SKIN

BRIEF ARTICLE

Steatocystoma Multiplex Suppurativa Treated Successfully with Adalimumab and Topical Tofacitinib

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ABSTRACT

Steatocystoma multiplex (SM) is a rare chronic dermatologic condition which may cause significant distress and negatively impact quality of life. Currently, there is no consensus for treatment. When inflammatory, it is called steatocystoma multiplex suppurativa, an even more rare condition that has shown reported success with anti-TNF-alpha therapy. We describe a patient case wherein adalimumab prescribed for autoimmune arthritis resulted in SMS remission, further supported with evidence of sustained remission with topical tofacitinib. This patient case supports other evidence showing adalimumab as a treatment option for SMS and highlights the potential of tofacitinib as a novel treatment.

INTRODUCTION

SM appears as multiple, usually asymptomatic, flesh or yellow colored cystic nodules filled with sebum, typically where the pilosebaceous apparatus is highly developed. Onset is typically during early adolescence.[1] Diagnosis of SM is often made by history and physical alone. An inflammatory variant known as SMS can present with great risk for infection and scarring. Goals of treatment include reduction of size and pain, prevention of recurrence, and improved cosmetic outcome. No FDA approved treatment exists, but success has been documented with carbon dioxide laser, surgical interventions, cryotherapy, and medical management. Successful medical management has included isotretinoin, oral tetracycline, topical clindamycin, or benzoyl peroxide wash.[2]

SM has been reported concomitantly with hidradenitis suppurativa (HS) in some patients, which some suggest could be due to a defect in follicular proliferation or shared genetic link.[3,4] The pathophysiology of HS is not well understood, with only some patients responding well to anti-TNF-alpha therapies.[5] Due to a proposed shared link between SM and HS, anti-TNF-alpha therapy has been considered for SMS management in some patients with comorbidities. Currently, few case reports exist of treatment of SMS with anti-TNF-alpha therapies. SM associated with HS was successfully treated with adalimumab in three cases, including both inflammatory and non-inflammatory skin lesions. A theoretical mechanism is that the compromised pilosebaceous structures benefit from the medication's pro-apoptotic and anti-lipogenic effect.[4] No cases have been reported regarding the use of tofacitinib in SMS to date.

CASE PRESENTATION

A 67 year old male with personal history of rheumatoid arthritis (RA) presented with a past medical history of SMS, originating during adolescence as a solitary inflamed cystic lesion on his chest. He had a family history of HS. At age 27, he was diagnosed with SM, consisting of many full flesh colored nodules over the back, arms and chest **(Figure 1)**. Over years, he developed severe SMS. Prior to our assumption of care, the patient reported multiple infected lesions, two complicated by sepsis, and six hospitalizations within a single year related to SMS. Acute minor infections had been treated with antibiotics (doxycycline or cephalexin), coupled with an anti-inflammatory dosage of dapsone 175 mg daily. He completed two full courses of isotretinoin.

On presentation, the patient began treatment with dapsone 150 mg once daily. Zinc 220 mg daily added anti-neutrophilic effect while bleach baths were recommended for anti-microbial benefit. The first 5 years of treatment included dapsone gel twice daily to active lesions, ammonium bituminosulfonate paste, mupirocin ointment, topical desoximetasone, and retapamulin 1% topical antibiotic, with topical chlorhexidine antiseptic and a sulfacetamide shampoo to cleanse the skin daily. Active lesions were treated in office through incision and drainage, as well as injections of intralesional Kenalog (ILK). Systemic treatments included hydroxychloroquine 400 mg daily, doxycycline 100 mg twice daily, clindamycin 100 mg daily, and multiple prednisone courses.

After lack of durable efficacy from aforementioned treatments and continued frequent painful flaring, adalimumab therapy was considered. Counseling with rheumatology solidified adalimumab as an option to mitigate progression of the patient's RA, with hope of simultaneous remission of SMS. The patient thus began 40 mg biweekly for the next two years.

Following adalimumab initiation, the patient had multiple periods of 3-6 months with few or no SM flares, a significant improvement. This included his longest ever remission period since the condition first manifested. Despite being eventually discontinued after two years due to the complication of new frequent herpes simplex virus (HSV) flaring, the patient reported the lowest SM discomfort level to date with adalimumab, further highlighting the importance of decreasing systemic inflammatory response to increase quality of life. The patient had a focal flare on his right forearm shortly after this period (treated with ILK/cephalexin).

