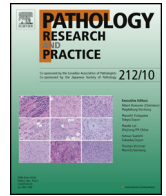




Contents lists available at ScienceDirect

Pathology – Research and Practice

journal homepage: www.elsevier.com/locate/prp



Original article

Clinicopathologic, immunophenotyping and cytogenetic analysis of Sweet syndrome in Egyptian patients with acute myeloid leukemia

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ARTICLE INFO

Article history:

Received 11 March 2016

Keywords:

Sweet syndrome
Leukemia
Subcutaneous
Genetic
Chemotherapy

ABSTRACT

Background: Sweet syndrome (SS) is an uncommon dermatologic disorder that could be associated with hematologic malignancies.

Objective: To describe the clinicopathologic, immunophenotyping and cytogenetic characteristics of SS in Egyptian patients with acute myeloid leukemia (AML).

Methods: The study was conducted during the period from April 2011 to March 2015. For each patient, a clinical evaluation and histological assessment of cutaneous lesions were recorded. Diagnostic investigations, immunophenotyping and cytogenetic features of leukemia were analyzed. Therapeutic monitoring and follow up of both diseases were registered.

Results: The study included 13 patients (7 males and 6 females) with a mean age of 44.4 ± 17.49 years. Fever was recorded in 10 cases and most of the lesions (61.5%) appeared during the post remission period. Clinically, the lesions were more frequently located on the extremities (61.5%), presented as solitary lesion (53.8%) and mostly tender (69.2%). Atypical presentations were observed in 5 cases and included ulcerative lesion, indurated mass and a gangrenous mass. Histological assessment revealed two patterns of inflammatory reactions described as classic (dermal) form (38.5%) and deep (subcutaneous) form (61.5%). Laboratory investigations showed leukocytosis in 61.5%, neutropenia in 38.5%, anaemia in 92.3%, and thrombocytopenia in 84.6%. Bone marrow aspiration and biopsy showed suppressed trilineage hematopoiesis in 84.6% and blast cell count $>50\%$ in 69.2%. The common subtypes of AML included M2 and M4 (23.1% for each). Cytogenetic studies revealed genetic abnormalities in 69.2% of cases. Most of the cases (76.9%) showed a poor response to oral prednisolone but responded well to alternative therapies, including dapsone, colchicine and cyclosporine.

Conclusion: Sweet syndrome associated with AML may show atypical clinical forms that have an aggressive course and is mostly associated with subcutaneous involvement. Although chemotherapy of AML may play a significant role in the development of SS, the exact mechanism remains unclear. The disease is considered a steroid refractory and genetic abnormalities may have a role in altering the classic nature of the disease.

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1. Introduction

Sweet syndrome (SS) is a characteristic dermatologic disorder described as acute febrile neutrophilic dermatosis. The disease is characterized clinically by a sudden onset of fever and painful

succulent red-brown plaques favoring face, shoulders and arms [1]. Histologically, SS is characterized by diffuse dermal infiltrate, mostly neutrophils, and is associated with prominent papillary dermal edema. Classically, the infiltrate is confined to the reticular dermis, and there are no signs of vasculitis [2].

Sweet syndrome may occur as an idiopathic reaction but it is commonly associated with other conditions, such as infections, autoimmune disorders, malignancy and drugs. It was reported that 21% of SS cases are associated with cancer; out of this percentage, approximately 85% are associated with hematologic malignancies.

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<http://dx.doi.org/10.1016/j.prp.2016.10.008>

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Table 1
Demographic and clinical characteristics of 13 patients with SS-associated AML.

No	Sex	Age	Fever	Symptoms/sings	Clinical characteristics of lesions (number – morphology – distribution)
1	M	25	+	Hepatoasplenomegaly	Solitary Ulcerative lesion Leg
2	M	41	+	Regional lymphadenopathy	Solitary Erythematous plaque Trunk
3	M	55	–	Chest pain	Multiple Erythematous plaques Upper limbs – Face
4	F	27	+	Gum hypertrophy	Solitary Indurated mass Arm
5	F	69	+	Ecchymosis	Multiple Erythematous nodules Upper extremities
6	M	56	++	Pallor	Multiple Ulcerative lesions Lower extremities
7	F	28	+	Gum hypertrophy	Solitary Gangrenous mass Thigh
8	F	81	+	Regional lymphadenopathy	Multiple Erythematous plaques Lower extremities
9	M	51	+	Ecchymosis	Solitary Indurated mass Trunk
10	F	42	–	Gum hypertrophy	Solitary Erythematous plaque Face
11	M	33	+	Pallor	Solitary Erythematous plaque Thigh
12	M	43	–	Chest pain	Multiple Erythematous plaques Trunk – upper limbs
13	F	26	++	Arthralgia	Multiple Erythematous plaques Trunk

Genitourinary tract malignancy constituted the most common solid tumor in association with SS [3].

