Histiocytoid Giant Cellulitis-Like Sweet Syndrome At The Site of Sternal Aspiration: A Case Report

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Case report

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Abstract

Background: Sweet syndrome with both histiocytoid pathology and giant cellulitis-like lesion feature is extremely rare and has only been reported once. Our case is different from the previous report because the cutaneous lesion was caused by local invasive irritation, which made it much more difficult to distinguish from cellulitis.

Case presentation: A 52-year-old male was diagnosed with myelodysplastic syndrome with multilineage dysplasia (MDS-MLD) associated with myelofibrosis (MDS-F) as well as angioneurotic edema of the tongue and floor of the mouth. Seven days after sternal aspiration, a cellulitis-like lesion was formed at the puncture site. Since he had neutropenia, history of glucocorticoid use and didn't keep the site dry and clean after aspiration, cellulitis was diagnosed, followed by broad-spectrum antibiotics and debridement. However, the lesion continued to expand, associated with blisters formation, accompanied by chills and fever. Blood cultures and blister smears didn't detect any pathogens. Biopsy of the lesion was performed and histiocytoid Sweet syndrome was diagnosed. He received prednisone treatment, and the fever relieved within 24 hours and the cutaneous lesion resolved within one week. He has had no recurrence during two-month follow-up.

Conclusions: This case can provide help for timely diagnosis and treatment and a reference for further summarizing the characteristics of this rare variant.

Background

Histiocytoid Sweet syndrome (H-SS) is an uncommon histopathologic variant of Sweet syndrome, which is characterized by an infiltration mostly composed of immature myelomonocytic cells with histiocytoid morphology[1]. Cutaneous lesions of H-SS are mostly similar to those of classical neutrophilic Sweet syndrome (N-SS), consisted of tender erythematous plaques and nodules on the extremities and trunk accompanied by systemic symptoms such as fever, arthritis, episcleritis and diarrhea[2, 3]. Giant cellulitis-like Sweet syndrome, a morphologically distinctive clinical variant, was reported recently. Its infiltrate of the dermis consists mainly of mature neutrophils[4]. Sweet syndrome with both histiocytoid pathology and giant cellulitis-like lesion feature is extremely rare and has only been reported once[5]. Here we report a patient with a cutaneous lesion caused by local invasive irritation, which is different from the previous report and much more difficult to distinguish from cellulitis.

Case Presentation

A 52-year-old male was admitted to our hospital due to progression of fatigue for one month and tongue swelling for one day. His tongue was swollen and painful after eating conch flesh the day before admission, and he has no fever, rash, or dyspnea. He didn’t have any positive past medical history. Positive physical examination findings were pallor, swollen tongue and splenomegaly (3cm below the left costal margin). Blood routine test revealed pancytopenia (WBC 1.82×10^9/L, absolute neutrophil count

1.07×10^9/L, platelet count 19×10^9/L, Hb 53g/L). Other laboratory findings included elevated lactate dehydrogenase (LDH, 351 U/L) and C-reactive protein (CRP, 84.4mg/L). Bone marrow aspiration yielded nearly “dry” tap even at the sternal. Combined with bone marrow biopsy, he was diagnosed with MDS-MLD associated with MDS-F. Angioneurotic edema of the tongue and floor of the mouth was diagnosed after dental consultation. After five days of treatment with dexamethasone (10mg/day) and loratadine, the swelling of the tongue was relieved.

Seven days after sternal aspiration (two days after discontinuation of dexamethasone), an erythematous nodule was formed at the puncture site, about 4 cm in diameter, sharply delimited, warm and tenderness. Considering the patient had neutropenia, history of glucocorticoid use and didn't keep the site dry and clean after aspiration, cellulitis was diagnosed, followed by broad-spectrum antibiotics and debridement. However, the lesion continued to expand to over 20cm in diameter, associated with blisters formation, accompanied by chills and fever, with a maximum body temperature of 40°C. Over three blood cultures for bacteria and fungi were negative, and blister smears didn't find any bacteria, fungi, or mycobacteria. Histopathology of the cutaneous lesion presented moderate edema in the papillary dermis, no evidence of vasculitis, and dense, inflammatory infiltrate involving the superficial and mid-dermis predominantly composed of MPO^+ $CD163^-$ mononuclear cells with vesicular twisted nuclei and scant eosinophilic cytoplasm (Fig. 1). Next-generation sequencing of the biopsy specimen didn't detect any pathogens. He was diagnosed with histiocytoid giant cellulitis-like Sweet syndrome and treated with prednisone (1mg/kg). The fever relieved within 24 hours and the skin lesion resolved within one week (Fig. 1). Thalidomide, erythropoietin and stanozolol were used to treat MDS, and prednisone was gradually tapered. He has had no recurrence during two-month follow-up.

