SKIN

BRIEF ARTICLE

Improvement of Suspected Linear Morphea Seen with Topical Ruxolitinib

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ABSTRACT

Linear morphea is an autoimmune skin condition characterized by localized sclerosis and discoloration. Current standard of care includes topical and intralesional steroids, phototherapy, and immunosuppressive agents; however, no gold-standard treatment exists. We present a 34-year-old female who presented with a hypopigmented and indurated plaque in an en coup de sabre configuration on her forehead, suspicious of linear morphea, that improved significantly in thickness and hypopigmentation after eight weeks of 1.5% ruxolitinib cream.

INTRODUCTION

Morphea is a rare autoimmune skin condition associated with localized sclerosis and discoloration.[1,2] There are many clinical presentations of morphea (linear, circumscribed, generalized, and mixed) that can affect individuals physically (often seen with other comorbidities) and mentally.[2–4]

Moreover, the management of linear morphea en coup de sabre, a subtype of linear morphea present on the forehead, has been challenging due to limited evidence and no gold standard of treatment.[1,5] Current treatments focus on inhibiting active progression of disease and improving induration. Treatment options such as topical and intralesional steroids, phototherapy, and immunosuppressive agents (methotrexate, hydroxychloroquine, cyclosporine), and surgical reconstructive treatment have shown variable efficacy and risk of potential side effects.[1,5]

JAK inhibitors have been shown to inhibit TGF-beta signaling, a crucial cytokine for the pathogenesis of dermal fibrosis.[6,7] We hypothesize that ruxolitinib cream, a JAK1/2 inhibitor recently approved for eczema and non-segmental vitiligo, may be used to treat linear morphea with less potential for side effects compared to other currently available treatment options.[1,8]

CASE REPORT

A 34-year-old female presented with a 2-month linear plaque on her forehead. She reported that the lesion had progressively worsened over time despite treatment with calcipotriene ointment and petroleum jelly. Physical examination revealed a solitary hypopigmented, indurated, pruritic plaque measuring 5.4 cm extending from her hairline

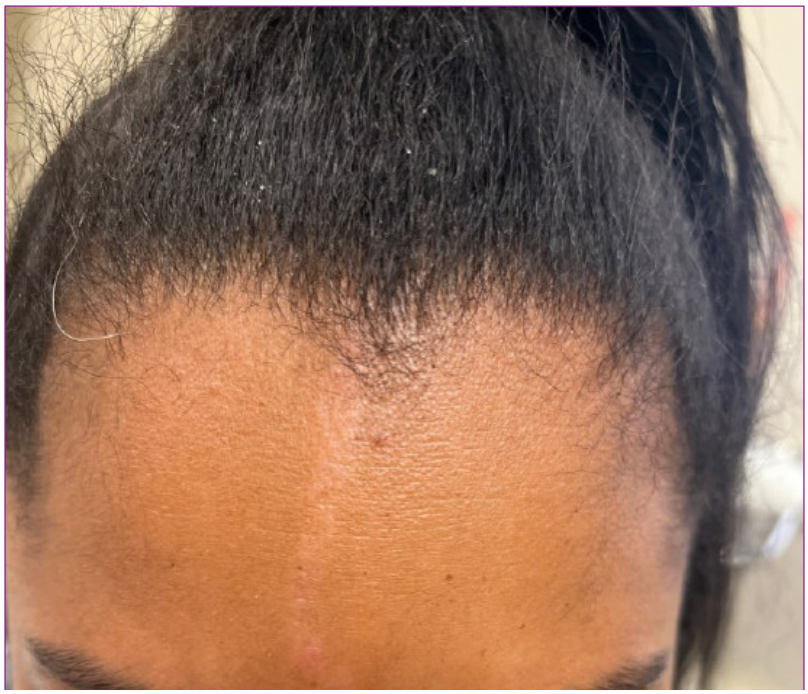

Figure 1. Frontal view taken before treatment

downward in an en coup de sabre configuration (**Figure 1**). There were no associated neuromuscular and ocular symptoms. Concomitantly, the patient also had pruritic, erythematous patches on her chin that failed to improve with moisturizers, resembling atopic dermatitis. A skin biopsy was not taken as the lesion was located in a cosmetically sensitive area, however, her presentation was suspicious for linear morphea. 1.5% ruxolitinib cream was prescribed for the lesions on her chin, and after discussing several options with accompanying risks and benefits for her forehead, the patient opted to start a trial of 1.5% ruxolitinib cream twice a day. After 8 weeks of application, significant improvement in induration and hypopigmentation was noted (**Figure 2**). Pruritus on the forehead and chin persisted. However, no systemic or cutaneous side effects from ruxolitinib were observed.

DISCUSSION

In this case, we show a 34-year-old female with suspected linear morphea with significant improvement in skin thickness and hypopigmentation after 8 weeks of treatment with 1.5% ruxolitinib cream with no accompanying side effects.

This improvement may be due to the decreased activation of two pathways mediated by JAK1/2/STAT inhibition: TGF-beta and IFN-gamma signaling.[1] TGF-beta activates the JAK/STAT pathway, and previous in-vitro and in-vivo studies have shown that inhibition of JAK leads to reduced collagen deposition and dermal thickness.[1] Also, morphea in its early stages is highly inflammatory with increased production of cytokines (IFN-gamma, IFN-alpha, TNF-a), and this inflammatory response may lead to the development of discoloration or depigmentation.[9] Ruxolitinib has been shown

Figure 2. Frontal view (A) and side view (B) taken after 8 weeks of 1.5% ruxolitinib cream twice a day

to decrease IFN-gamma signaling and the production of cytotoxic immune cells.[8]

Based on this case, ruxolitinib may be a potential treatment for linear morphea. We recognize that a skin biopsy was not performed to confirm the diagnosis but the rash exhibited the prototypic en coup de sabre morphea presentation without any known traumatic insult to the skin.[2] Other diseases and skin conditions were considered, but the lack of symptoms other than pruritus made them less likely.

This patient responded rapidly to ruxolitinib cream despite linear morphea typically affecting the deeper dermis; however, the timing of improvement may vary.[10] Further research is needed to establish ruxolitinib's efficacy as a possible treatment option for morphea.

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