

Persistent Pruritic Eruptions in Adult-onset Still's Disease: Additional Histopathological Findings

Adult-onset Still's disease (AOSD) is a systemic inflammatory disorder of unknown etiology that occasionally involves atypical eruptions such as persistent pruritic eruptions (PPEs) (1). We report two cases of AOSD that simultaneously showed typical eruptions and PPEs and we describe the possible histopathological findings in PPEs.

Case 1 was a 53-year-old Japanese woman who was referred to our Department for diagnosis of eruptions associated with a spiking fever ($>39^{\circ}\text{C}$) that had been present for 9 days. Salmon-pink erythema without itching on the abdomen (Figure 1, a) and pruritic persistent erythematous papules and plaques on the waist and buttocks (Figure 1, b) were observed. The patient complained of pharyngodynia, swollen neck lymph nodes, and arthralgia in the knees, elbows, and shoulders. Histopathology was consistent with salmon-pink erythema of AOSD (Figure 1, c). Histopathology of the erythematous plaques showed parakeratosis, acanthosis, scattered necrotic keratinocytes, prominent exocytosis of eosinophils, and perivascular lymphocytic infiltrate with many neutrophils and eosinophils in the superficial dermis (Figure 1, d). Abnormal laboratory results included leukocytosis (white blood cells (WBC), 24000 / μL ; neutrophils, 22280 / μL), liver dysfunction (aspartate aminotransferase, 114 U/L; alanine aminotransferase, 86 U/L), and elevated serum C-reactive protein (CRP) (19.79 mg/dL) and ferritin (11278 ng/ml).

Case 2 was a 63-year-old Japanese woman who was referred to our Department for diagnosis of eruptions associated with a spiking fever ($>39^{\circ}\text{C}$), pharyngodynia, and arthralgia in the knees, elbows, and hands that had been present for 3 days, 2 weeks, and 3 weeks, respectively. Salmon-pink erythema without itching on the left forearm (Figure 2, a) and pruritic persistent erythematous papules and plaques on the waist and buttocks (Figure 2, b) were observed. Histopathology was consistent with salmon-pink erythema (Figure 2, c). Histopathology of the erythematous plaques showed findings similar to those in case 1 (Figure 2, d, e). Abnormal laboratory results included

leukocytosis (WBC, 14700 / μL ; neutrophils, 13090 / μL) and elevated serum CRP (26.22 mg/dL) and ferritin (6335 ng/mL).

In both cases, serum antinuclear antibodies and rheumatoid factor were negative, and infection, col-

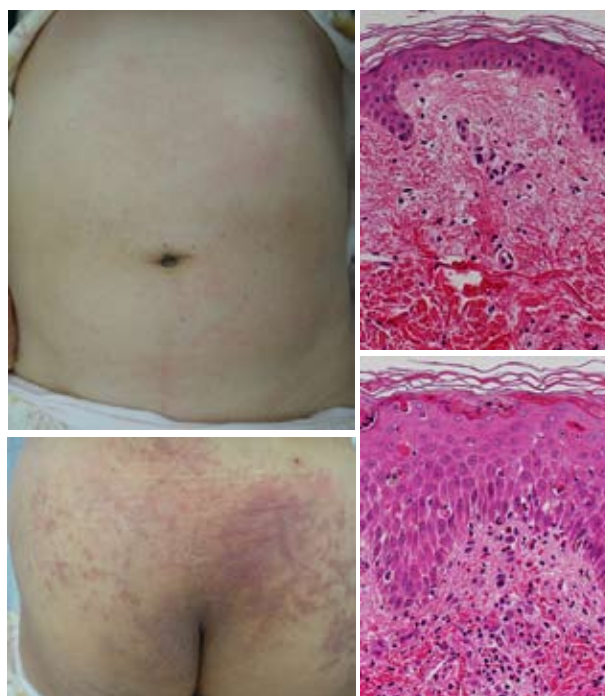


Figure 1. Clinical photographs (a, b) and histological studies (hematoxylin and eosin, original magnification $\times 200$) (c, d) in case 1. (a) Salmon-pink erythema without itching on the abdomen and (b) pruritic persistent erythematous papules and plaques on the waist and buttocks. (c) Histopathology of the erythema on the abdomen showed considerable superficial dermal edema and a mild perivascular lymphocytic infiltrate with scattered neutrophils in the superficial dermis. (d) Histopathology of the erythematous plaques showed parakeratosis, acanthosis, scattered necrotic keratinocytes, prominent exocytosis of eosinophils, and perivascular lymphocytic infiltrate with many neutrophils and eosinophils in the superficial dermis.

lagen disease, and internal malignancies were excluded. Therefore, we diagnosed both cases as AOSD (2). The skin lesions disappeared with oral steroid treatment for AOSD.