Discussion And Conclusions

H-SS has a high frequency of association with hematological malignancies, especially myelodysplastic syndrome (MDS), and cutaneous lesions could precede the hematological diagnosis[6, 7]. The histopathologic differential diagnosis should be established with leukemia cutis and other inflammatory conditions characterized by a dermal infiltrate mostly composed of histiocytes[8]. The recurrence rate of H-SS patients is significantly higher than that of N-SS[3]. Treatment of the underlining disease is effective in reducing the risk of recurrence.

Giant cellulitis-like Sweet syndrome, a morphologically distinctive clinical variant, characterized by relapsing episodes, multifocal and widespread infiltrated plaques with bullous appearance, was reported recently. The infiltrate of the dermis consists mainly of mature neutrophils, and its relationship with malignancy remains to be clarified [4, 9, 10]. Histiocytoid giant cellulitis-like Sweet syndrome is extremely rare, the only reported case was a 72-year-old woman who developed a single lesion on her leg while gradually reducing the dose of prednisone for foot pain, and then diagnosed with unclassifiable myelodysplastic/myeloproliferative neoplasm[5]. Based on the clinical characteristics of these two patients, this unique variant seems to have an unifocal lesion and may closely related to myeloid malignancies, especially MDS. Our case is different from the previous report because the cutaneous

lesion was caused by local invasive irritation, in addition, the patient had neutropenia and received glucocorticoid therapy, the above conditions made it much more difficult to distinguish from cellulitis. Based on ineffective broad-spectrum antibiotics and debridement, no pathogenic evidence, rapid response to systemic corticosteroids, and combined with biopsy, the diagnosis can be confirmed. Early diagnosis will significantly reduce the suffering of patients. For this patient, hypersensitivity reaction to cutaneous irritation may be one of the predisposing factors. The previous angioneurotic edema also reflected his allergic physique. More cases and long-term follow-up are needed for summarizing the characteristics of this rare variant.

Abbreviations

CRP: C-reactive protein; H-SS: Histiocytoid Sweet syndrome; Hb: Hemoglobin; LDH: Lactate dehydrogenase; MDS-MLD: Myelodysplastic syndrome with multilineage dysplasia; MDS-F: Myelodysplastic syndrome associated with myelofibrosis; MDS: Myelodysplastic syndrome; MPO: Myeloperoxidase; N-SS: Neutrophilic Sweet syndrome; WBC: White blood cell.

Declarations

Ethics approval and consent to participate

All procedures performed were in accordance with the ethical standards of the institutional and national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. Informed consent was obtained from the patient.

Consent for publication

The patient consented to the publication of this case report and written informed consent was obtained from the patient.

Availability of data and materials

The datasets used in this case report are available from the corresponding author on reasonable request.

Competing interests

The authors declare that they have no competing interests.

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Authors' contributions

DZ and XS examined and treated the patient. JN was responsible for pathological diagnosis. DZ wrote the manuscript and made all the figures. XS supervised the manuscript. All authors read and approved the final manuscript.

