INFLAMMATORY SKIN DISEASES (OTHER THAN ATOPIC DERMATITIS & PSORIASIS)

AN INTRIGUING CASE OF EXTENSIVE ARBORIZING NEKAM'S DISEASE

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BACKGROUND: Keratosis lichenoides chronic (KLC) or Nekam's disease is a rare, chronic and progressive mucocutaneous keratinizing dermatosis. Typical lesions are red to violaceous papulonodular, hyperkeratotic, and covered with gray scales which frequently coalesce to form linear and reticulate arrays of warty lichenoid lesions. In extensive Nékam's disease, the lesions tend to be bilaterally symmetrical. It is resistant to most therapeutic modalities and no effective treatment has been established.

OBSERVATION: A 34-year-old otherwise healthy woman presented with intensely itchy eruptions, insidious in onset and gradually progressing since 20 years. The lesions appeared on trunk and extremities. There is history of multiple topical and oral medications with no satisfying outcomes. On cutaneous examination there were widespread hyperkeratotic erythematous to violaceous plaques with thick adherent scales present over the posterior aspect of trunk , buttocks, thighs, anterior aspect of lower legs and dorsum of foot distributed in a bilaterally symmetrical and arborizing pattern. Histopathological examination showed basket weave hyperkeratosis, focal hypergranulosis, irregular acanthosis, eosinophilic civatte bodies around dermo-epidermal junction and lymphohistiocytic infiltrate in upper dermis, periadnexal and perivascular areas and extensive fibrosis in mid and lower dermis. On the basis of clinical and histopathological findings diagnosis of extensive Nekam's disease was made.

KEY MESSAGE: KLC is characterized by its chronic presentation and refractoriness to treatment. This case is remarkable and extremely rare due to the extensive bilaterally symmetrical and striking arborizing pattern which is not yet described in literature also uncommon feature of extensive fibrosis in mid and lower dermis.