Received: 24 November 2021 | Revised: 11 January 2022 | Accepted: 13 January 2022

DOI: 10.1002/cia2.12227

Journal of Cutaneous Immunology and Allergy

WILEY

CORRESPONDENCE

Dyskeratosis, a characteristic histopathological feature, seen in a patient with systemic juvenile idiopathic arthritis

Systemic juvenile idiopathic arthritis (SJIA) develops before 16 years of age and is characterized by persistent arthritis and fever lasting for at least 6 and 2 weeks, respectively. It is also accompanied by one or multiple skin rash, lymphadenopathy, hepatosplenomegaly, and serositis.[1] This disorder was described first by Dr. George F. Still in 1897, thereafter termed as Still's disease.[2] The similar condition in adults was described and now known as adult-onset Still's disease (AOSD),[3] which shares clinical manifestations and laboratory findings with SJIA.[4,5] However, the histopathology of SJIA is not well recognized compared to that of AOSD because of scarcity of patients.[6] Here, we experienced a case of SJIA, who provided us a knowledge of the histopathological features.

A 9-year-old boy presented with complaints for the period of 3 weeks of remittent fever, polyarthralgia, waxing, and waning urticarial erythematous macules. The skin lesions developed on the face, neck, and extremities following fever and frequently showed a linear configuration along with scratching marks (Figure 1A). Laboratory data were as follows: white blood cell count, 16.4×10^3/μl (normal range of $3.3\text{-}8.6 \times 10^3$/μl) with neutrophilia; platelet count, 43.6×10^4/μl ($15.8\text{-}34.8 \times 10^4$/μl); lactate dehydrogenase, 378 U/L (121-222 U/L); C-reactive protein, 8.57 mg/dl (≤0.14 mg/dl); erythrocyte sedimentation rate, 119 mm/h (ESR 2-10 mm/h); ferritin, 2411 ng/ml (39.9-465 ng/ml); negative for anti-cyclic citrullinated peptide antibody. Computed tomography scan revealed widespread lymphadenopathy with enlargement of the cervical, axillary and inguinal lymph nodes. Histopathological findings taken from his right arm showed mild acanthosis, dyskeratosis in the upper and lower epidermis, focal vacuolar interface change, superficial, and mid-dermal mixed infiltrates of neutrophils and lymphocytes (Figure 1B). With the constellation of quotidian fever along with inflammatory signs including polyarthritis and lymphadenopathy, he was diagnosed with SJIA and treated with naproxen. Although the fever, arthralgia, and skin rash continued during the initial treatment, the patient responded well to intravenous administration of methylprednisolone at a dose of 30 mg/kg/day for 3 days, which was followed by oral prednisolone at a dose of 0.9 mg/kg/day and was slowly tapered. Ten months after the onset, all his symptoms resolved.

Cutaneous histopathology in AOSD included dermal infiltrates, which varied among neutrophil- or lymphocyte-rich and mixed cells.[7] Other findings included focal vacuolar interface changes and acanthosis associated with the presence of dyskeratotic cells in the upper epidermis.[7] SJIA and AOSD show two histopathological similarities: dyskeratosis mainly is in the superficial epidermis and a sparse, superficial dermal infiltrates are composed of neutrophils without vasculitis.[6] Dyskeratosis may be a feature of increased apoptosis that has been also found in peripheral blood lymphocytes of AOSD patients and was associated with up-regulation of proinflammatory cytokines, especially IL-18.[8]

Since the diagnosis of SJIA is difficult and frequently delayed, a recognition of these characteristic clinical and histopathological features may aid to diagnose this disease. In particular, dyskeratosis in the epidermis can help making the diagnosis when only mild or localized lesions are present.

