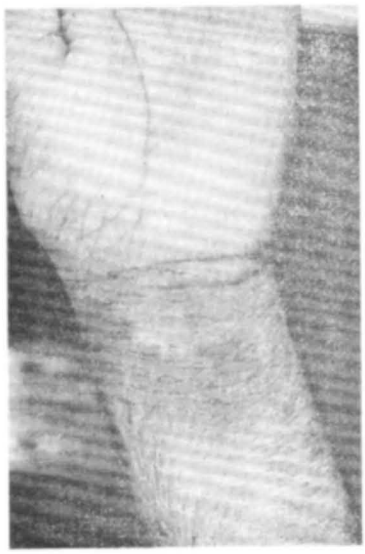

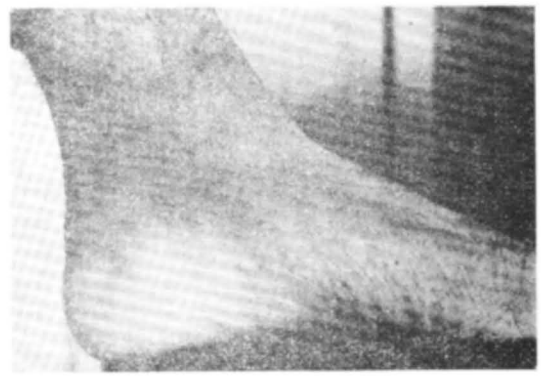

Figs. 1 and 2. Ill-defined hyperpigmented and hyper-keratotic lesions over the wrist and ankle.

some inconstant features associated with autosomal dominant ichthyosis vulgaris.[2,5] The appearance of hyperpigmented hyperkeratotic localized lesions during the summer months and remission with the onset of monsoon in ADIV cases is perhaps not reported so far. We have no explanation for this paradoxical phenomenon of ichthyosis vulgaris alternating with hyperkeratotic pigmented lesions in the same individuals during different seasons.

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