PRIMARY PAPULOERYTHRODERMA OF OFUGI IN A 60-YEAR-OLD MALE PATIENT

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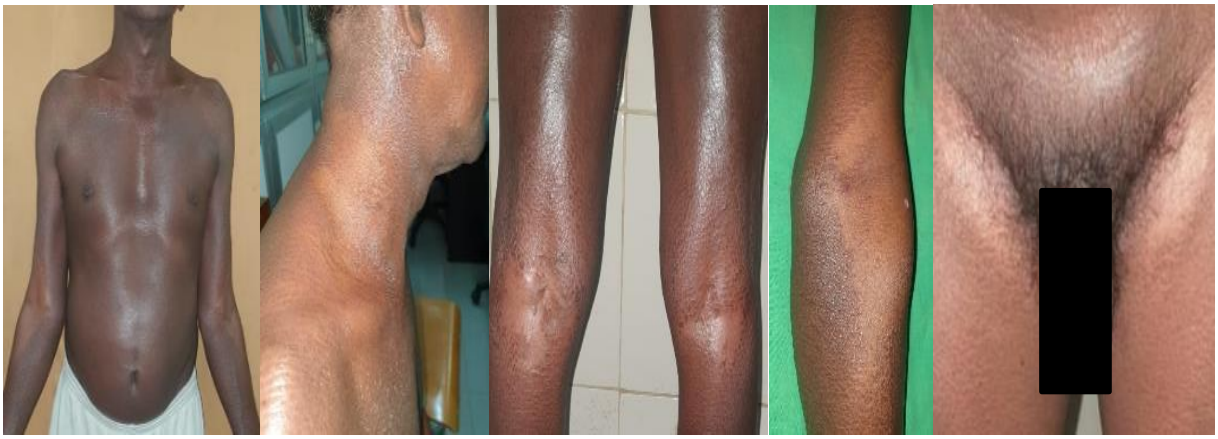
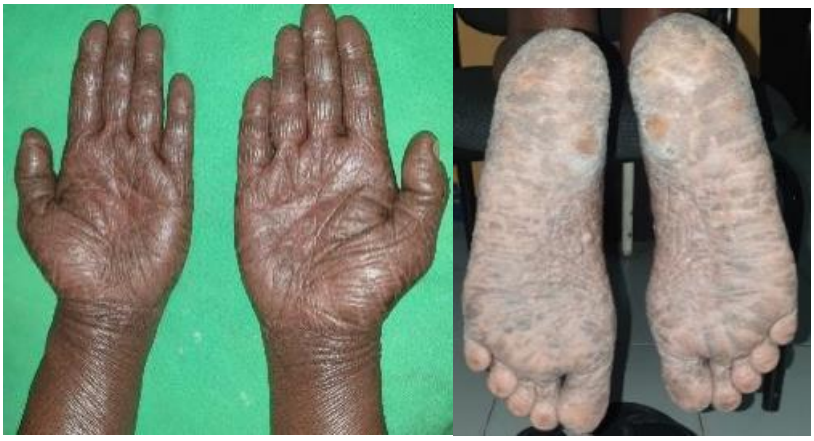
60-year-old male presented with progressive, intensely itchy generalized papular skin eruption for 2 years. He had no history of atopy, other skin disease or constitutional symptoms.

Figure 1: Erythroderma with exfoliative dermatitis involving more than 90% body area with flexural sparing (deck-chair sign)



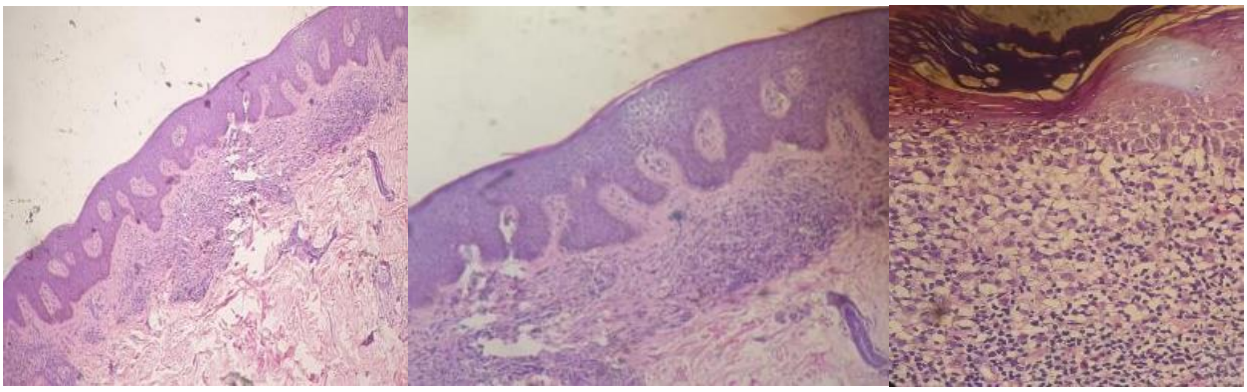
Examination revealed erythematous to hyperpigmented papules coalescing giving rise to cobblestone like appearance over torso and limbs but sparing the flexures.

