Annals of Hematology (2019) 98:787–788
https://doi.org/10.1007/s00277-019-03613-1

LETTER TO THE EDITOR

Giant cellulitis-like Sweet syndrome as an initial clinical presentation of acute myeloblastic leukemia with t(6;9)(p23;q34): DEK-CAN and internal duplications of FMS-like tyrosine kinase 3

Shuhei Okuyama[1] · Toshiya Nito[2] · Naoki Yanagawa[3] · Katsushi Tajima[1]

Received: 4 January 2019 / Accepted: 9 January 2019 / Published online: 18 January 2019

Dear Editor,

A 56-year-old female farmer knocked her right knee against the ground and suffered from stinging pain, while she was working in a rice field. The next day, she developed a painful erythematous eruption on the right knee. She consulted a dermatologist and was diagnosed with cellulitis, and was treated with flomoxef antibiotic orally. However, on the following day, the right knee was still painfully inflamed, and she was transferred to a local hospital. On admission, she presented with an extensive cellulitis-like eruption on the right leg and mild fever. She had a history of sub-total gastrectomy, due to gastric cancer without chemotherapy 19 years ago. Her laboratory findings and clinical course are described in Fig. 1. A computed tomography or magnetic resonance image showed subcutaneous to fascia lesions without fluid formation and mild edematous swelling with multiple thrombophlebitis in the right leg. Intensive treatments, including administrations of meropenem, clindamycin, and daptomycin and two local skin debridements, seemed to transiently improve the skin lesions. Recurrent tissue cultures of the dissected skin lesions and her blood revealed no pathogens, including bacteria and fungi. However, the skin lesions were not remedied despite these intensive treatments, and therefore, the patient transferred to our hospital for further treatments and evaluations.

In our hospital, once a re-debridement was performed, the inflamed skin lesion and laboratory data gradually improved without antibiotics, and she was discharged. However, 2 weeks later, she developed new erythematous and partially purpuric lesions with pain and edema on the contralateral left leg and pancytopenia (Fig. 1). These findings strongly suggested an undetected underlying systemic disease. A bone marrow examination revealed that she had acute myeloblastic leukemia (AML) with the chromosomal translocation t(6;9)(p23;q34), DEK-CAN and internal duplications of the FMS-like tyrosine kinase 3 (FLT/ITD). A skin biopsy showed the infiltration of neutrophils in the dermis and adipose tissue, superficial dermal edema, intact epidermis, and the absence of vasculitis or massive necrotic change, compatible with a diagnosis of giant cellulitis-like Sweet syndrome (SS) [1–3]. Chemotherapy with prednisolone and indomethacin relieved the skin lesions and thrombophlebitis (Fig. 1).

SS sometimes appears as a paraneoplastic neutrophilic dermatosis in connection with malignancies, either before, during, or after the definite diagnosis [4]. Recently, six cases of giant cellulitis-like SS, characterized by relapsing widespread giant lesions with bullous appearances, have been reported [1], although this pathogenesis remains unknown. This new variant of SS is likely to be misdiagnosed as acute infectious cellulitis, when the skin lesions precede the discovery of an underlying malignancy. In this case, the skin lesions seemed to transiently respond to the intensive treatments, and the initial-onset episode of the skin lesion and the late-onset of cytopenia without pathological cells in the peripheral blood might have postponed her precise diagnosis. We could not deny the possibility that the comorbidity of thrombophlebitis exacerbated and extended her skin lesions. In conclusion, this is the first reported case of giant cellulitis-like SS with AML carrying t(6;9), DEK-CAN and FLT/ITD duplications [5]. Clinicians

Shuhei Okuyama and Katsushi Tajima contributed equally to this work.

✉ Shuhei Okuyama
s-okuyama@ypch.gr.jp

[1] Department of Hematology, Yamagata Prefectural Central Hospital, Yamagata 990-2292, Japan

[2] Department of Orthopedics, Yamagata Prefectural Central Hospital, Yamagata, Japan

[3] Department of Pathology, Yamagata Prefectural Central Hospital, Yamagata, Japan

