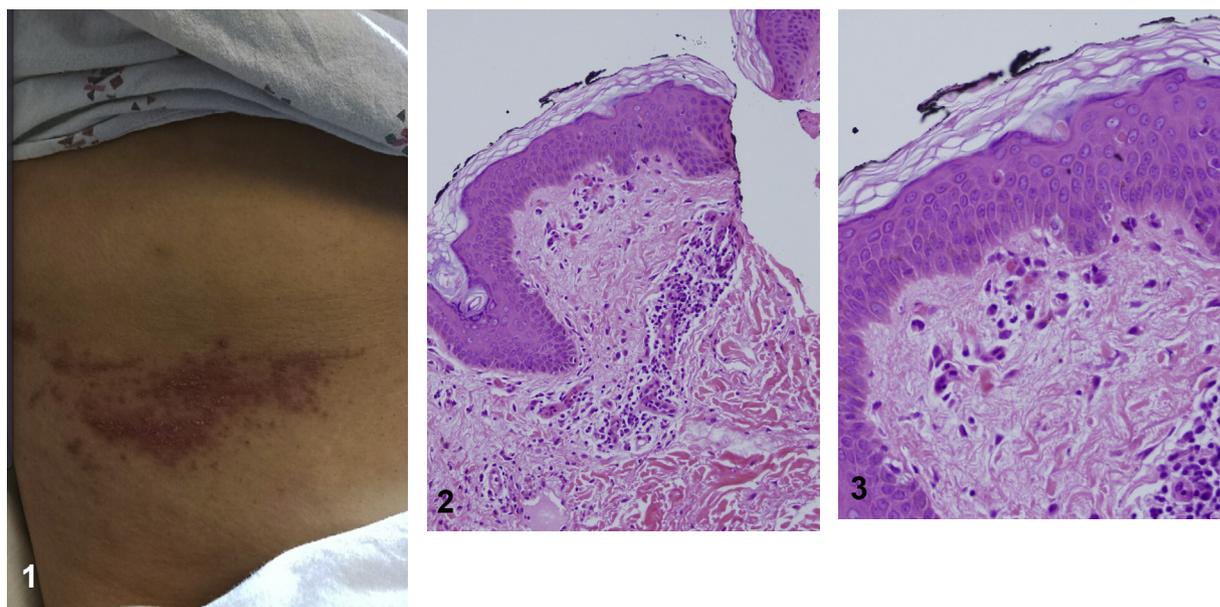


Joint pain with violaceous papules and plaques



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Key words: adult-onset Still disease; arthralgias; arthritis; persistent plaques; Still disease; violaceous plaques.



CASE PRESENTATION

A 39-year-old woman presented with a 2-week history of new-onset bilateral leg pain, arthralgias, and difficulty arising from bed. She reported a 2-day history of a pruritic rash on her right knee, right flank, posterior neck, and anterior scalp, along with a 3-day history of throat pain and fevers (daily maximum temperature, 39.4°C). Laboratory values included white blood cell count, 11.1 (normal range [NR], 4-11) with 90% polymorphonuclear neutrophils (NR, 40-80%); platelets, 469 (NR, 150-450); ferritin 3,662 (NR, 12-300); erythrocyte sedimentation rate, 115 (NR, 0-29); C-reactive protein, 225 (≤ 10); aspartate aminotransferase, 91 (NR, 10-40); alanine aminotransferase, 67 (NR, 7-56); negative antinuclear antibodies (ANA) and rheumatoid factor. Physical examination found excoriated, linear, and ill-defined violaceous papules and plaques (Fig 1). Biopsy of the flank revealed interface dermatitis (Figs 2 and 3).

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Question 1. What is the most likely diagnosis?

- A. Dermatomyositis
- B. Mixed connective tissue disease
- C. Interstitial granulomatous dermatitis
- D. Adult-onset Still disease (AOSD)
- E. Systemic lupus erythematosus (SLE)

Answers:

A. Dermatomyositis – Incorrect. Although dermatomyositis may present with proximal muscle weakness, the cutaneous findings often include a heliotrope rash and/or Gottron papules. Pathology findings show mucin deposition in the dermis, sparse lymphocytic infiltrate, epidermal atrophy, and vacuolar changes of the basal keratinocytes.¹

B. Mixed connective tissue disease – Incorrect. In the absence of positive antinuclear antibodies and elevated anti-ribonucleoprotein antibodies, this disease is unlikely. Mixed connective tissue disease typically presents with Raynaud phenomenon, arthralgias, esophageal dysfunction, and muscle weakness.