Journal of Cutaneous Immunology and Allergy

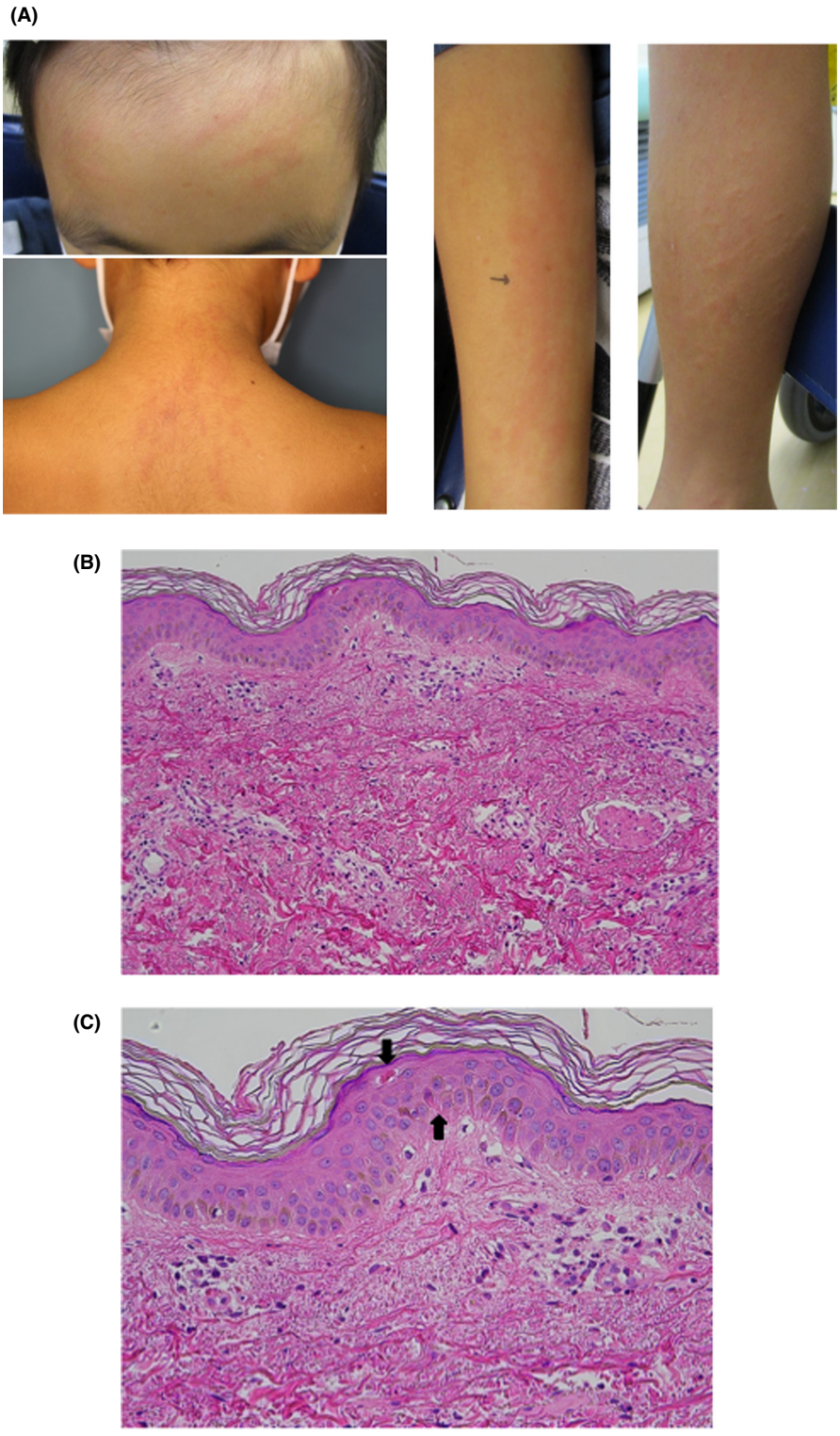

FIGURE 1 (A) Clinical findings at the first presentation. Linear urticarial erythema on the forehead, the posterior neck, the right arm, and the right leg. Linear lesions become confluent on the arm. (B, C) Histopathological findings of urticarial erythema on the right arm (Hematoxylin-eosin staining; original magnifications: ×200 and ×400 for B and C, respectively). Perivascular infiltrates of neutrophils and lymphocytes in the upper and lower dermis and focal interface change. Dyskeratotic cells are observed in the upper and lower epidermis (arrows)

DECLARATION SECTION

Informed Consent: All informed consent was obtained from the subject.
Approval of the research protocol: N/A.
Registry and the Registration No. of the study/trial: N/A.
Animal Studies: N/A.

