Sweet's Syndrome Associated with Chronic Myelomonocytic Leukemia

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Abstract **More than 85 per cent of patients with malignancy-associated Sweet's syndrome have a hematologic disorder, most commonly acute myelogenous leukemia. Only a few cases of this syndrome have been reported to be associated with chronic leukemia. We report a 52-year-old female who had Sweet's syndrome after having diagnosed of accelerated phase of chronic myelomonocytic leukemia for one month. She developed fever, swelling and erythrematous circumscribed plaque and reside at both cheeks and left arm. Fluid examination of the vesicle showed no organism. Biopsy of the skin was done and was consistent with Sweet's syndrome. Prednisolone therapy was administered with improvement. However, the patient developed uncontrolled bleeding due to complicated sepsis and DIC.**

เรื่องย่อ **Sweet's Syndrome Associated with Chronic Myelomonocytic Leukemia**

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ผู้ป่วยที่เป็น Sweet's syndrome ที่พบร่วมกับโรคมะเร็ง มากกว่าร้อยละ 85 พบร่วมกับมะเร็งของระบบเลือด โดยเฉพาะ acute myelogenous leukemia มีผู้ป่วยเพียงไม่กี่รายที่เป็น Sweet's syndrome ที่มีความสัมพันธ์กับ chronic leukemia คณะผู้รายงานขอเสนอผู้ป่วยหญิงอายุ 52 ปี ที่ได้รับการวินิจฉัยว่าเป็น accelerated phase ของ chronic myelomonocytic leukemia ต่อมามีไข้ ผื่นบวมแดงเป็นปื้นที่บริเวณแก้มและแขนซ้าย จากการตรวจน้ำที่ vesicle ไม่พบเชื้อที่เป็นสาเหตุ ผล skin biopsy เข้าได้กับ Sweet's syndrome รอยโรคที่ผิวหนังดีขึ้นอย่างรวดเร็วหลังจากได้รับการรักษาด้วย เพรดนิโสโลนวันละ 30 มก. 2 สัปดาห์ต่อมา ผู้ป่วยอาการเลวลงอย่างรวดเร็ว เนื่องจากมีเลือดออกทั่วไปซึ่งเป็นผลมาจาก sepsis และ DIC

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Sweet's syndrome or acute febrile neutrophilic dermatosis (AFND) was first described by Sweet in 1964[1]. The characteristics of this syndrome are fever, neutrophilia, multiple tenderness erythematous papule, nodule, vesicle or plaque with sharp demarcated border mainly at face, neck, upper extremities and occasional at lower extremities. Histology shows a dense dermal infiltration with mature neutrophil polymorphs. These lesions respond dramatically to corticosteroids with absence of scarring.

Sweet's syndrome is commonly associated with upper respiratory tract infections, ulcerative colitis, benign monoclonal gammopathy, myeloproliferative disorder or malignancy[2]. More than 85 per cent of patients with malignancy-associated Sweet's syndrome have a hematologic disorder, most commonly acute myelogenous leukemia[3-15]. Only a few cases have been reported in association with chronic myeloid leukemia[16-20] but no cases associated with chronic myelomonocytic leukemia (CMMoL) have been previously mentioned.

We report a case of CMMoL who developed Sweet's syndrome.

Table I. Clinical feature and response to corticosteroids of AFND during hematological disorders : a review of previously reported cases.

Case (reference)	Age/Sex	Hematological disorder	Location of lesions	Treatment
1 (7)	16/F	AML	extremities	cortisone (healed)
2 (8)	38/F	ALL	ears,tongue,forearm	resolved spontaneously
3 (8)	52/F	ALL	breast,head,neck, extremities	prednisone (resolved)
4 (3)	10/M	APL	neck,ear,shoulder	prednisone (resolved)
5 (3)	45/F	AML	legs,arms,lip,thigh	prednisone (resolved)
6 (15)	47/F	AML	shoulders,upper back, face,legs	prednisone (resolved)
7 (9)	55/F	AML	face,arms,shoulders	prednisone (resolved)
8 (10)	50/F	AML	hand,forearm,wrist, leg	prednisone (resolved)
9 (4)	39/M	AML	hand,forearm,scalp, face,neck,ears	prednisone (resolved)
10 (11)	45/F	AMMoL	face,neck,chest, mouth,extremities	prednisone (resolved)
11 (11)	51/F	AML	wrists,hands,arms, face,neck	prednisone (resolved)
12 (5)	73/M	AML	face,neck,chest, arm,legs	resolved spontaneously
13 (12)	50/F	AML	not specified	corticosteroid (resolved)
14 (6)	27/F	AMMoL	face,arms	prednisone (resolved)
15 (6)	29/M	AMMoL	face,head,hands	prednisone (resolved)
16 (13)	50/F	AMMoL	arm,wrist,legs,	prednisone (resolved)