A review of the histology of AOSD-associated PPEs showed that this disease is characterized by multiple individual necrotic keratinocytes, including the normal or parakeratotic horny layer, infiltration of lymphocytes and neutrophils in the superficial and middle dermis, and an absence of eosinophils

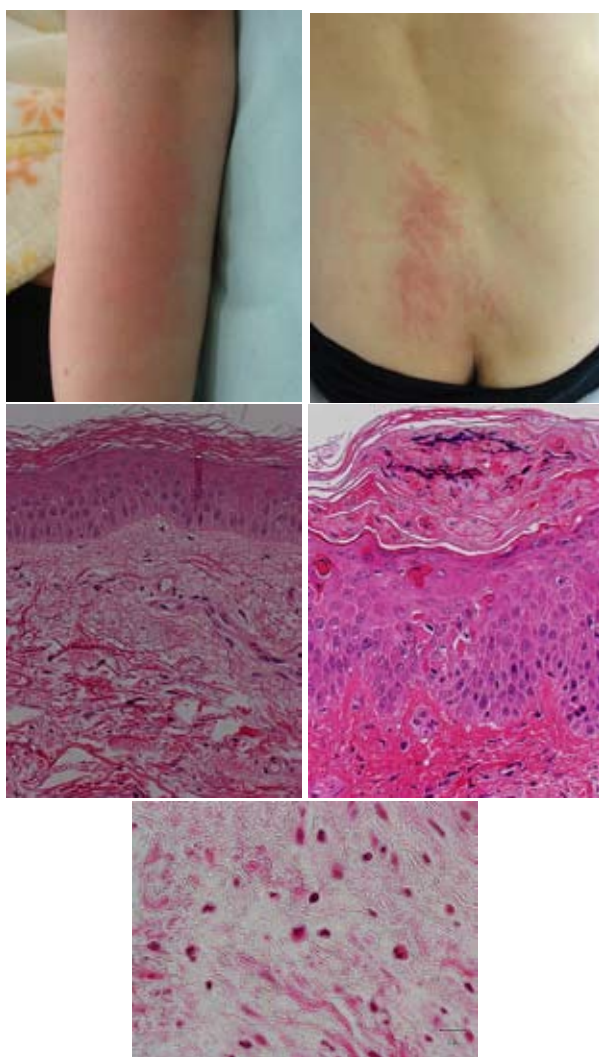


Figure 2. Clinical photographs (a, b) and histological studies (hematoxylin-eosin, original magnification $\times 200$) (c-e) in case 2. (a) Salmon-pink erythema without itching on the left forearm and (b) pruritic persistent erythematous papules and plaques on the waist and buttocks. (c) Histopathology of the erythema on the left forearm showed findings similar to those of faint erythema in case 1. (d) Histopathology of the erythematous plaques on the buttocks showed findings similar to those for the erythematous plaques in case 1. (e) A perivascular lymphocytic infiltrate with eosinophils was also seen in the superficial dermis

(1,2). To the best of our knowledge, there have been 2 cases with AOSD-associated PPEs with presence of eosinophils in the English language literature (3,4). In the reports, a dermal infiltrate including lymphocytes, neutrophils, and eosinophils was seen. Eosinophilic spongiosis was also confirmed (3). In our cases, these findings were also observed.

The pathophysiology of itch involves a complex network of cutaneous and neuronal cells; keratinocytes, mast cells, and cells of the inflammatory infiltrate, including lymphocytes and eosinophils, which may interact with neuronal cells (5). Therefore, an eosinophil infiltrate in PPEs is a possible finding that may depend on the severity of the eruption. Thus, the two cases described here illustrate possible additional histopathological findings in PPEs.

References

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