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Figures

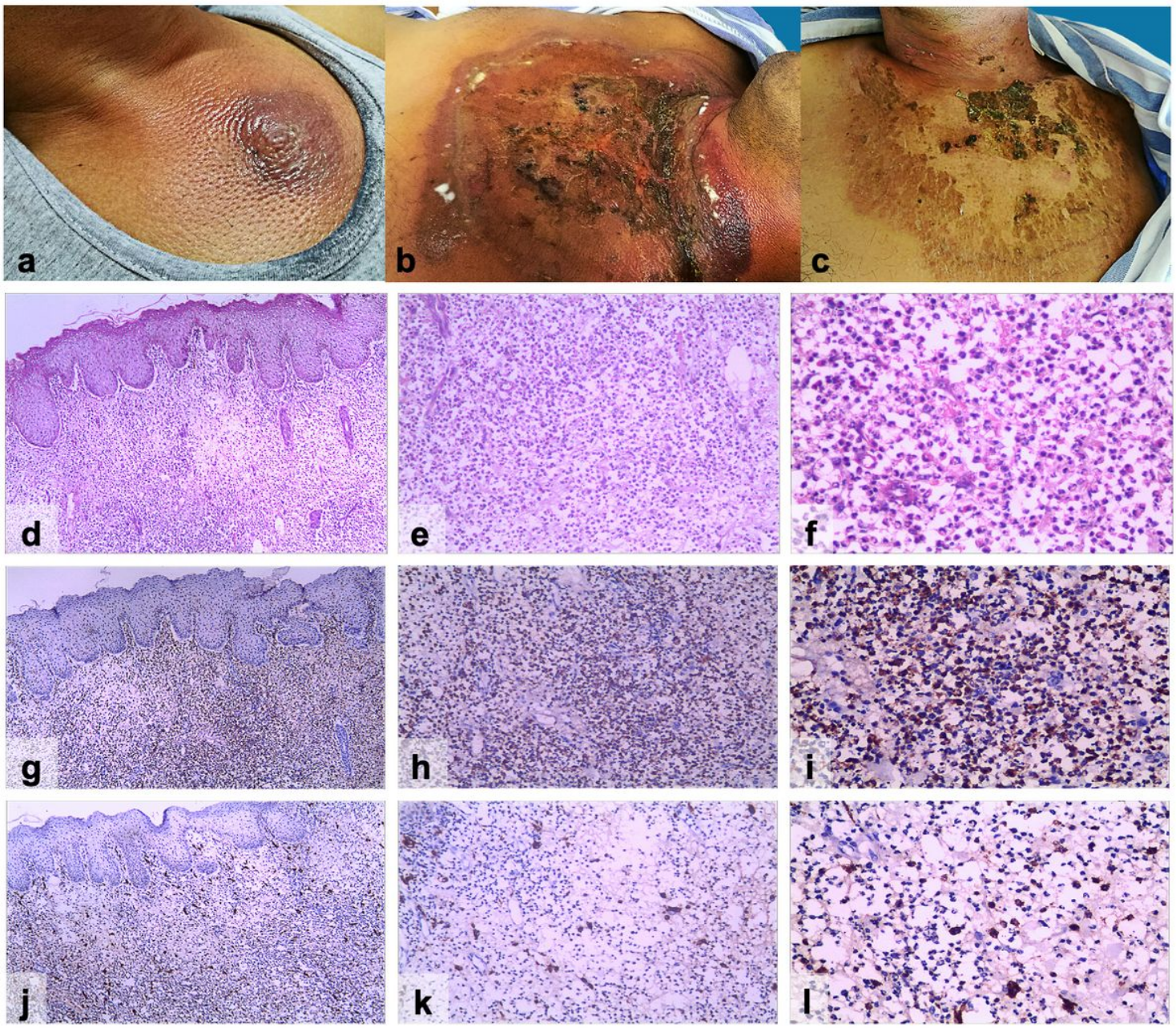

Figure 1

The cutaneous manifestation and pathological characteristics. a-c Characteristics and changes of the lesion. d-f Hematoxylin-eosin–stain showed moderate edema in the papillary dermis, the infiltrate was composed of mononuclear cells with vesicular twisted nuclei and scant eosinophilic cytoplasm (×40, 100 and 200 magnification from left to right). g-i Immunohistochemistry showed most of the infiltrating cells expressed MPO in the cytoplasm. j-l, Cells expressed CD163 in the cytoplasm constituted the scattered cells round the infiltrate. *MPO: myeloperoxidase