CASE REPORTS

Neutrophilic Dermatosis of the Dorsal Hands

C Laguna, JJ Vilata, and B Martin
Servicio de Dermatología, Hospital General Universitario de Valencia, Spain

Abstract. Neutrophilic dermatosis of the dorsal hands is a recently described disorder. It is still debatable if it constitutes a separate entity, a variant of Sweet syndrome or pyoderma gangrenosum or an overlap disorder of both. We report the clinical features, histopathological findings, and evolution of a 35-year-old patient with the diagnosis of neutrophilic dermatosis of the dorsal hands and distant lesions on the forehead.

Key words: neutrophilic dermatosis of the dorsal hands, Sweet syndrome, pyoderma gangrenosum.

Introduction

Neutrophilic dermatosis of the dorsal hands is a recently described disorder. Clinical presentation, laboratory data, histopathological findings, and response to systemic corticosteroids suggest that this entity is in fact a localized variant of Sweet syndrome and that it is identical to atypical pyoderma gangrenosum when it presents on the hands.1

Case Description

We present the case of a 35-year-old woman who came to the emergency department because of linearly distributed blood-filled blisters, which initially presented as a rash, on the dorsum of the right wrist. She also presented small papulovesicular lesions on the dorsal hands (Figure 1) and indicated that she had had no previous trauma. The most notable aspect of her clinical history was that she was allergic to penicillin and had taken benzodiazepines due to a stressful family situation. The initial suspicion was of stings and she was prescribed sulfated water, an oral macrolide, and topical mupirocin. Her symptoms had worsened 48 hours later with the appearance of exudative erosions on the wrist, and violet erythematous pustules and nodules with an edematous border, some of which had central ulceration, on both dorsal hands (Figure 2). She also had an edematous erythematous plaque with a raised edge on the forehead (Figure 3).

The patient remained afebrile at all times, was in a good general state of health, and had no associated enlarged lymph nodes or systemic symptoms. The laboratory workup revealed leukocytosis with a cell count of 14,500, a neutrophil count of 10,800, and an erythrocyte sedimentation rate of...
54 mm/h. A biopsy revealed a dense polymorphonuclear infiltrate in the dermis with no leukocytoclastic vasculitis (Figure 4). The definitive diagnosis was neutrophilic dermatosis on the dorsa of the hands with distant lesions. Oral prednisone was prescribed at a dose of 1 mg/kg/d and the lesions resolved immediately (Figure 5). The remaining complementary tests—biochemistry, antinuclear antibodies, virus serology, tumor markers, lymphocyte counts, thyroid hormones, and chest radiograph—showed no abnormalities, with the exception of a urine sediment indicating an asymptomatic urinary infection. The patient is currently under observation without treatment and, to date, has not had a relapse.

Discussion

In 1995, Strutton et al² coined the term pustular vasculitis of the dorsal hands to refer to the appearance of painful violaceous erythematous nodular lesions with pustules and blisters in this area. Patients presented with or without associated systemic involvement. Histological examination revealed a dense polymorphonuclear inflammatory infiltrate associated with leukocytoclastic vasculitis. In 2000, Galaria et al³ presented 3 patients with similar lesions that were refractory to antibiotic therapy and responded well to systemic corticosteroids. Recurrences were treated effectively with dapsone. The histopathological study revealed a polymorphonuclear infiltrate with no leukocytoclastic vasculitis; thus, the authors preferred the term neutrophilic dermatosis of the dorsal hands, suggesting that this could be a local variant of Sweet syndrome or an overlap of Sweet syndrome and pyoderma gangrenosum. Sharareh⁴ used the term pyoderma gangrenosum of the dorsal hands to refer to this condition and included it in the atypical manifestations of pyoderma gangrenosum. In their recent review of 52 published cases, Walling et al¹ show that 27% of cases had associated neoplastic disease, 21% of which
were hematological neoplasms. Inflammatory bowel disease affected 15% of patients. Most presented fever or leukocytosis and/or an increase in the erythrocyte sedimentation rate. Treatment with systemic corticosteroids was successful in 71% of cases. Most of the published cases were initially diagnosed as infection but antibiotic therapy failed.

At present, it is unknown whether these symptoms are indeed a variant of Sweet syndrome, pyoderma gangrenosum, an overlap of the two, or a separate entity. However, Walling et al believe that neutrophilic dermatosis of the dorsal hands is a variant of Sweet syndrome and that it is identical to atypical pyoderma gangrenosum and pustular vasculitis of the hands; therefore, they propose the term neutrophilic dermatosis of the dorsal hands as the most suitable when referring to these symptoms, preferring it to neutrophilic dermatitis of the hands, as proposed by Weening et al, to reflect the primary localization in most cases.