Case (reference)	Age/Sex	Hematological Disorder	Location of lesions	Treatment
17 (14)	38/M	AML	neck,shoulder (only sweat glands)	resolved spontaneously
18 (15)	62/F	AML	hand,forearm,vulvar area	prednisone (90% resolved)
19 (6)	49/M	RAEB	face,neck,back, chest,arms	prednisone (resolved)
20 (6)	72/M	RAEB	face,arms,upper trunk,thighs,legs	prednisone (resolved)
21 (23)	58/F	NHL[1]	legs	prednisone (resolved)
22 (24)	58/M	NHL[2]	not specified	corticosteroid (resolved)
23 (25)	32/M	MF	not specified	corticosteroid (resolved)
24 (15)	66/F	MF	arms,axillas,breast	triamcinolone (90% resolved)
25 (16)	19/F	CML-B	not specified	corticosteroid (resolved)
26 (17)	46/F	CML-B	not specified	corticosteroid (resolved)
27 (18)	33/F	CML-A	shoulder,face,neck	prednisone (60% resolved)
28 (20)	64/F	CML-C	hand,fingers	prednisone (resolved)
29 (19)	52/M	CML-C	face,neck,arm, trunk,legs	prednisone (resolved) Naproxen
39 (this article)	52/F	CMMoL-A	face,arm	prednisone (resolved)

AML - Acute myeloid leukemia ; AL - acute leukemia (not specified) ; APL - acute promyelocytic leukemia ; AMMoL - acute myelomonocytic leukemia ; RAEB - refractory anemia with excess of blast ; MF - idiopathic myelofibrosis ; HNL[1] - non-Hodgkin's lymphoma (diffuse mixed lymphocytic-histiocytic), NHL[2] - non-Hodgkin's lymphoma (disffuse histiocyte) ; CML-A - chronic myeloid leukemia (accelerated phase) ; CML-B - chronic myeloid leukemia (blastic phase) ; CML-C - chronic myeloid leukemia (chronic phase) ; CMMoL-A - chronic myelomonocytic leukemia (accelerated phase)

Case report

A 52-year-old female, who had been healthly until March 1991, developed fatique, anorexia, low graded fever and anemia for two weeks. On examination, T 38.5°C, she was found marked anemia, liver 2 F.B. below right costal margin with 12 cms span and palpated tip of spleen. No lymphadenopathy was noted. Laboratory examination showed Hct 12%, leukocytes 11,750/cu.mm with 52% neutrophil, 12% lymphocytes, 1% basophils, 1% monocytes, 1% promylocyte and 40% blasts, platelet 285,000/cu.mm. Bone marrow demonstrated slightly increased cellularity and moderate increase of megakaryocytes with clumpping of platelet, 22% blasts (myeloblast

and monoblast), 13% monocyte, 12% lymphocytes, 17% histiocytes, 2% plasma cells and 10% erythroid series. She was diagnosed as having CMMoL and was treated with 6-mercaptopurine 50 mg twice daily and blood transfusion. Three weeks later, she developed high fever, frequently rising over 38.5°C, swelling and erythematous papule, vesicle and plaque with sharp demarcated border at face and right arm without tenderness. Laboratory investigation showed Hct 17%, leukocyte 19,500/cu.mm (96% neutrophils, 4% lymphocytes, platelet 165,000/cu.mm. Vesicular fluid examination showed no organism and culture were negative for bacteria and fungus. Blood culture was done and reported no growth. She was first diagnosed as having cellulitis and cloxacillin plus gentamicin were started. A diagnosis of AFND was made by skin biopsy. She received prednisolone 30 mg/d and skin lesions was rapidly improved in 3 days. Unfortunately 4 days later she developed pneumonia, sepsis, DIC and rapidly deterioration because of uncontrol bleeding. On her relatives request, the patient was discharged from the hospital without any further treatment.

Discussion

The cutaneous manifestation myelogenous leukemia can be divided into two groups, the leukemid or nonspecific lesion and the true leukemic infiltration or specific lesion[21]. Sweet's syndrome belongs to the first group. Skin lesions at Sweet's syndrome should be differentiated from the true leukemic infiltration, cellulitis, erythema multiforme, panniculitis, erythema nodosum, herpes simplex and SLE. If one cannot distinguish Sweet's syndrome from those conditions, skin biopsy should be done.

The etiology of Sweet's syndrome is still unknown. Abnormal chemotactic stimuli from leukemic cells or abnormal chemotactic responses of neutrophil have been postulated to be the cause of Sweet's syndrome[22] in myelogenous leukemia. An increase production of factor(s) responsible for abnormal chemotactic stimuli from leukemic cells may precipitate the occurrence of the skin lesion. Some patients have spontaneous resolved of skin lesions within 2 weeks to four months[6] but mostly respond rapidly to corticosteroid without scarring. NSAID may be an alternative treatment for patients with Sweet's syndrome who do not respond to corticosteroids[20] and for those who are immunocompromised.

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