Successful treatment of psoriasis with risankizumab in a patient with telangiectasia macularis eruptiva perstans

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INTRODUCTION

Telangiectasia macularis eruptiva perstans (TMEP) is a rare cutaneous form of mastocytosis that is most commonly found in adults.[1] Symptoms are caused by mast cell degranulation and can include rash, itching, flushing, diarrhea, and dyspnea. TMEP is usually confined to the skin but may have systemic involvement of bone marrow, gastrointestinal tract, liver, and spleen. The treatment of TMEP depends on the presence or absence of systemic involvement and organ dysfunction. TMEP does not have a Food and Drug Administration-approved medication; currently available treatment options are guided toward symptom management and rash reduction. Identification and avoidance of patient individual triggers such as alcohol, temperature and drugs can be useful in the management of the disease.[1] Also, H1 antagonists, topical and oral corticosteroids, psoralen and longwave ultraviolet radiation, for rash and associated pruritus have all been reported in the literature with some effectiveness help treat TMEP.[1] Iatrogenic immunosuppression with cyclosporin in renal transplant patients and tumor necrosis factor-α inhibitors in chronic immune-mediated disorders have been associated with an increased risk of lymphoproliferative disorders and hematologic malignancies.[2,3] Therefore, traditional biologic therapy is generally not recommended in patients with active TMEP disease. However, biologics used in dermatology have become increasingly more targeted with potentially less immunosuppression and side effects. In fact, IL-17 and 23 antagonists have shown superior safety profiles in numerous studies without an appreciable malignancy risk beyond that of background rate.[4] Little is known about how biologic therapy can alter the natural course of TMEP in the context of psoriasis treatment. Herein, we report a case of cutaneous TMEP with moderate-to-severe plaque psoriasis that was successfully treated with risankizumab without an exacerbation of symptoms or blood parameters related to his TMEP.

Abbreviation used:

TMEP: elangiectasia macularis eruptiva perstans

CASE REPORT

A 58-year-old man with uncontrolled moderate-to-severe plaque psoriasis and hypertension presented to our dermatology clinic for the treatment of psoriasis present for 20 years. The patient had predominantly guttate- and plaque-type psoriasis, involving most of his trunk and upper extremities, with approximately 6% body surface area involvement and an investigator global assessment score of 3, without evidence of psoriatic arthritis. Of note, patient was a former smoker, consumed 1-2 alcoholic beverages daily and had a history of numerous nonmleanoma skin cancers. Years after the diagnosis of psoriasis, he was noted to have numerous, asymptomatic, scattered, red-to-brown papules and macules on his back and abdomen (Fig 1, *A* and *B*). The patient reported flushing and itching with alcohol consumption, along with intermittent diarrhea, but was otherwise

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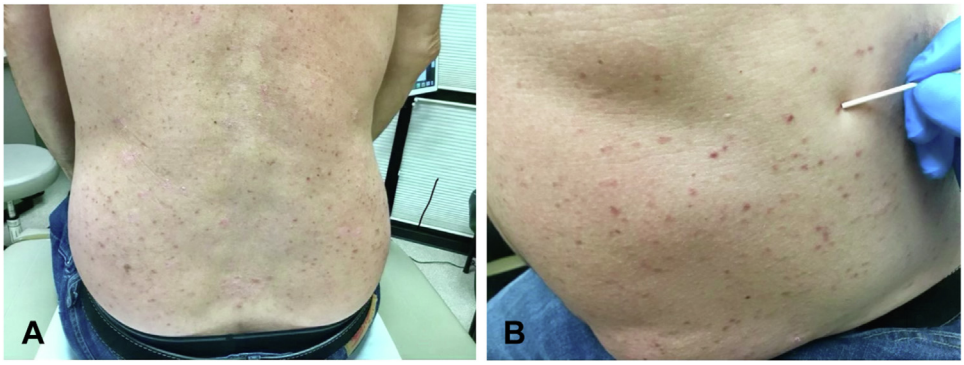

Fig 1. Photos of the patient from the time of original diagnosis of telangiectasia macularis eruptiva perstans. The characteristic ruddy brown telangiectatic macules were appreciated on (**A**) his back and (**B**) abdomen.

asymptomatic. A shave biopsy was performed, which showed histopathologic changes consistent with TMEP. He was referred to hematology-oncology where bone-marrow biopsy was performed, which did not reveal any evidence of systemic mastocytosis. Laboratory results revealed a normal complete blood cell count, comprehensive metabolic panel, and a tryptase level of 9.4 μg/L (reference range, 2.2-13.2 μg/L).

The patient had previously failed topical steroids and was a poor candidate for light therapy given his extensive history of non-melanoma skin cancers, and methotrexate because of his drinking history. The decision to start risankizumab, a humanized immunoglobulin G 1 monoclonal antibody that targets the interleukin 23A cytokine, was made because of its highly selective nature, risk and side effect profile, and favorable dosing schedule (lower injection frequency per patient preference) with other biologic options. At his 4-week follow-up, the patient was completely clear of his psoriasis. The patient is now 13 months into treatment with risankizumab and continues to be completely clear of psoriasis. Furthermore, he remains asymptomatic, and his follow-up tryptase level has remained stable, 9.8 μg/L (reference range, 2.2-13.2 μg/L), with no appreciable change in TMEP lesions.

DISCUSSION

TMEP is a rare form of cutaneous mastocytosis that can cutaneous mastocytosis that can not infrequently be associated with an underlying systemic mastocytosis. In a recent retrospective study of 243 patients with cutaneous mastocytosis in a tertiary care center, TMEP was observed in 14% of all cases of cutaneous mastocytosis, with 47% of those cases associated with systemic involvement.[5] The total tryptase level may be the most relevant laboratory value to measure when checking for systemic involvement in the cutaneous forms.[1,5] Tryptase levels have shown correlation with positive bone-marrow biopsies, with 100% being positive, when the total tryptase level was >75 ng/mL and about 50% being positive, when the total tryptase level was 20-75 ng/mL.[1] Annual monitoring of complete blood count, comprehensive metabolic panel, serum tryptase levels, possibly, 24-hour urinary histamine levels are recommended to track progression and or transformation to malignant hematologic disease.[3] Our patient posed a clinical conundrum, as the literature regarding safety and outcomes of biologic therapy in patients with psoriasis and rare forms of mastocytosis is limited. Given the rarity of co-existent psoriasis and TMEP, most of our experience will most likely come from registry studies and case reports. We hope that our case adds to the current dearth of literature and will provide some guidance for future cases.

Conflicts of interest

Cara Barber has no conflicts of interest to report. Dr. Eingun James Song, MD, has been a consultant, speaker or investigator for the following companies: AbbVie, Janssen, Amgen, Lilly, SUN, UCB, Incyte, Novartis, Sanofi & Regeneron, Castle Biosciences, and Pfizer.

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