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Letter to the editor

A case of adult-onset Still's disease with psoriasiform eruptions

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Adult-onset Still's disease (AOSD) is a rare, acute systemic inflammatory disease, which is characterized by spiking fever, polyarthralgia and a salmon-pink evanescent rash.[1] Recently, it was revealed that not only does a typical skin rash appear with AOSD, but so do several atypical skin eruptions.[2] Some AOSD skin lesions are persistent and consist of papules and erythematous plaques with scales or crusts, and these are described as "persistent pruritic papules and plaques".[2] Here we report a female AOSD patient who presented with psoriasiform eruptions during the course of AOSD.

A 42-year-old woman presented with a spiking fever of up to 39℃, pharyngeal pain, polyarthralgia, a salmon-pink evanescent rash on the thigh, linear erythema on the back (Fig. 1a) and psoriasiform eruptions (erythema with thick scales) on the upper chest (Fig. 1b, c). Until this episode, she had no history of skin diseases including psoriasis vulgaris. The present case was included in the case series of our previous research article on phosphorylated signal transducer and activator of transcription 3 (STAT3) expression in AOSD lesions.[3] Laboratory examinations revealed the following: increased leukocytosis (12,500/mm^3, normal range: 3800-8500), mildly elevated C-reactive protein (3.0mg/dL, normal range: <0), liver abnormality (AST 157IU/L, normal range: 13-33; ALT 53IU/L, normal range: 6–27; lactate dehydrogenase 1,595IU/L, normal range: 119–129), and hyperferritinemia (36,560ng/mL, normal range: 3-129). A skin biopsy from the erythema with thick scales showed acanthosis, elongation of rate ridges and microabscesses in the stratum corneum (Fig. 1d, e). The histological findings were consistent with psoriasis vulgaris. Systemic computed tomographic scans showed hepatosplenomegaly (Fig. 1f).

The present case satisfied the diagnostic criteria of AOSD proposed by Yamaguchi

et al.[1] We administered oral prednisolone (PSL, 1.0mg/kg/day), and the eruptions improved and the fever abated. However, while tapering the PSL (0.2mg/kg/day) after a period of improvement, the skin symptoms and polyarthralgia recurred. Finally, the symptoms were successfully treated with an interleukin-6 antagonist.

We considered the psoriasiform eruptions in the present patient to be a previously unreported, distinct type of AOSD skin lesions, although the linear erythema on the back is thought to be a persistent pruritic eruption in AOSD. Dyskeratotic cells in the upper epidermis were reported as a characteristic histological feature of persistent pruritic eruptions.[2] In contrast, dyskeratotic cells in the upper epidermis were not seen, but plaque-type psoriasis-like inflammatory reactions were observed histopathologically in the psoriasiform eruptions of the present case. The present psoriasiform lesions occurred and regressed synchronously with the AOSD and the persistent pruritic eruptions of AOSD, and psoriasiform eruptions never appeared independently from symptoms of AOSD. Thus, we consider the psoriasiform eruptions to be AOSD skin lesions. In the literature, we were unable to find any reports of psoriasis vulgaris complicated with AOSD, although there was one case report of generalized pustular psoriasis complicated with AOSD.[4] In conclusion, we reported a case of AOSD with psoriasiform eruptions, and we should keep in mind that there could be a wide variety of skin lesions in AOSD patients, other than only persistent pruritic eruptions.