Acute myelogenous leukemia (AML), chronic lymphocytic leukemia and multiple myeloma are considered the most common hematologic malignancies associated with SS [4]. In 10–15% of cases, an associated leukemia, usually of acute myelomonocytic type, is present or develops later. Hairy cell leukemia developed in one case. In some of these cases, features of atypical pyoderma gangrenosum may be present, and it has been suggested that Sweet's syndrome and pyoderma gangrenosum may be at opposite ends of the spectrum of one process [5].

To the best of our knowledge, there is no previous report describing the characteristics of SS in Egyptian patients. In this study, we describe the clinicopathological features of SS associated with AML and also describe the pattern of AML along with the cytogenetic and molecular abnormalities in these patients.

2. Patients and methods

During the period from April 2010 to March 2015, all patients with SS associated with AML were enrolled in this study. The study was conducted in three dermatology centers at different locations in Egypt. The study was approved by the local ethical committee and institutional review boards. Informed consent was obtained from each patient, and medical photography was performed for some patients after written consent.

Data recorded for each patient included age, sex; onset, course, location, description, and duration of the lesions. Skin biopsy was obtained from each lesion, and histological examination was evaluated. The workup of underlying hematologic malignancy, including bone-marrow aspiration and biopsy, immunophenotyping and cytogenetic studies, was registered. Therapeutic response and follow up information were also recorded for each patient.

For histological examination, skin specimen was fixed in 4% formalin, embedded in paraffin wax, cut and stained with hematoxylin-eosin. The histologic hallmark of the disease was the presence of moderately dense neutrophilic infiltrate in the reticular dermis associated with papillary dermal edema. Assessment of other histologic features, such as subcutaneous involvement, signs of vasculitis and presence of bacterial colonies, was reported. Special staining with Gram's, Ziehl–Neelsen (ZN) and Periodic acid Schiff (PAS) was performed routinely for all cases to exclude bacterial or fungal infections.

Laboratory investigations were recorded with more concern of total and differential leukocyte count and erythrocyte sedimentation rate (ESR). Radiological examination of the chest was performed for some cases. Blood culture was done for all cases with fever. Blood film was performed for all cases for identification of myeloid cells. Bone marrow aspiration and biopsy were collected

from the pelvic bone (iliac crest) under local anaesthesia. The samples were drawn into EDTA tube and heparin tube for cytogenetic studies and immunophenotyping.

Flow cytometric analysis was performed using a general panel of fluorescent antibodies against the antigens more specified of acute leukemia panel which included CD2, CD3, CD7, CD10, CD13, CD14, CD15, CD19, CD20, CD22, CD33, CD34, CD45, CD56, CD64, CD79a and CD117 (*APC Mouse Anti-Human*, *BD Pharmingen™*, *BD Biosciences, UK*). Other antibodies that were also used included Kappa and Lambda light chains, IgD, sIgM, Myeloperoxidase (MPO), terminal deoxynucleotidyl transferase (TdT) and HLA-DR (*SSP UniTray® Kit*, *Thermo Fisher Scientific, USA*). Samples were analyzed, and data acquisition and analysis were conducted by BD Cellquest Pro software [6].

For cytogenetic analysis, 0.5 ml of each bone marrow sample was collected in lithium heparin for fluorescence-in-situ hybridization (FISH) test. Samples were examined on direct short-term (24 h) cultures with at least 20 metaphases being analyzed. The technique was performed using LSI (local specific identifier) dual color translocation probe (*Abbott Molecular/Vysis, Des Plaines, IL, USA*) designed to detect any cytogenetic abnormalities [7].

The regimen of treatment was proposed for each patient according to the severity of the lesions and general condition of the patient. The standard treatment was administration of oral prednisolone (0.5 to 1 mg/kg/day). Monitoring of therapeutic response was assessed, and relapse rate was recorded.

3. Results

Out of 171 cases diagnosed as SS during the study period, 13 cases (8%) were associated with AML. The study included 7 males (54%) and 6 females (46%) with a mean age of 44.4 ± 17.49 years (ranged from 25 to 81 years). Fever was recorded in 10 cases (77%), and all cases showed a sudden onset of lesions (Table 1). In 3 cases (23%), SS was the presenting sign of AML, while in 2 cases (15%) the lesions developed during induction therapy. In 8 cases (62%), the lesions appeared during the post remission period; after the end of induction treatment and before consolidation therapy.

Clinically (Fig. 1), 7 cases (54%) presented with solitary lesion, while 6 cases (46%) presented with multiple lesions. The majority of lesions (61%) were located on the extremities (8 cases). Lesions were tender in 9 cases (69%), slightly pruritic in 3 cases (23%) and asymptomatic in one case (8%). The morphology of the lesions was variable, with a common presentation of classic erythematous and edematous nodules and/or plaques in 8 cases (61%).

Atypical presentations were observed in 5 cases (38%) in the form of ulcerative lesion (Fig. 2a), indurated mass (Fig. 2b) and gangrenous mass (Fig. 2c). Chest pain and arthralgia were a significant complaint in 3 cases (23%). Other clinical findings included gum

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