CONFLICT OF INTEREST

The authors declare no conflict of interest. Dr. Shigetoshi Sano is a member of the Journal of Cutaneous Immunology and Allergy Editorial Board. Management of the peer review process, and all editorial decision making, for this article was undertaken by Editor-in-Chief.

KEYWORDS

dyskeratosis, systemic juvenile idiopathic arthritis, adult-onset Still's disease, polyarthralgia, urticaria

Kimiko Nakajima MD, PhD[1]
Hideki Nakajima MD, PhD[1]
Mariko Araki MD, PhD[2]

Mikiya Fujieda MD, PhD[2]
Shigetoshi Sano MD, PhD[1]

[1]Department of Dermatology, Kochi Medical School, Kochi University, Nankoku, Japan
[2]Department of Pediatrics, Kochi Medical School, Kochi University, Nankoku, Japan

Correspondence
Hideki Nakajima, Department of Dermatology, Kochi Medical School, Kochi University, Oko-cho, Nankoku, Kochi, 783-8505 Japan.
Email: nakajima@kochi-u.ac.jp

These authors contributed equally to this work.

ORCID

Kimiko Nakajima https://orcid.org/0000-0002-0341-3252
Hideki Nakajima https://orcid.org/0000-0001-6406-962X
Shigetoshi Sano https://orcid.org/0000-0002-9812-0216

REFERENCES

1. Giancane G, Consolato A, Lanni S, Davi S, Schiappapietra B, Ravelli A. Juvenile idiopathic arthritis: diagnosis and treatment. Rheumatol Ther. 2016;3(2):187–207.
2. Still GF, Garrod AE. On a form of chronic joint disease in children. Med Chir Trans. 1897;80:47.
3. Bywaters EGL. Still's disease in the adult. Ann Rheum Dis. 1971;30(2):121–33.
4. Luthi F, Zufferey P, Hofer MF, So AK. Adolescent-onset Still's disease: characteristics and outcome in comparison with adult-onset Still's disease. Clin Exp Rheumatol. 2002;20(3):427–30.
5. Pay S, Türkçapar N, Kalyoncu M, Şimşek İ, Beyan E, Ertenli İ, et al. A multicenter study of patients with adult-onset Still's disease compared with systemic juvenile idiopathic arthritis. Clin Rheumatol. 2006;25(5):639–44.
6. Fortna RR, Gudjonsson JE, Seidel G, DiCostanzo D, Jacobson M, Kopelman M, et al. Persistent pruritic papules and plaques: a characteristic histopathologic presentation seen in a subset of patients with adult-onset and juvenile Still's disease. J Cutan Pathol. 2010;37(9):932–7.
7. Larson AR, Laga AC, Granter SR. The spectrum of histopathologic findings in cutaneous lesions in patients with Still disease. Am J Clin Pathol. 2015;144(6):945–51.
8. Qiao J, Zhou S, Li S, Xue R, Pan Y, Tang S, et al. Histopathological diagnosis of persistent pruritic eruptions associated with adult-onset Still's disease. Histopathology. 2019;74(5):759–65.

How to cite this article: Nakajima K, Nakajima H, Araki M, Fujieda M, Sano S. Dyskeratosis, a characteristic histopathological feature, seen in a patient with systemic juvenile idiopathic arthritis. J Cutan Immunol Allergy. 2022;5:136–138. doi:10.1002/cia2.12227