C. Interstitial granulomatous dermatitis – Incorrect. This rare disease manifests with arthritis and a wide spectrum of cutaneous manifestations, most commonly erythematous papules and plaques.² The histopathologic findings include dense, often palisading, histiocytic infiltrate in the dermis with some scattered neutrophils and eosinophils.

D. AOSD – Correct. AOSD can have this atypical presentation with persistent violaceous papules and plaques, as opposed to the classic evanescent salmon-colored rash.³ In addition to having clinical and histopathologic findings that were consistent with AOSD, this patient met the Yamaguchi criteria for AOSD.⁴ She met 2 major criteria (presence of arthralgias for 2 weeks and elevated white blood cell count with greater than 80% polymorphonuclear neutrophils) and 3 minor criteria (presence of sore throat, abnormal liver function, and negative ANA and rheumatoid factor.)

E. SLE – Incorrect. The typical manifestations in this disease include a malar rash, discoid rash, photosensitivity, arthritis, presence of a positive ANA, and systemic findings. Although SLE is an interface dermatitis, the necrotic keratinocytes are consistent with the atypical presentation of AOSD.

Question 2. Which of the following histopathologic findings would most likely be seen in this patient?

- A. Multiple necrotic keratinocytes in the epidermis and a mixed dermal infiltrate with numerous neutrophils
- B. Epidermal atrophy, degeneration of the basement membrane, vacuolar basal keratinocytes, dermal mucin deposits, and dermal lymphocytic infiltrate
- C. Vacuolar changes in basal layer, apoptotic keratinocytes, and epidermal atrophy
- D. Hyperkeratosis, irregular acanthosis, liquefactive degenerative changes of the basal layer, and lymphocytic infiltrate at the dermoepidermal junction
- E. Spongiosis, focal vacuolar degenerative changes of basal keratinocytes, superficial dermal edema, and dermal perivascular lymphocytic infiltrate

Answers:

A. Multiple necrotic keratinocytes in the epidermis and a mixed dermal infiltrate with numerous neutrophils – Correct. These findings are associated with the atypical variant of AOSD that presents as persistent violaceous papules and plaques, as seen in our patient.³ The classic evanescent rash of AOSD presents as an interstitial and perivascular mixed infiltrate with numerous neutrophils on histopathology.

B. Epidermal atrophy, degeneration of the basement membrane, vacuolar basal keratinocytes, dermal mucin deposits, and dermal lymphocytic infiltrate – Incorrect. These findings are more consistent with dermatomyositis, which is another type of interface dermatitis.¹

C. Vacuolar changes in basal layer, apoptotic keratinocytes, and epidermal atrophy – Incorrect. These findings are associated with another type of interface dermatitis, cutaneous lupus erythematosus.¹

D. Hyperkeratosis, irregular acanthosis, liquefactive degenerative changes of the basal layer, and lymphocytic infiltrate at the dermoepidermal junction – Incorrect. These findings are consistent with lichen planus, which is another type of interface dermatitis.⁵

E. Spongiosis, focal vacuolar degenerative changes of basal keratinocytes, superficial dermal

edema, and dermal perivascular lymphocytic infiltrate — Incorrect. These findings are more characteristic of erythema multiforme, a different type of interface dermatitis.⁶

Question 3. Which of the following cytokines is a strong mediator in this disease and is correlated with disease activity?

- A. Interleukin (IL)-8
- B. IL-1
- C. IL-23
- D. Interferon (IFN)- γ
- E. IL-10

Answers:

A. IL-8 — Incorrect. Patients with AOSD can have high serum levels of IL-8; however, this cytokine is not typically associated with disease activity.⁷

B. IL-1 — Correct. Patients with AOSD often have high serum levels of IL-1.⁷ IL-1 and IL-6, both important mediators in the pathogenesis of AOSD, are associated with clinical symptoms and disease activity. For this reason, biologics that target IL-1, such as anakinra, are used in the treatment of AOSD. However, IL-1 receptor antagonists are not considered a first-line of therapy for AOSD. Oral corticosteroids remain the first-line agents for AOSD. Our patient was started on high-dose oral prednisone, and methotrexate was added to her treatment regimen as the prednisone was tapered. Her symptoms, including her rash and joint pain, improved on this regimen. She continues to follow up with rheumatology.

C. IL-23 — Incorrect. This cytokine is typically involved in the pathogenesis of other inflammatory diseases, such as psoriasis.

D. IFN- γ — Incorrect. Patients with AOSD can have high serum levels of IFN- γ ; however, this cytokine is not typically associated with disease activity.⁷

E. IL-10 — Incorrect. This cytokine is not a major mediator in the pathogenesis of AOSD.⁷

Abbreviations used:

ANA: antinuclear antibodies

AOSD: adult-onset Still disease

SLE: systemic lupus erythematosus

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