



SKIN MANIFESTATIONS OF INTERNAL DISEASE

HISTOPATHOLOGIC CHARACTERIZATION OF THE CUTANEOUS MANIFESTATIONS OF HYPEREOSINOPHILIC SYNDROME

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Introduction: Hypereosinophilic syndrome (HES) is a rare disease that is characterized by elevated eosinophilic counts above $1.5 \times 10^9/l$ for over 6 months without any underlying causes. Cutaneous involvement is a prominent feature and may be the presenting sign of the disease. Despite this, histopathologic findings in skin biopsies have not been systematically characterized in a large, heterogeneous HES disease cohort.

Objective: We sought to systematically characterize the cutaneous features of HES through analysis of histopathology.

Materials and Methods: Subjects with dermatologic manifestations of HES were identified by retrospective chart review of 326 subjects referred for evaluation of HES between 1983 and 2017. Clinical characteristics were recorded and available skin biopsies reviewed by a single pathologist.

Results: Consistent with prior reports, 46% of subjects with HES had skin manifestations, ranging from urticaria to necrotic ulcers. 51 biopsies were available and systematically evaluated. Eosinophilic infiltration, as assessed by H&E staining, was absent (0-1/hpf) in 24% of subjects and $>10/hpf$ in only 47% of biopsies. When present, the pattern was typically superficial, dermal and predominantly perivascular across HES subtypes. Of those with no eosinophils on biopsy, the majority of subjects were on systemic corticosteroids.

Conclusions: Although cutaneous manifestations are common in HES, skin histology does not always reveal dense eosinophilic infiltration and other cell types, including lymphocytes and neutrophils, may predominate. HES biopsies were heterogeneous in nature reflecting the varied skin manifestations even within a similar HES subtype category. Lack of eosinophils on histology was associated with treatment with corticosteroids despite presence of peripheral eosinophilia ($>500/\mu L$) in half of the subjects at the time of biopsy. No individual histologic feature such as pattern of inflammation, depth or location of inflammation was able to distinguish between subtypes of HES patients. Future work





correlating histologic features with clinical manifestations and disease severity are ongoing.

