PAPULOERYTHRODERMA OF OFUJI

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A 60-year old male with papulo erythroderma of Ofuji ia presented here for its rarity. The characteristic 'deck chair' sign was present. Response to PUVA therapy was noted.

Key Words: Papuloerythroderma, Ofuji's disease, 'Deck chair' sign

Introduction

Papuloerythroderma is a rare intensely pruritic eruption of unknown etiology, consisting of widespread sheets of uniform erythematous papules with characteristic feature 'deck chair' sign.[1] We report here a case of papuloerythroderma of Ofuji with its characteristic clinical and histological feature, for its rarity.

Case Report

A 60-year old male presented with history of flat-topped erythematous papules and nodules all over the body progressing to uniform sheets of thickened skin. There was history of intense itching without any diurnal variation. There was no history suggestive of hypertension, diabetes mellitus or any other chronic ailments. He gave history of intensely pruritic papules and nodules going in for infarcted skin ulceration mainly over the shoulder and buttock.

Cutaneous examination revealed thickened and infiltrated look of face, extremities and abdomen. There were infarcts over shoulder, back of trunk, few of them healed with scars. There was characteristic sparing of the abdominal folds, the so called 'deck chair' sign. Skin biopsy revealed endothelial swelling of capillaries, leakage of RBC, and at places perivascular infiltrate.

Complete haemogram revealed peripheral eosinophilia (-21%). Abdominal USG, X-ray chest, LFT and renal function tests were within normal limit. A clinical and histological diagnosis of papuloerythroderma was made and PUVA and antihistamine therapy was strated.

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Discussion

Ofuji's erythroderma is an intensely pruritic eruption of unknown etiology. The characteristic feature is 'deck chair' sign i.e., sparing of compressed abdominal folds.[1-4] There may also be small infarcts of nail folds, and larger infarcts over the buttocks as in the present case. Infaracts may be due to dermal capillary endothelial swelling, but not due to thrombi.

There may be circulating eosinophilia, as noted in the present case. Other associated features may be raised I gE, lymphopenia and development of T-cell lymphoma.[2]

Patient was treated successfuly with PUVA therapy and oral antihistamines as it was the only alternative left with us. Earlier patient was prescribed oral steroid without any result. There was marked reduction in pruritus, thickening and ulceration of skin by the end of eighth exposure to PUVA.

References

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