

AUTOIMMUNE CONNECTIVE TISSUE DISEASES

DETERMINING DISEASE COURSE IN MORPHEA: A PROSPECTIVE COHORT STUDY

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Introduction: Morphea (localized scleroderma) results in skin sclerosis, producing devastating impairment in those affected. Understanding the course of morphea is necessary to determine how to counsel patients and to plan outcome and interventional studies.

Objective: To determine the disease course of morphea using validated clinical outcome measures.

Materials and Methods: Prospective cohort study of 130 participants from the Morphea in Adults and Children (MAC) cohort with at least two years of follow-up and modified localized scleroderma skin severity scores (mLoSSI) scores recorded. Disease activity was defined as mLoSSI score greater than 0. Time to recurrence of disease activity from the first visit with inactive disease was compared between the linear and generalized subtype using survival analysis with the logrank test. All statistical analysis was performed using GraphPad Prism 6.0.

Results: 130 participants (663 study visits) were included. Fifty had at least 5 years of follow-up. Mean total follow-up was 4.3±1.7 years. The majority of participants were Caucasian (72%), female (77%), and had linear (55%) or generalized (31%) subtype. Median baseline mLoSSI score was 5. The mean time to first recurrence of disease activity after initial resolution of activity was 1.1 years for generalized morphea and 2.3 years for linear morphea. Overall, 43% of those with generalized morphea had a recurrence of disease activity compared to 21% of those with the linear subtype (Hazard ratio 2.79, 95% CI 1.48-7.94). Of the 50 participants followed for at least five years, 36% had a recurrence of disease activity.

Conclusions: Disease activity improves in the majority of morphea patients over time. Some patients have a monophasic disease course in which activity resolves. However, a substantial number of patients, particularly those with generalized morphea, have a relapsing remitting course over many years. Patients with morphea warrant monitoring for disease flares for extended periods of time.





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