Nekam's disease (keratosis lichenoides chronica)

R W Ragunathan[1]

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Nekam's disease is a very rare, chronic inflammatory dermatosis that combines the features of a seborrhoeic dermatitis like eruption of the scalp and face with progressive lichenoid papulonodular dermatosis affecting the trunk, buttocks and limbs. It is persistent and typically does not respond to treatment. The face and scalp lesions are erythematous, greasy and scaly and bear no resemblance to those found on the trunk and extremities which are erythematous or violaceous lichenoid scaly papules present in a confluent, reticulate or linear distribution[1].

Papulonodular and inlfiltrated plaques are sometimes present. Scarring is not a feature. Genital involvement has been documented[1]. In extensive Nekam's disease, the lesions tend to be symmetrical. Associated features include oral ulceration, nail dystropy and palmoplantar keratoderma. (Nekam's original case had these associated features)[2]. Nail can become thickened, longitudinally ridged and are prone to paronychia. Oral lesions seen in 50% of patients include apthous ulcers, large chronic ulcers or erythrokeratotic papules[2]. Histologically changes are often consistent with a chronic dermatitis but lichenoid features can be seen[2]. The course of the dermatosis is chronic and progressive and very resistant to therapeutic approaches but has shown a favourable response to photo chemotherapy and etretinate[2]. Commonest age group affected is 20-40 years. Children are affected occasionally. This disease is extremely difficult to treat[3].

Nekam's disease (Keratosis lichenoides chronica) in a 60 years old male is reported. It's clinical and histological features are discussed. This condition has not been documented previously in Sri Lanka.

Case report

A 60 year old male presented in January 1999 with an itchy skin rash of four years duration. On examination he had erythematous, greasy and scaly rash mainly affecting his face scalp and neck resembling seborrhoeic dermatitis. His skin rash (Fig.1) gradually worsened over the next few months with very poor response to conventional treatment.

Subsequently he developed an itchy lichenoid papular eruption mainly affecting the trunk and extremities (Fig. 2). He also had hyperkeratosis, of palms and soles. Some of his nails were thickened and also had longitudinal ridging. Routine investigations of haematological, hepatic and renal functions were within normal limits. Because of poor response to treatment and different morphology, skin biopsies were performed from both sites. Histologically the scalp lesions showed spongiosis with exocytosis, patchy parakeratosis and acanthosis suggestive of chronic dermatitis. Lichenoid lesions demonstrated hyperkeratosis, occasional parakeratosis, acanthosis, lymphocytic infiltration in upper dermis and basal cell degeneration. A few cytoid bodies were also noted. These features were suggestive of lichen planus.

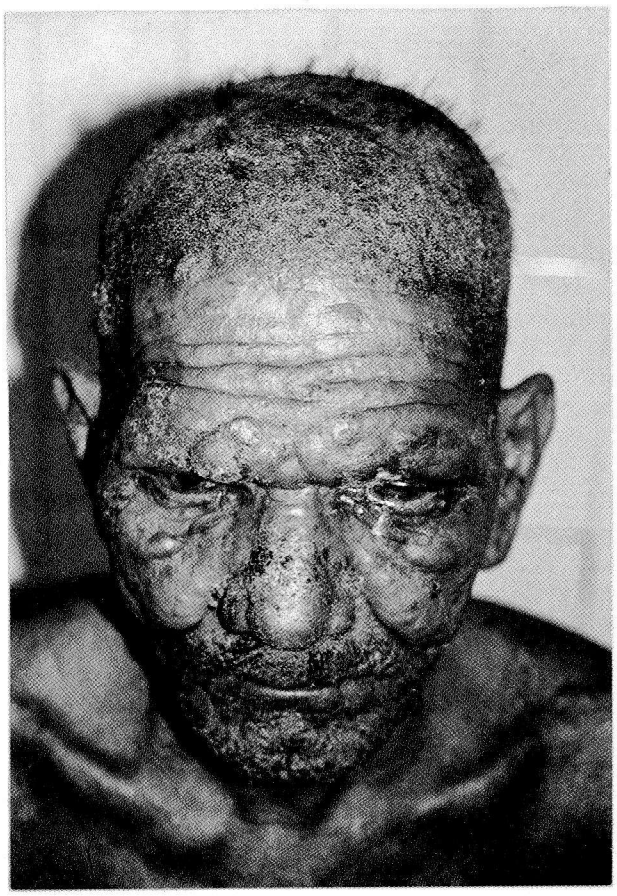

Figure 1. Lesions on face and scalp

[1] *Consultant Dermatologist, Teaching Hospital, Karapitiya, Galle.*

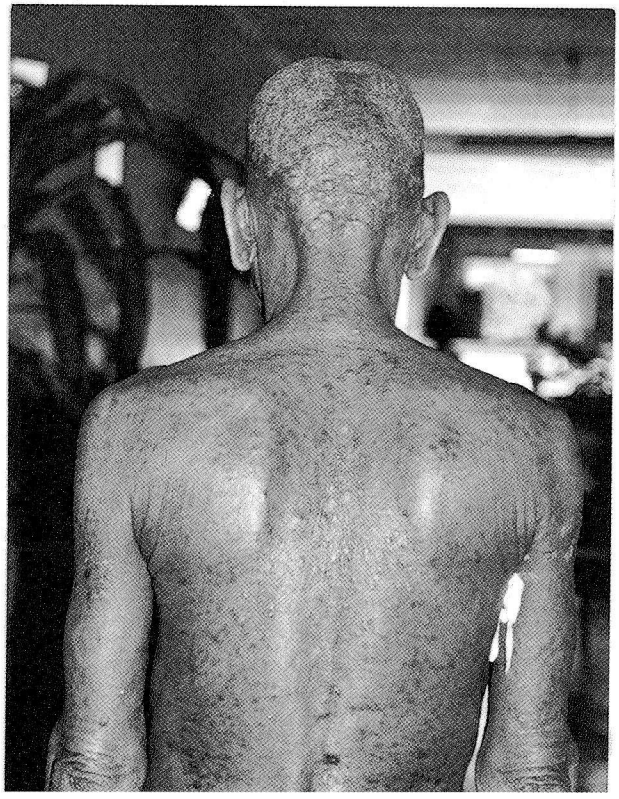

Figure 2. Lichenoid eruption

Discussion

Keratosis lichenoides chronica was first described in 1895, by Kaposi, as a case of lichen verrucosus et reticularis. Since then several authors (Wise & Rein, 1936; Nekam, 1938; De Asprer et al, 1974, Degos et al. 1974; Margolis et al 1972) have suggested alternative nomenclature. Kersey and Ive believe that the condition is an unusual variant of lichen planus[4]. Many authors have used the term keratosis lichenoides chronica to describe this condition. More recently the name Nekam's disease was established. Despite several changes in nomenclature both its aetiology and association with other diseases remain obscure[4]. Braun-Falco et al consider that keratosis lichenoides chronica does not show any pathological relationship to any other dermatosis and suggested that it is a distinct entity[5]. A limited variant of Nekam's disease presenting with erythematous hyperkeratotic papules and plaques on the face has been described in two young siblings[6]. Curiously, the facial eruption had improved in the summer months in some cases of typical Nekam's disease[7].

The case described by the author has the clinical and histological features described by most authors. Clinical features, poor response to treatment and histological findings of this patient is compatible with Nekam's disease.

References

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