Ind J Dermatol Venereol Leprol 1995; 61 : 299-300

PAPULOERYTHRODERMA OF OFUJI

D Parsad, R K Gautam, H K Kar, R K Jain, M Bhardwaj*

A 62-year-old female had intensely pruritic eruption consisting of widespread coalescing sheets of uniform erythematous papules with characterstic sparing of body folds (deck-chair sign). Papuloerhthroderma of Ofuji is a rare disease of unknown aetiology.

Key Words : Papuloerythroderma of Ofuji, Deck-chair sign

Introduction

Papuloerythroderma of Ofuji is a rare disease reported recently as an eruption with severe pruritus and characteristic sparing of body folds. Ofuji in 1981[1] reported it first as an eruption which typically consists of coalescing sheets of uniform, flat-topped, erythematous papules with striking sparing of flexural regions, skin creases and compressed abdominal folds (deck-chair sign).[2] The histological features reported include dense perivascular infiltrate comprising of lymphocytes, histiocytes, plasma cells and eosinophils.[1-3] Exudates and extravasation of RBC's in the keratin layer have also been recorded.[2] There are only seven documented cases so far and only once it has been reported in a female.[4] The of onset has ranged from 57-75 years.[3]

Case Report

A-62 year-old female patient presented with debilitating pruritic eruption involving the chest, abdomen, trunk and extremities for the last 4 months. There was nopersonal history of atopy, drug reaction, diabetes mellitus and hypertension. On exanubatuib, she had diffuse erythema, oedema and scaling of the skin involving the above mentioned sites. The body folds i.e., submammary region, compressed abdominal folds, cubital fossae were conspicuous by their sparing (Fig.1). There were numerous

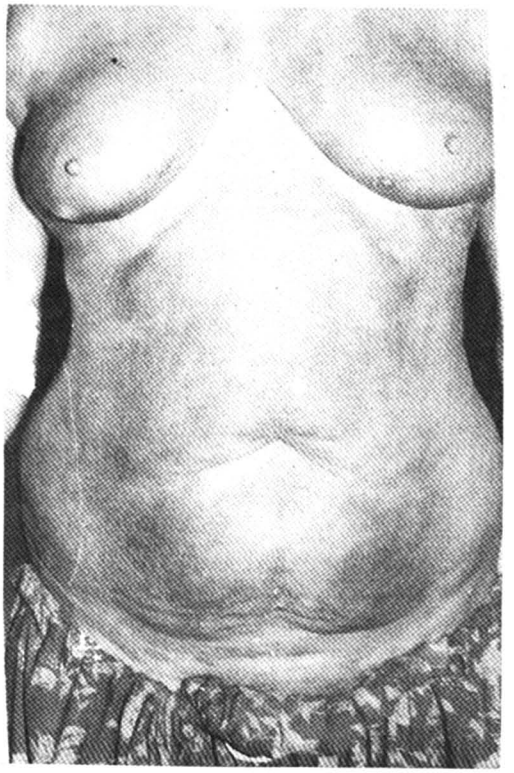

Fig. 1. Sparing of compressed abdominal folds (deck-chair sign).

sheets of flat topped coalescing papules located on the chest, abdomen and back. There were a few excoriation marks on the above mentioned sites. The nails, mucous membranes and hair were normal. The

From the Department of Dermatology and STD, and pathology*, Dr R M L Hospital, New Delhi-100 001 India.

Address correspondence to : Dr H K Kar

systemic examination was noncontributory. The histopathological examination of papule from the abdomen revealed mild hyperkeratosis, patchy parakeratosis, mild acanthosis with irregular elongated rete ridges. Several neutrophils along with fibrinous exudate were seen in the keratin layer. The superficial dermis showed oedema and sparse perivascular infiltrate.

Discussion

The diagnosis of papuloerythroderma of Ofuji was clinched on the basis of characterstic clinical features. These included very itchy coalescing sheets of papules with sparing of body folds. It was further supported by the histological findings. To the best of our knowledge this is the first case being reported from India.

The nosologic status of papuloerythroderma of Ofuji is still undecided, however, from a purely symptomatologic point of view, it constitutes a distinctive entity.[5] The nature of the disease is also undecided and it may not be more specific than any other form of erythroderma and thus may be a manifestation of a variety of underlying diseases, eczema or psoriasis in elderly patients[3] or skin lymphoma.[5] Further, it could be that papuloerythroderma is a peripheral T cell skin lymphoma different from mycosis fungoides and Sezary syndrome.[5] Anbother very important aspect of this entity is the dramatic response to steroids which was also observed in our case. In a further follow up of 6 months the disease has not relapsed in our case.

References

1. Ofuji S, Furukawa F, Miyachi Y. Papuloerythroderma. Dermatologica 1984; 169: 125-30.
2. Farthing CF, Staughton RCD, Rowland Payne CME. Papuloerythroderma-a further case with the 'deck chair sign'. Dermatologica 1986; 172: 65-6.
3. Haris DWS, Spencer MJ, Tidman MJ. Papuloerythroderma-clinical and ultrastructural features. Clin Exp Dermatol 1990; 15: 105-6.
4. Langtry JAA, Rowland Payno CME, Harper JI. Papuloerythroderma in a woman. J Royal Soc Med 1985; 81: 170-7.
5. Grob JJ, Collet-Villete AM, Horchowski MD, et al. Ofuji papuloerythroderma J Am Acad Dermatol 1989; 20: 927-31.