RESEARCH LETTER

An Evaluation of Long-term Outcomes in Adults With Pediatric-Onset Morphea

The prevalence of morphea in childhood is not well established, but reports suggest that approximately two-thirds of linear morphea cases occur before age 18 years, and 20% to 30% of morphea cases overall begin in childhood.1,2 Studies have described substantial morbidity in children with morphea, but to our knowledge none has addressed the disease effects when the patients reach adulthood (age ≥18 years). To assess the impact of childhood morphea in adults, we searched the Morphea Registry and DNA Repository at the University of Texas Southwestern Medical Center (UTSW) for adults with pediatric-onset morphea (APOMs), and we report our findings herein.

Methods. Patients were enrolled between September 2007 and November 2009, and all were examined by the same physician (H.T.J.). Disease-related quality of life (QOL) was measured with the Dermatology Life Quality Index (DLQI).3 Symptoms of itch, pain, and numbness were assessed with visual analog scale (VAS). Data on disease progression, activity, and inactivity were collected. We confirmed the presence of concomitant disorders, symptoms, and permanent sequelae by direct physical examination of the patients or by extraction from medical records. Each patient was assigned a deep modifier, if present.

We determined antinuclear antibody (ANA) titers in collaboration with the laboratory of Frank Arnett, MD, using immunofluorescence on Hep-2 cells. Cases with positive ANA findings (titer ≥1:80) were tested for antibodies to extractable nuclear antigens, topoisomerase, and RNA polymerase.

Results. A total of 27 of 200 patients met enrollment criteria. Mean age at the time of study enrollment was 30.6 years (range, 18-78 years; median age, 26 years), and mean age of morphea onset was 11.5 years (age range, 3-17 years; median age, 13 years). Twenty of 27 patients had linear morphea (74%) (including 7 with en coup de sabre and 2 with partial facial hemiatrophy). Five patients had generalized morphea (19%), and 2 had plaque type (7%). Seven of 27 patients had at least 1 autoimmune disease (26%), and 13 of 27 had a family history of autoimmune disease. Overall, 78% of patients had noncutaneous symptoms (21 of 27); 67% had musculoskeletal complaints (18 of 27); 44% had neurologic manifestations (12 of 27); and 25% had Raynaud-like phenomena (7 of 27). On physical examination, it was noted that 56% of patients had permanent sequelae (15 of 27) (limited range of motion, 11 of 15; deep atrophy, 6 of 15; limb-length discrepancy, 1 of 13; and joint contracture, 2 of 15). All permanent morbidities occurred in linear morphea cases (15 of 20). Eighty-one percent of patients (21 of 27) had at least 1 symptom related to morphea (pain, itch, or numbness).

The mean DLQI score was 3.5 (median, 2; range, 0-12). Six of 27 scores (22%) indicated moderate to very large effect on QOL. Patients with moderate to very large QOL effect had a greater mean number of lesions than those with low to no QOL effect (8 vs 5), although the difference was not significant (P = .07). Limited range of motion was significantly associated with a lower disease-related QOL (r = .39; P = .05). Ten of 27 patients had positive ANA findings (37%), predominantly in a speckled pattern. Antibodies associated with the speckled-pattern ANA (nonchromatolin protein antigens) were not present.

All 27 patients received treatment, 13 with topical corticosteroids. Methotrexate was used in 8 of 20 cases of linear morphea; systemic corticosteroids in 6 of 20 linear cases and 1 generalized case; and physical therapy in 3 of 20 linear cases.

Eighty-nine percent of patients developed new or expanded lesions over time (24 of 27). Seven reported an inciting event (trauma at the site of the lesion, cessation or taper of systemic therapy, and pregnancy) (29%). Time to recurrence from initial disease onset ranged from 6 to 18 years. Sixteen patients described periods of remission and exacerbation, while 8 described continuous activity of existing lesions or formation of new lesions. Overall, with increasing age, lesions increased number and/or size (Figure 1).

Comment. This study represents the first examination of APOMs from the UTSW Morphea Registry and DNA Repository. The predominant morphea subtype in our group was linear, reflecting the predominance of this subtype in children.4,5 Patients maintained this subtype into adulthood but developed more lesions (Figure 2). Patients with generalized morphea had plaque subtype at disease onset that progressed to meet criteria for generalized disease over time. This underscores the progressive nature of morphea.

Figure 1. Morphea activity in 18 of 27 patients over time. Each colored line represents 1 patient; colors and data point shapes hold no significance but to visually distinguish the patient lines from each other. Disease activity was measured by number of episodes of new lesion development.

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Adults with pediatric-onset morphea have an autoimmune phenotype. Studies have demonstrated that patients with adult morphea have increased prevalence of autoimmunity compared with the general population. Children with morphea have increased prevalence of familial autoimmunity but relatively less risk of personal autoimmunity disease. An unanswered question was whether children with morphea have increased autoimmunity as adults. Our study suggests that APOMs develop autoimmune disorders in adulthood. Overall, 7 of 27 APOMs had autoimmune disorders (26%), 5 of them having developed these disorders as adults. This is similar to rates reported for adults with morphea and higher than childhood rates.

Adults with pediatric-onset morphea have lowered QOL, especially if they also have functional impairment \((P = .05)\) and/or a large number of lesions. Furthermore, permanent functional impairment, persistent disease activity, and extracutaneous symptoms were all present in most of our patients. These data underscore the progressive, disabling course of pediatric-onset morphea in adults.

The predominance of topical therapy in this group suggest that the condition is being undertreated, which possibly affects long-term outcome. Even in those APOMs treated with immunosuppressive agents, 2 of 7 cases flared during treatment tapering or after treatment cessation. Systemic treatment may inhibit disease activity but does not preclude subsequent disease reactivation.

In contrast to the results of prior studies, we found that 89% of our cohort had continued disease activity (24 of 27 patients). To mitigate referral bias, prospective cohort studies are needed to determine accurate prevalence. Nonetheless, our data indicate the presence of a subset of patients with active disease in adulthood who need lifelong evaluation and repeated courses of aggressive treatment to prevent the substantial morbidity noted in our group. Parents of children with morphea should be counseled on the possibility of recurrence and on the need for vigilance in identifying and seeking treatment for new activity.

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**VIGNETTES**

**Atypical Fibroxanthoma in an African American Woman**

Report of a Case. A 70-year-old African American woman was seen in our clinic with a 3- to 4-month history of a...