A 72-year-old man was admitted to the hospital with a fever as high as 39°C; mild dyspnea; and tender, erythematous, vesicular skin lesions on his lower legs, forearms, and neck (Fig 1). A chest radiograph showed patchy consolidation in the right upper, right lower, and left lower lobes. Medical history was remarkable for a 1-year history of myelodysplastic syndrome. On admission, his hemoglobin level was 8.5 g/dL, and WBC count was 6 × 10^9/L, with a differential of 52% segmented neutrophils, 20% lymphocytes, 26% monocytes, and 2% eosinophils. The platelet count was 80 × 10^9/L. Appropriate blood, sputum, urine, and skin cultures were obtained, and the patient was empirically started on IV ampicillin.

Over the next 36 h, the skin lesions and dyspnea worsened. Culture results were negative. A skin biopsy revealed a sterile, diffuse, nodular infiltration of the dermis by mature neutrophils. BAL revealed a WBC count of 11.2 × 10^9/L, with a differential of 90% neutrophils, 3% eosinophils, and 7% macrophages. No pathogenic organisms or malignants cells were found. Transbronchial biopsy revealed interstitial infiltration by large numbers of neutrophils with occasional eosinophils and small numbers of lymphocytes. There was no evidence of granulomatous inflammation. At this point, which of the following is the correct treatment?

A. Trimethoprim-sulfamethoxazole  
B. Ceftazidime and erythromycin  
C. Methylprednisolone  
D. Paclitaxel and carboplatin  
E. Colchicine
Answer: C. Methylprednisolone.

This patient has Sweet’s syndrome (acute febrile neutrophilic dermatosis) with lung involvement. If recognized and treated, a dramatic response to corticosteroids is typical.

Sweet’s syndrome was originally described in 1964 and is characterized by fever, peripheral neutrophilia of the blood, multiple tender cutaneous plaques on the skin, and a dense infiltration of the dermis by mature neutrophils. Typical skin lesions, as seen in our patient, are raised erythematous plaques or nodules that range in size from 0.5 to 12.0 cm in diameter and may appear vesicular or bullous. Sweet’s syndrome is frequently observed in association with other illnesses. In 10 to 20% of cases, the syndrome may be associated with a malignancy, especially hematologic diseases—acute myelogenous leukemia in nearly half of these.

Extracutaneous manifestations such as episcleritis, arthralgias, proteinuria, and hepatitis may occur in up to 50% of malignancies associated with Sweet’s syndrome. Neutrophilic infiltration of the lung, however, is not common. Histologically, the pulmonary inflammation resembles that of the skin, consisting predominantly of mature neutrophils infiltrating the interstitium in the absence of pathogenic organisms. BAL analysis shows evidence consistent with a florid neutrophilic alveolitis.

Because use of systemic corticosteroids is the treatment of choice for Sweet’s syndrome, the other therapeutic options are not appropriate.

Suggested Readings