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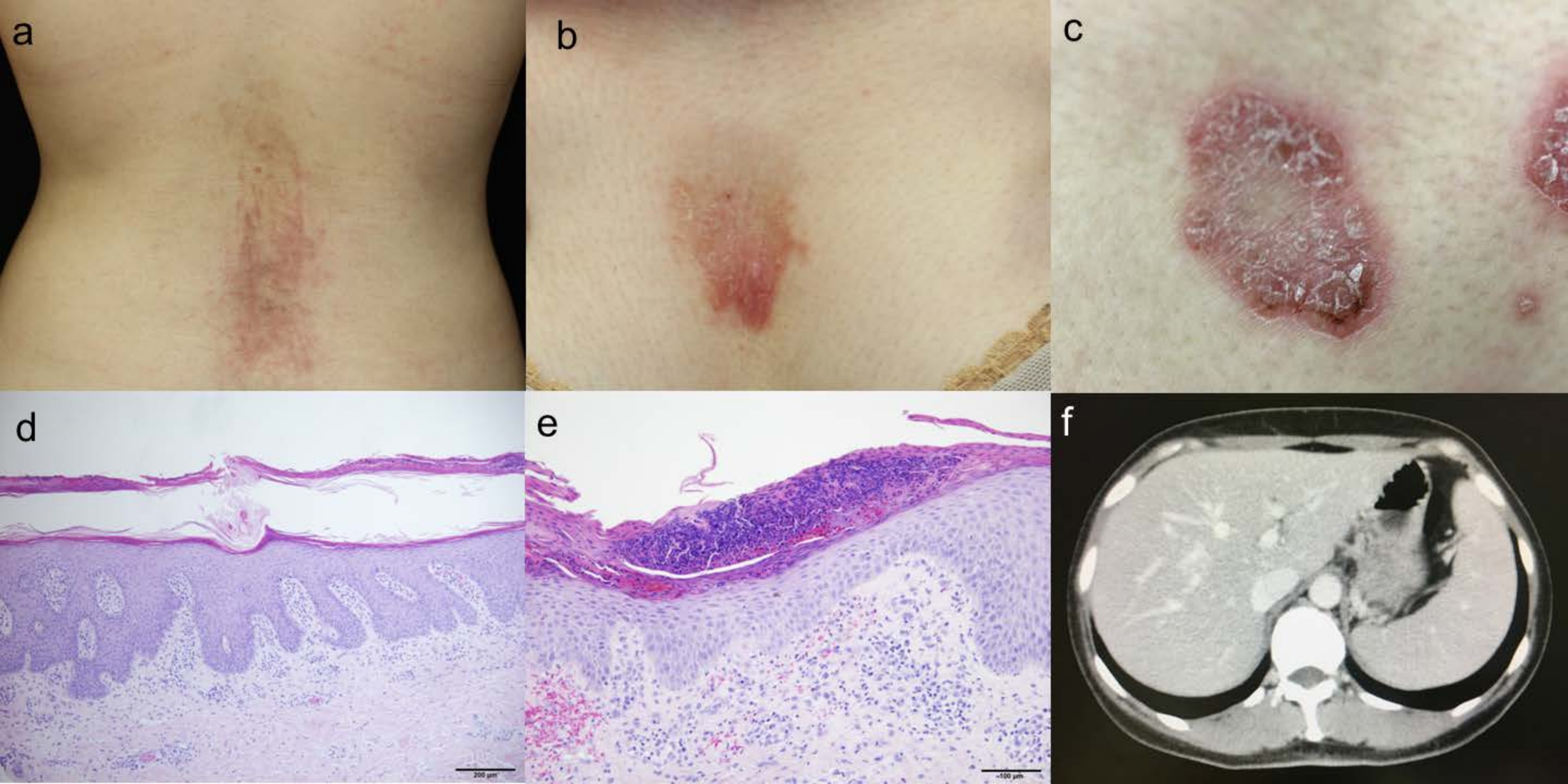
Figure legends

Figure 1. The clinical features of the patient

(a) Sharply defined linear erythema on the back after scratching. (b, c) Scaly hyperkeratotic erythema (b) and scaly erythematous plaques and pustules (c) on the chest. (d) A skin biopsy specimen shows parakeratosis and hyperkeratosis in the epidermis. Scale bar: 200μm. (e) Neutrophilic pustules are observed within the stratum corneum. Scale bar: 100μm. (f) Hepatomegaly and splenomegaly are detected by computed tomography.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