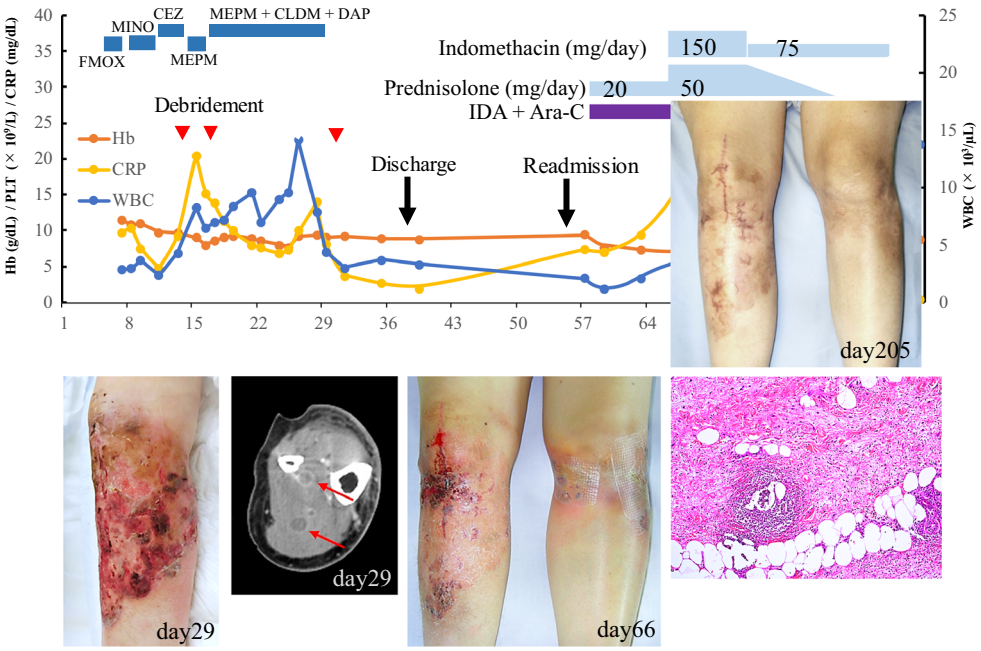

Fig. 1 The patient's clinical course. The primary right leg lesion seemed to have transiently improved, but a new painful lesion with edema was recognized on the left leg on day 66. A skin biopsy showed inflammatory infiltration of neutrophils in the dermis and adipose tissue and a small abscess was seen in part (hematoxylin and eosin stain, × 100 magnification). FMOX, flomoxef; MINO, minomycin; CEZ, cefazolin; MEPM, meropenem; CLDM, clindamycin; DAP, daptomycin; IDA, idarubicin; Ara-C, cytarabine

should attend to this variant and perform a skin biopsy promptly in unusual manifestations including no pathogens detected by recurrent microbiologic examinations.

Funding information This work was supported by a grant-in-aid for scientific research, KAKENHI Grant Number 80292423 (K. Tajima), from the Ministry of Education, Culture, Sports, Science, and Technology of Japan.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

Ethical approval This article does not contain any studies with animals performed by any of the authors.

Informed consent Informed consent was obtained from the patient included in this article.

References

1. Surovy AM, Pelivani N, Hegyi I, Buettiker U, Beltraminelli H, Borradori L (2013) Giant cellulitis-like Sweet Syndrome, a new variant of neutrophilic dermatosis. JAMA Dermatol 149(1):79–83. https://doi.org/10.1001/2013.jamadermatol.548
2. Kaminska EC, Nwaneshiudu AI, Ruiz de Luzuriaga A, Tsoukas M, Bolotin D (2014) Giant cellulitis-like Sweet syndrome in the setting of autoimmune disease. J Am Acad Dermatol 71(3):e94–e95. https://doi.org/10.1016/j.jaad.2014.03.025
3. Ozdogu H, Yeral M, Boga C (2017) An unusual giant leg ulcer as a rare presentation of Sweet's syndrome in a patient with hairy cell leukemia successfully managed by splenectomy. Turk J Haematol 34(3):270–271. https://doi.org/10.4274/tjh.2016.0416
4. Rochet NM, Chavan RN, Cappel MA, Wada DA, Gibson LE (2013) Sweet syndrome: clinical presentation, associations, and response to treatment in 77 patients. J Am Acad Dermatol 69(4):557–564. https://doi.org/10.1016/j.jaad.2013.06.023
5. Kazmi SM, Pemmaraju N, Patel KP, Cohen PR, Daver N, Tran KM, Ravandi F, Duvic M, Garcia-Manero G, Pierce S, Nazha A, Borthakur G, Kantarjian H, Cortes J (2015) Characteristics of Sweet Syndrome in patients with acute myeloid leukemia. Clin Lymphoma Myeloma Leuk 15(6):358–363. https://doi.org/10.1016/j.clml.2014.12.009