

characterized by lesions that are morphologically distinct or appear in other locations. These atypical forms include vesicular, purpuric, inverse, unilateral, and palmoplantar PR. Palmoplantar involvement in PR is very rare, and very few cases are described in the literature. In some such cases the palms and soles are affected in the context of a more typical eruption on the trunk.<sup>2,3</sup> Others consist of palmoplantar involvement in the form of vesicular lesions,<sup>4</sup> or of more typical, exclusively palmoplantar lesions.<sup>5</sup> We consider our case to correspond to the latter group, diagnosis of which can be difficult to establish. In all cases of PR with palmoplantar involvement the main differential diagnosis is secondary syphilis. For this reason, serological and histological approaches were used to rule out secondary syphilis in our patient and help establish diagnosis. Histology of PR is nonspecific. In our patient biopsy revealed findings that could be considered compatible with an eczematous process. However, given the clinical appearance of the lesions, the absence of pruritus, and the resolution without treatment, this entity was excluded from the differential diagnosis.

Treatment of PR is controversial. Some data support treatment with erythromycin.<sup>6</sup> However, given the natural course of the disease alternative options include symptomatic treatment of pruritus with topical corticosteroids or oral antihistamines and therapeutic abstention, which was selected in the present case.

We present a case compatible with palmoplantar PR, a rare variant of PR of which very few cases are described in the literature. Despite their infrequent nature, atypical variants of PR can simulate other conditions, and therefore knowledge of these entities is of the utmost importance.

## References

1. Chuang TY, Ilstrup DM, Perry HO, Kurland LT. Pityriasis rosea in Rochester, Minnesota, 1969 to 1978. *J Am Acad Dermatol.* 1982;7:80–9.
2. Deng Y, Li H, Chen X. Palmoplantar pityriasis rosea: two case reports. *J Eur Acad Dermatol Venereol.* 2007;21:406–7.
3. Bukhari I. Pityriasis rosea with palmoplantar plaque lesions. *Dermatol Online J.* 2005;11:27.
4. Singh V, Sharma M, Narang T, Madan M. Vesicular palmoplantar pityriasis rosea. *Skinmed.* 2012;10:116–8.
5. Zavar V. Acral pityriasis rosea in an infant with palmoplantar lesions: a novel manifestation. *Indian Dermatol Online J.* 2010;1:21–3.
6. Sharma PK, Yadav TP, Gautam RK, Taneja N, Satyanarayana L. Erythromycin in pityriasis rosea: a double-blind, placebo-controlled clinical trial. *J Am Acad Dermatol.* 2000;42:241–4.

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## Good Response to Tofacitinib in Refractory Amyopathic Dermatomyositis<sup>☆</sup>



### Dermatomiositis amiopática refractaria con buena respuesta a tofacitinib

To the Editor:

Dermatomyositis (DM) is an idiopathic inflammatory myopathy characterized by muscle inflammation, skin involvement, and systemic manifestations that are often a very broad spectrum in nature.<sup>1</sup> Amyopathic DM, also known as DM sine myositis, affects a subset of DM patients and presents with cutaneous symptoms but without myopathy.<sup>2</sup>

Our patient was a 49-year-old white woman with a history of thalassemia, for which she had been in follow-up for 7 years. She presented with asthenia, itching, and skin lesions located predominantly in photo-exposed areas. Physical examination revealed heliotrope erythema, Gottron papules, Gottron sign, shawl sign, V sign, and Holster

sign. A detailed medical history was taken and a systemic review performed, as well as multiple additional tests, including nuclear magnetic resonance imaging of the pelvic girdle, an electromyogram, a muscle biopsy, and determination of muscle enzymes. All results were normal, and the patient was diagnosed with amyopathic DM. Screening for antinuclear, anti-synthetase, anti-TIF-1 $\gamma$  (transcription intermediary factor 1  $\gamma$ ), and anti-MDA-5 (melanoma differentiation-associated protein 5) antibodies and occult tumors was negative throughout the follow-up period.

Owing to poor disease control, the patient had undergone multiple treatments to which she responded poorly, including topical and oral corticosteroids (maximum dose, 1 mg/kg/d), azathioprine, methotrexate, hydroxychloroquine, mepacrine, rituximab, oral tacrolimus, intravenous immunoglobulins, and mycophenolate mofetil. She was being simultaneously treated with oral prednisone, hydroxychloroquine, mepacrine, and mycophenolate mofetil up until 7 months before the consultation. Given the progressive worsening of her condition and the severity of the skin lesions (Fig. 1) and pruritus, only treatment with low-dose prednisone (2.5 mg/d) was maintained, and tofacitinib (TOF) treatment (5 mg/12 h) was started.

Clinical improvement was observed 2 weeks later and, at the time of writing, all skin lesions have improved significantly (Fig. 2) and the pruritus has disappeared.

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**Figure 1** Skin lesions prior to beginning tofacitinib therapy. A, Gottron papules, periungual erythema, and hemorrhagic crusting on the hands. B, Facial erythema and scaling and heliotrope erythema of the eyelids. C, V sign on the chest.



**Figure 2** Skin lesions after 7 months of tofacitinib therapy. A, Flattening of Gottron papules and disappearance of periungual erythema. B, Discrete heliotrope erythema. C, Poikiloderma on the chest with no evidence of V sign.

