

Dermatomyositis-Like Lesion: An Atypical Cutaneous Presentation in Adult-Onset Still Disease

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Case Presentation

A 56-year-old female with no prior dermatological history presented with persistent pruritic erythematous scaly plaques accompanied by fever, myalgia, and generalized lymphadenopathy for over 20 days. The rash had started on the neck and chest and had rapidly spread to the face, back, and thighs (Figure 1A-D). Histopathology found dyskeratotic keratinocytes of the upper third of epidermis. Muscle, bone marrow lymph node biopsies, and PET-CT found no substantial abnormality. The patient was initially diagnosed as having dermatomyositis. However, based on Yamaguchi criteria, especially the highly specific and sensitive skin histopathology, clinical presentation and subsequent examinations, the final diagnosis was adjusted to adult-onset Still's disease (AOSD). During our 10-month follow-up after treatment with glucocorticoids and siltuximab, laboratory results, including complete blood count, liver function, and ferritin levels, showed improvement; the patient no longer had fever or any discomfort.

Teaching Point

AOSD is a rare systemic inflammatory disease with diverse clinical presentations. Skin rash is a key feature, which is typically characterized as evanescent, non-pruritic eruption, and often followed by spiking fever[1]. Here, we report an atypical case of AOSD with persistent pruritic dark-red lesions on the face, neck, chest, and back, which are frequently associated with poor prognosis[2]. Dyskeratotic cells of the upper-to-mid-epidermis in histopathology is a crucial clue for diagnosis (Figure 1E). Dermatomyositis is the primary disease for differential diagnosis, and hematologic malignancies should be ruled out. Early recognition of these clinical and pathological signs can aid in timely clinical decision-making.

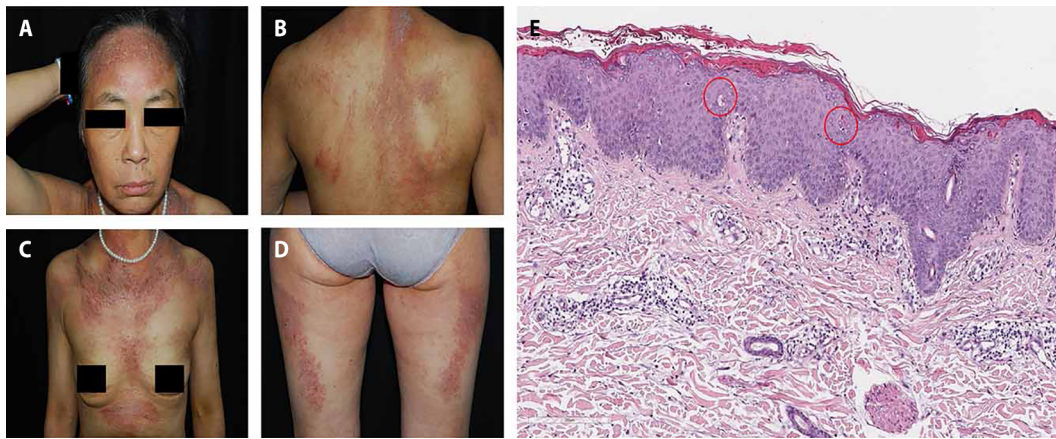


Figure 1. (A-D) The patient presented erythema scale rash on the forehead, face, chest, back, and lateral thigh. (E) Histopathology found several dyskeratotic keratinocytes of the upper third of the epidermis and a few infiltrations of lymphocytes, neutrophils, and eosinophils in the superficial dermis (H&E, 400x).

Reference

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