



## Sweet's syndrome and myelodysplasia: two entities with a common pathogenetic mechanism? A case report

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A 60-year old woman presented with fever and painful skin lesions of the lower extremities. At physical examination she had large elevated and infiltrated plaques ranging from 4 to 6 cm in diameter, on the knees, feet and pretibial regions (Figure 1). Her white blood cell (WBC) count was  $12 \times 10^9/L$  with 77% polymorphonuclear cells; hemoglobin (Hb) was 10.7 g/dL, platelet count  $86 \times 10^9/L$ , erythrocyte sedimentation rate was 103 mm/hr. Bone marrow aspiration showed a picture consistent with a diagnosis of acute myeloid leukemia (AML) probably secondary to a myelodysplastic syndrome (MDS). A biopsy of a skin lesion showed a dense dermal infiltrate characterized by neutrophils with nuclear dust, vasodilatation with swollen endothelial cells without signs of vasculitis (Figure 2). These findings enabled us to diagnose Sweet's syndrome (SS), according to Su and Liu's criteria revised by Von den Driesch.<sup>1</sup> Treatment with steroids (betamethasone 4 mg i.v. and then, 8 mg i.v.) led to disappearance of the fever and improvement of skin lesions until complete resolution and four months later a hematologic evaluation showed: WBC  $17 \times 10^9/L$  (72% of polymorphonuclear cells), Hb 13.3 g/dL, platelets  $229 \times 10^9/L$ . A bone marrow examination at that time showed a decrease of the percentage of myeloid blasts to less than 20%. After a further six months, the patient presented with skin lesions on the eyelids and a worsening of the hematologic picture with a WBC count of  $58 \times 10^9/L$  (88% of myeloid blasts), a platelet count of  $15 \times 10^9/L$ , and about 90% of myeloid bone marrow blasts. She underwent chemotherapy, but did not respond and died of an infective complication.

SS is an acute febrile neutrophilic dermatosis characterized by fever, neutrophilic leukocytosis, cutaneous erythematous and painful plaques, histologic findings of a dense neutrophilic dermal infiltrate without vasculitis, and a prompt response to systemic corticosteroid therapy.<sup>1</sup> This condition can be idiopathic, para-inflammatory, paraneoplastic, or pregnancy-related.<sup>1</sup> In the majority of the paraneoplastic cases the associated neoplasm is hematologic.<sup>2</sup>

The pathogenesis of SS is still undefined. Inappropriate cytokine secretion has been hypothesized as a possible mechanism: endogenous serum levels of cytokines (G-CSF, IL-6) have been described to correlate with the different phases of SS.<sup>3</sup> Furthermore, the development of SS after G-CSF therapy in patients with hematologic diseases has been



Figure 1. Sweet's syndrome: large elevated and infiltrated plaques on lower limbs.

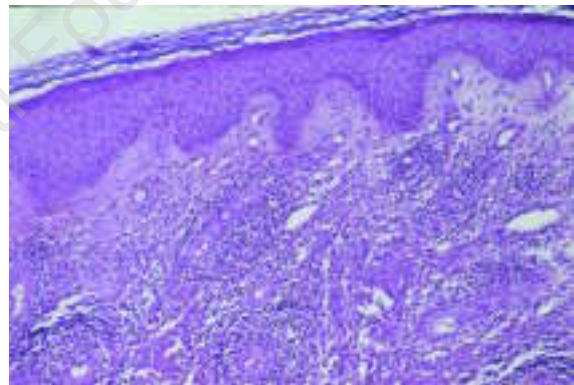


Figure 2. Skin lesions biopsy.

described.<sup>4,5</sup> On the other hand, there are reported cases of progression of MDS to an overt blastic phase after G-CSF therapy.<sup>6</sup>

The peculiarity of the case reported here, is the unusual concurrent course of the hematologic and dermatologic syndromes. In fact, at presentation, during the acute phase of the SS, the bone marrow examination showed a picture of AML. By contrast, as the skin lesions improved, after the administration of steroids alone, the percentage of blastic cells in the bone marrow fell to less than 20%.

So, although we did not measure the serum levels of G-CSF, it is possible to hypothesize that the presence of high levels of G-CSF may represent a triggering factor of the acute phase of both syndromes.

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