Two Pediatric Cases of Nonbullous Histiocytoid Neutrophilic Dermatitis Presenting as a Cutaneous Manifestation of Lupus Erythematosus

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Background: Nonbullous neutrophilic dermatoses are seen infrequently in association with lupus erythematosus (LE). A recently described histopathologic variant of Sweet syndrome, to our knowledge, histiocytoid Sweet syndrome (HSS) has not been described in either pediatric or adult patients with LE.

Observations: We describe 2 pediatric patients with nonbullous histiocytoid neutrophilic dermatitis in the setting of LE. One case represents the initial presentation of subacute cutaneous LE, while the other case represents a manifestation of established systemic LE. Both cases demonstrate histopathologic findings of HSS.

Conclusions: We believe that the dermatosis observed in these 2 patients represents a nonbullous histiocytoid neutrophilic dermatosis that is best termed HSS. This entity may represent a distinct and important cutaneous manifestation of LE. Additional study is needed to further elucidate the relationship between neutrophilic dermatitis and LE.

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ever, the DIF slides did reveal nuclear staining with IgG of keratinocytes, consistent with the positive results of an antinuclear antibody test and the presence of anti–ribonucleoprotein antibodies, which this patient had.

Treatment was initiated with pulsed methylprednisolone and dapsone. The patient responded well to the corticosteroids and experienced a slow but almost complete response to dapsone after 6 months of therapy.

CASE 2

A previously healthy 5-year-old girl presented with an asymptomatic facial eruption of several months’ duration. It initially appeared on the ears and eventually spread to involve the nasal bridge and malar cheeks. She had no systemic symptoms. She had received multiple courses of antibiotics for presumed cellulitis, but the eruptions seemed to resolve and recur on average every 3 months regardless of antibiotic treatment.

Physical examination findings revealed well-defined erythematous plaques of the helix, both annular and linear in configuration. There were several arcuate edematous and erythematous plaques with overlying crust extending over the nasal bridge onto the malar cheeks. (Figure 2A).

Initial laboratory examination revealed a normal white blood cell count and normal hemoglobin and complement levels. Findings for anti–Sjogren syndrome antigen B anti-
Neutrophilic dermatoses are commonly seen in association with connective tissue disease, but they are infrequently reported as a manifestation of LE. Bullous SLE is a well-documented but rare neutrophilic dermatosis that manifests as a vesiculobullous eruption in the setting of SLE. Palisaded neutrophilic granulomatous dermatitis (PNGD), also known as interstitial granulomatous dermatitis with arthritis, represents an uncommon condition characterized by a nonbullous symmetric papular eruption and has been described as a manifestation of connective tissue disease, including LE.1 One case of PNGD in a pediatric patient with LE has been reported.3 In addition, isolated case reports of pyoderma gangrenosum in association with SLE are described in the literature.1,5

Sweet syndrome and Sweet-like eruptions with histopathologic findings including a diffuse neutrophilic infiltrate and papillary dermal edema have also been described as uncommon manifestations of LE.6-13 However, many of the patients with LE who present with these neutrophil-rich eruptions do not meet the diagnostic criteria for classic Sweet syndrome. Gleason et al7 have postulated that this nonbullous neutrophilic dermatosis, or Sweet-like eruption, is an underrecognized variant of cutaneous LE.

Of interest, the histopathologic findings of all cases of neutrophilic dermatosis in the context of established LE suggested LE.5,7,8 Conversely, the biopsy specimens from patients with no history of LE did not reveal such
Evidencing the spectrum of histopathologic findings. They coined the term histiocytoid Sweet syndrome, thus extending histiocytoid Sweet syndrome, both histopathologically and immunohistochemically.

One of the distinctive aspects of our 2 pediatric LE cases is that the histopathologic findings in both revealed not only interstitial and diffuse infiltrates of segmented neutrophils but also monocytoid cells resembling histiocytes. These histiocytelike cells were confirmed to be of myeloid lineage by positive staining for myeloperoxidase and CD68. In 2005, Requena et al first coined the term histiocytoid Sweet syndrome, thus expanding the spectrum of histopathologic findings. They described 41 cases of cutaneous lesions consistent with Sweet syndrome in which biopsy specimens showed cutaneous infiltrates, including many histiocytelike cells that were confirmed to be of myeloid lineage by immunohistochemical analysis. Six of the 41 patients had associated malignant neoplasms. None of the patients were children. The course of the disease was similar to that of classic Sweet syndrome, with prompt resolution of the cutaneous lesions after administration of systemic corticosteroid therapy. Chow et al more recently elaborated on this phenomenon as they reported 6 additional cases of inflammatory skin disease characterized by dermal and/or subcutaneous infiltrates of histiocytoid myeloid cells. Most of their cases did not fulfill the strict criteria for a diagnosis of classic Sweet syndrome, and therefore they referred to this spectrum of observations as “histiocytoid neutrophilic dermatoises and panniculitides.”

Although the histopathologic findings from both of our pediatric patients with LE were consistent with HSS as described by Requena et al, the criteria for a diagnosis of Sweet syndrome were not met by either of our patients. In addition, because the clinical picture of our patients was not typical of classic Sweet syndrome, we do not believe that Sweet-like represents the best descriptive term for these 2 cases. We have chosen the designation nonbullous histiocytoid neutrophilic dermatitis to generally describe the observed cases in this report, which we believe is both descriptive and accurate. The presence of an interstitial infiltrate of histiocytoid myeloid cells in the setting of connective tissue disease raises a differential diagnosis that includes interstitial granulomatous dermatitis and interstitial granuloma annulare (Table). Intertstitial granulomatous dermatitis differs histopathologically from HSS and nonbullous histiocytoid neutrophilic dermatitis in that it is characterized by a mixed infiltrate that includes both histiocytes and neutrophils, with histiocytes sometimes palisaded around foci rich in neutrophils or around a vasculitic nidi. The clinical morphologic characteristics of interstitial granulomatous dermatitis also differ from those observed in our patients. In interstitial granuloma annulare, the infiltrate is composed of authentic histiocytes rather than histiocytoid myeloid cells, and thus the findings are distinct from HSS nonbullous histiocytoid neutrophilic dermatitis, both histopathologically and immunohistochemically.

In conclusion, we report 2 pediatric cases of nonbullous neutrophilic dermatitis, a variant of the recently described HSS, presenting as a cutaneous manifestation of LE. We believe that these cases further support the conclusions of Gleason et al,6 that nonbullous neutrophilic dermatitis should be considered a possible skin manifestation of LE. Moreover, the histopathologic findings of a histiocytoid neutrophilic dermal infiltrate may represent an important clue to the diagnosis of LE.

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