Although adalimumab had been relatively successful at subduing the patient's SM for two years, he ultimately manifested modestly increased flare frequency. In addition, the treatment likely exacerbated his proclivity for HSV flares. During a four-month period, he experienced 1-2 flares per month, whereas previously he would have a few per year. In the past, his HSV was treated with abortive valacyclovir. This was altered to maintenance therapy with valacyclovir of 500 mg daily, with abortive therapy during acute flares.

Due to decreased response to adalimumab and increased HSV flare frequency, the patient requested all systemic immunosuppressive therapies be discontinued to observation. In place of systemic therapy, compounded topical tofacitinib 2%/niacinamide 2% cream, a topical Janus kinase (JAK)-inhibitor, was initiated given that topical JAK inhibitors have been useful in many inflammatory conditions.

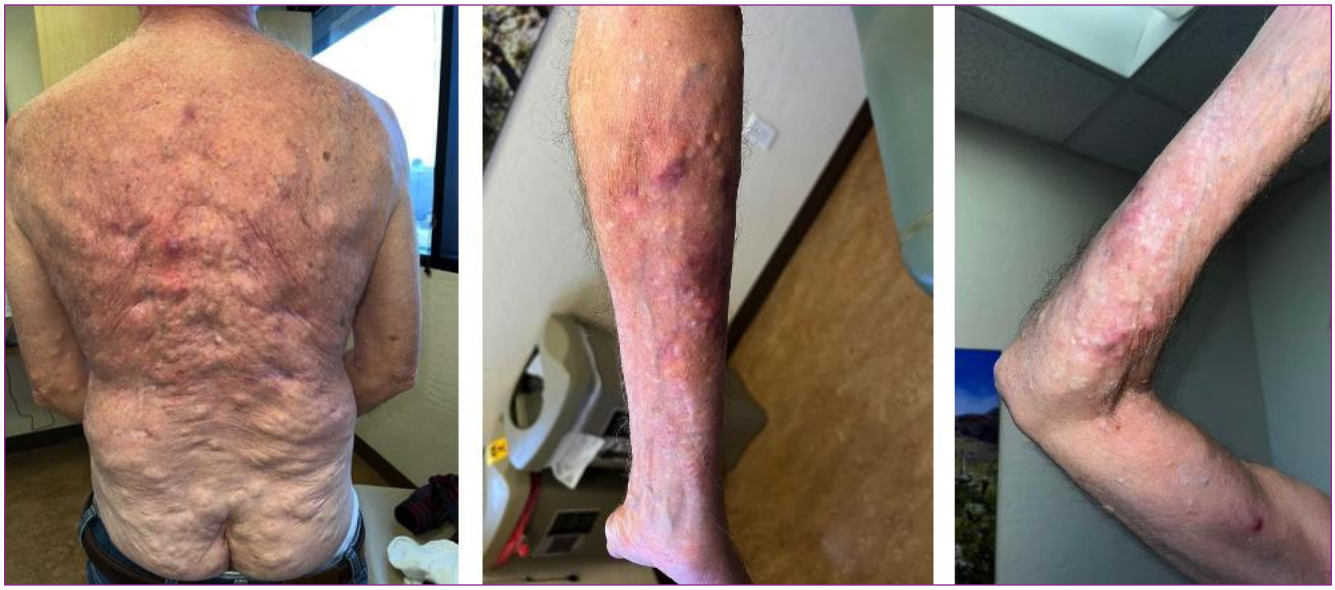

Figure 1. Steatocystoma Multiplex Suppurativa after adalimumab treatment. Multiple subcutaneous cystic nodules are visible as well as scars from previous surgical interventions.

Following a few months of use of topical tofacitinib alone, the patient reported significant reduction in inflamed SMS lesions. At his most recent clinic visit, the patient desired continuing only topical tofacitinib twice daily as needed, but future plans entail considering another course of isotretinoin and dapsone dual therapy.

Topical JAK inhibitors have become increasingly relevant in the treatment of various inflammatory dermatological conditions. These directly target the JAK signaling pathway, which plays a crucial role in inflammation and immune responses. These medications can effectively alleviate symptoms associated with inflammatory skin conditions such as psoriasis, atopic dermatitis, vitiligo, and alopecia areata[6]. Clinical trials are currently underway for the use of both oral and topical JAK inhibitors in inflammatory conditions such as HS. At this time, there are no reported cases of JAK inhibitors being used to treat SMS.

DISCUSSION

This case demonstrates that SMS may be refractory to treatment and cause serious infection and even sepsis. Remission with adalimumab, followed by topical tofacitinib upon loss of remission, led to the longest disease-free period this patient ever experienced in over forty years. Treatment with topical tofacitinib demonstrated an unprecedented and durable response even when used alone, further emphasizing the need for additional research in the treatment of SMS.

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