DM is a difficult-to-manage multisystemic disease for which no standardized treatment algorithms have been developed owing to the scarcity of well-designed clinical trials. Furthermore, it is not uncommon for DM to overlap with other autoimmune diseases, posing both diagnostic and therapeutic challenges to physicians.<sup>3</sup>

TOF is an inhibitor of the Janus kinases JAK3 and JAK1 and attenuates signal transduction activated by interleukin (IL) -2, IL-4, IL-6, IL-7, IL-9, IL-15, IL-21, and type I and II interferons. Because the interferon signaling pathway is overexpressed in DM, TOF may be useful in the treatment of this disease.<sup>3,4</sup>

Kurtzman et al<sup>5</sup> reported promising results in 3 patients with recalcitrant cutaneous DM who were treated with TOF. Subsequently, Paik and Christopher-Stine<sup>4</sup> reported a good response to TOF treatment in an adult DM patient who had failed to respond to multiple therapies. In a review by Cinats et al<sup>6</sup> of emerging uses of JAK inhibitors in skin diseases, including psoriasis, atopic dermatitis, alopecia areata, vitiligo, DM, and graft versus host disease, adverse effects reported included infections in TOF-treated patients and anemia and thrombocytopenia in those treated with ruxolitinib.<sup>6</sup>

In the last year, clinical cases have been published demonstrating a therapeutic effect of TOF not only in skin manifestations, but also interstitial lung disease and severe refractory calcinosis, with no notable side effects<sup>1,3,7-9</sup> except for transient hypercalcemia and increased body mass index in 2 patients.<sup>1</sup>

The present case is the sixth published description of amyopathic DM treated with TOF. As in the 5 previously published cases,<sup>3,5,7</sup> the patient improved rapidly and treatment was well tolerated.

Clinical trials are required to reliably confirm the efficacy and safety profile of this treatment. A clinical trial is currently underway to evaluate the efficacy of TOF treatment of treatment-refractory DM.<sup>10</sup>

## Conflicts of Interest

The authors declare that they have no conflicts of interest.

## References

1. Wendel S, Venhoff N, Frye BC, May AM, Agarwal P, Rizzi M, et al. Successful treatment of extensive calcifications and acute pulmonary involvement in dermatomyositis with the Janus-Kinase inhibitor tofacitinib - A report of two cases. *J Autoimmun.* 2019;100:131-6.
2. Concha JSS, Tarazi M, Kushner CJ, Gaffney RG, Werth VP. The diagnosis and classification of amyopathic dermatomyositis: a historical review and assessment of existing criteria. *Br J Dermatol.* 2019;180:1001-8.
3. Moghadam-Kia S, Charlton D, Aggarwal R, Oddis CV. Management of refractory cutaneous dermatomyositis: potential

- role of Janus kinase inhibition with tofacitinib. *Rheumatology (Oxford).* 2019;58:1011-5.
4. Paik JJ, Christopher-Stine L. A case of refractory dermatomyositis responsive to tofacitinib. *Semin Arthritis Rheum.* 2017;46:e19.
  5. Kurtzman DJB, Wright NA, Lin J, Femia AN, Merola JF, Patel M, et al. Tofacitinib citrate for refractory cutaneous dermatomyositis: An alternative treatment. *JAMA Dermatol.* 2016;152:944-5.
  6. Cinats A, Heck E, Robertson L. Janus kinase inhibitors: a review of their emerging applications in dermatology. *Skin Therapy Lett.* 2018;23:5-9.
  7. Kurasawa K, Arai S, Namiki Y, Tanaka A, Takamura Y, Owada T, et al. Tofacitinib for refractory interstitial lung diseases in anti-melanoma differentiation-associated 5 gene antibody-positive dermatomyositis. *Rheumatology (Oxford).* 2018;57:2114-9.
  8. Hornig J, Weinhage T, Schmidt LH, Buerke B, Schneider U, Pavenstädt H, et al. Response of dermatomyositis with lung involvement to Janus kinase inhibitor treatment. *Z Rheumatol.* 2018;77:952-7.
  9. Kato M, Ikeda K, Kageyama T, Kasuya T, Kumagai T, Furuya H, et al. Successful treatment for refractory interstitial lung disease and pneumomediastinum with multidisciplinary therapy including tofacitinib in a patient with anti-MDA5 antibody-positive dermatomyositis. *J Clin Rheumatol.* 2019, <http://dx.doi.org/10.1097/RHU.0000000000000984>. Epub ahead of print.
  10. ClinicalTrials.gov. Study of tofacitinib in refractory dermatomyositis (STIR). Available in: <https://clinicaltrials.gov/ct2/show/NCT03002649?cond=dermatomyositis+tofacitinib&rank=1>. [accessed 23 May 2019].

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## Adult-Onset Eccrine Angiomatous Hamartoma: A Case Report With Ultrasound Findings<sup>☆</sup>



### Hamartoma angiomatoso ecrino en la edad adulta y sus hallazgos ecográficos

To the Editor:

Eccrine angiomatous hamartoma (EAH) is a rare, benign cutaneous tumor composed of vascular and eccrine ele-

ments. It is most commonly diagnosed during the first years of life, although there are reports of cases diagnosed in adults. We report the ultrasound findings of a 46-year-old patient with a left plantar lesion, histopathological diagnosis of which confirmed suspicion of EAH.

## Case Description

A 46-year-old woman was seen for mild discomfort in the sole of her left foot when walking. During the anamnesis, she reported a lesion in the midfoot, specifically the plantar support area, that had appeared approximately 10 years earlier. Clinical examination revealed a bluish, rubbery, compressible nodule of approximately 3 cm

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