The differential diagnosis of this skin eruption associated with fever in the setting of acute myelogenous leukemia (AML) included infection, drug hypersensitivity, erythema multiforme, and acute febrile neutrophilic dermatosis (Sweet's syndrome). Given the high-dose broad-spectrum antibiotics, viral etiologies were considered the most likely infectious agents. The presence of vesicles led to the suspicion of disseminated herpes zoster (VZV). Direct fluorescence antibody for VZV and viral cultures for VZV, herpes simplex, and cytomegalovirus were performed and found to be negative. Piperacillin and tazobactam for injection and vancomycin were known causes of acute, severe drug eruptions with blistering and were thus discontinued. However, a skin biopsy demonstrated a dense dermal neutrophilic infiltrate without evidence of vasculitis or bacterial or fungal organisms, which was typical of acute febrile neutrophilic dermatosis (AFND) or Sweet's syndrome.

**DISCUSSION**

Sweet's syndrome was first described by the British dermatologist Robert Douglas Sweet in 1964. Classically, Sweet's presents abruptly with multiple, distinct, painful erythematous plaques on the face, neck, upper chest, back, and extremities. The lesions are sometimes described as having a “mountain-range relief” appearance. Vesiculation, pustulation, and blistering can occur. Overall, 80% to 90% of affected patients present with fever.

**SUBTYPES**

Sweet's syndrome generally is classified into idiopathic, drug-induced, or malignancy-associated subtypes. These subtypes have distinct but overlapping clinical presentations. Thirty percent of Sweet's cases are malignancy associated, and of these 85% are hematologic—most commonly AML but also including chronic myelogenous leukemia, Hodgkin's and non-Hodgkin's lymphoma, and myelodysplastic syndrome. Compared with the idiopathic form, hematologic malignancy-associated Sweet's syndrome is more likely to be severe and to present with vesicles, bullae, or ulcers. Hematologic malignancy-type Sweet's syndrome commonly presents with abnormal platelet counts (thrombocytosis or thrombocytopenia) and anemia, both of which are rare in the idiopathic type. Finally, whereas the idiopathic type commonly presents with neutrophilia (80%), more often than not patients with hematologic malignancy do not have peripheral neutrophilia (47%).

**DIAGNOSTIC CRITERIA**

In 1986, Su and Liu proposed major and minor criteria for diagnosing Sweet's, which have gained general acceptance. The major criteria include: 1) abrupt onset of painful erythematous plaques or nodules; and 2) dense neutrophilic infiltrate on pathology without evidence of vasculitis. They also described minor criteria including: 1) underlying malignancy; 2) favorable response to glucocorticoids but not to antibiotics; 3) fever >38ºC; and 4) 3 of 4 abnormal laboratory results (erythrocyte sedimentation rate >20 mm/hr, C-reactive protein, white blood cell count >8000, neutrophils >70%). Both the major criteria and 2 of 4 minor criteria are necessary for definitive diagnosis.

**PROGNOSIS**

Patients with Sweet's syndrome respond well to systemic corticosteroids. Fever subsides within 2 to 3 days; skin lesions heal within 3 to 9 days. The rate of recurrence is 60% in patients with hematologic malignancies as long as the underlying malignancy is still present. Infection from breakage in the skin barrier is a major source of morbidity, and these manifestations are also responsive to corticosteroids.

In this case, we minimized further immunosuppression, which had already been induced by chemotherapy, and the patient was treated with prednisone 40 mg/day. There was improvement of skin lesions and fever within 2 days and clearing of the eruption in 7 days.

**References**

3. Cooper PH, Innes DJ Jr, Greer KE. Acute febrile neutrophilic dermatosis (Sweet's syndrome) and myeloproliferative disorders. Cancer. 1983;51:1518-1526
CASE OF THE MONTH

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The goal of Johns Hopkins Advanced Studies in Medicine and this activity is to provide continuing medical education to primary care physicians and reinforce their existing dermatology and clinical pathology knowledge base.

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