Figure II : Palmar planter hyperkeratosis



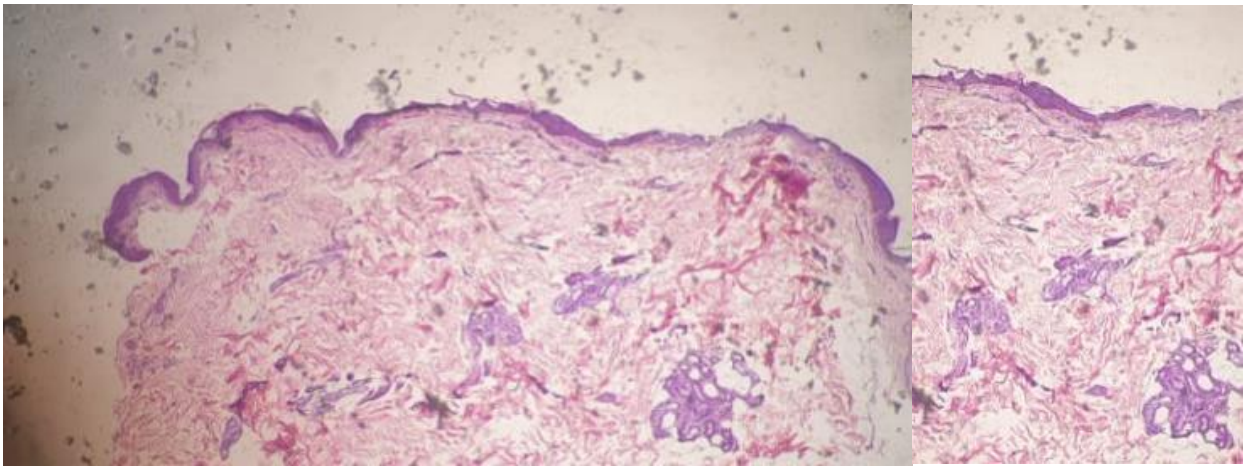
Moreover, painful fissured palmoplantar keratoderma was noted. He was not pale; no generalized lymphadenopathy or hepatosplenomegaly was present.

Figure III: Skin biopsy findings of involved skin in involved skin



Skin biopsy from the involved skin revealed focal parakeratosis, acanthosis, focal spongiosis and mixed inflammatory cell infiltrate in the dermis. No features suggestive of mycosis fungoides were noted.

Figure IV : Skin biopsy findings of normal skin from the flexural areas



Concurrent skin biopsy from the normal skin was unremarkable. Papuloerythroderma of ofugi(PEO) is a clinical entity, with unclear etiopathogenesis(1). Four etiological subtypes have been described. Diagnosis of PEO is based on ten diagnostic criteria and the idiopathic form of the syndrome requires the presence of all the 5 major criteria(2). Topical and/or systemic corticosteroids are the most frequently used but azathioprine, cyclosporine, retinoids and phototherapy have also been tried with variable success(3,4). Response to therapy and prognosis is usually determined by the aetiological associations(5).

Peripheral eosinophilia was noted with mild lymphopenia and elevated IgE levels but rest of the haematological and biochemical evaluation were unremarkable(6). Though the patient was extensively investigated for underling infections and malignancies, imaging and serological testing was within normal limits(7).

Based on the diagnostic criteria idiopathic PEO was diagnosed and patient was started on combination immunosuppressive therapy with steroids and methotrexate with good response. Deck chair sign /sparing of the flexures is considered characteristic for this entity, but it is not pathognomonic as other dermatoses like atopy and cutaneous lymphomas may have similar clinical appearance-so we had to exclude them actively in this patient. Though we have concluded this case as primary PEO, longitudinal evaluation of the patient was planned to rule out any associations and evolution.

REFERENCES

1. Torchia D, Miteva M, Hu S, Cohen C, Romanelli P. Papuloerythroderma 2009: Two New Cases and Systematic Review of the Worldwide Literature 25 Years after Its Identification by Ofuji et al. Dermatology [Internet]. 2010;220(4):311–20. Available from: https://doi.org/10.1159/000301915

2. Patel RH, Fagan K, Chitnavis P V, Grider D. Papuloerythroderma of Ofuji in a Young Man. Cureus. 2023;15(3):10–3.

3. Apalla Z, Koukouthaki A, Sidiropoulos T, Psarakis E, Nikolaidou C, Bobos M, et al. Dermoscopy of papuloerythroderma of Ofuji. Vol. 35, Photodermatology, photoimmunology & photomedicine. England; 2019. p. 127–8.

4. Li S, Yu X, Wang T. Papuloerythroderma of Ofuji. JAMA Dermatology [Internet]. 2020;156(12):1365. Available from: https://doi.org/10.1001/jamadermatol.2020.2973

5. Teraki Y, Aso Y, Sato Y. High Incidence of Internal Malignancy in Papuloerythroderma of Ofuji: A Case Series from Japan. Dermatology [Internet]. 2012;224(1):5–9. Available from: https://doi.org/10.1159/000335563

6. Terlikowska-Brzósko A, Paluchowska E, Owczarek W, Majewski S. Papuloerythroderma of Ofuji in a 41-year-old woman. Postep Dermatologii i Alergol. 2013;30(5):324–8.

7. Desai K, Miteva M, Romanelli P. Papuloerythroderma of Ofuji. Clin Dermatol [Internet]. 2021;39(2):248–55.
Available from: https://www.sciencedirect.com/science/article/pii/S0738081X20301966