Improvement of pterygium inversum unguis and Raynaud phenomenon with interdigital botulinum toxin injections

Daniel J. Kim, PhD, and Ian D. Odell, MD, PhD

Key words: botulinum; pterygium inversum unguis; raynaud phenomenon; scleroderma; systemic lupus erythematosus; systemic sclerosis; ventral pterygia.

INTRODUCTION

Pterygium inversum unguis (PIU), also termed ventral pterygium, is a rare condition characterized by extension and adherence of the hyponychium to the ventral surface of the nail plate, leading to obliteration of the distal nail groove. It most frequently arises in the setting of connective tissue diseases such as scleroderma (systemic sclerosis [SSc]) and systemic lupus erythematosus (SLE).[1] PIU, when symptomatic, can cause severe pain for patients. However, there are few treatments available for PIU. Here, we report a patient with SSc-SLE overlap whose PIU and refractory Raynaud phenomenon (RP) improved significantly after interdigital injections of botulinum toxin A (BTA).

Abbreviations used:

BTA: botulinum toxin A
PIU: pterygium inversum unguis
RP: Raynaud phenomenon
SLE: systemic lupus erythematosus
SSc: systemic sclerosis

CASE REPORT

In 2021, a 39-year-old man with SSc-SLE overlap and positive antibody titers (anti-nuclear antibody >1:1280 speckled pattern, RNP, Smith, and RNA polymerase III) presented to us with uncontrolled pain of the fingers associated with frequent episodes of RP. Management of his RP previously failed treatment with nifedipine, sildenafil, pentoxifylline, and topical nitroglycerin. On examination, he had mild skin fibrosis limited to thickening of the skin of his fingers on both hands, with a corresponding modified Rodnan skin score of 2.[2] He also had dactylitis, erythema of his nail folds, and ragged cuticles without dilated nail-fold capillaries. The ventral surfaces of his most painful digits showed overgrowth of the hyponychium and subungual thickening due to hyperkeratosis involving his left second and fifth fingertips and right third and fifth fingertips (Fig 1, *top*). Further diagnostic workup with fungal cultures of fingernail clippings was negative. Altogether, his examination findings supported a diagnosis of SSc-SLE-associated RP and PIU.

For management of his SSc-SLE-associated RP, we treated our patient with interdigital BTA 5 Units in 0.1-ml sterile saline per injection site at 10 and 2 o'clock at the base of the second-fifth fingers of both hands, similar to previously described.[3] Injection of the thumbs was not performed to avoid weakening thumb opposition strength. He tolerated the procedure well without hand weakness or pain from the procedure. Within 1 week, he reported significant improvement in pain of his fingers and quality of life by telephone interview. Improvement continued for 4 months, at which point he returned to clinic, and we observed improvement in his PIU and dactylitis as well (Fig 1, *bottom*). Injections were repeated at 6 months, and he has had continued successful control of his SSc-SLE-associated RP and PIU.

From the Department of Dermatology, Yale University School of Medicine, New Haven, Connecticut.

Funding sources: None.

IRB approval status: Not applicable.

Consent statement: Consent for the publication of all patient photographs and medical information was provided by the authors at the time of article submission to the journal stating that all patients gave consent for their photographs and medical information to be published in print and online and with the understanding that this information may be publicly available.

Correspondence to: Ian D. Odell, MD, PhD, Department of Dermatology, Yale School of Medicine, 330 Cedar St, LMP-5040, New Haven, CT 06520. E-mail: ian.odell@yale.edu.

JAAD Case Reports 2022;26:79-81.

2352-5126

https://doi.org/10.1016/j.jdcr.2022.06.009

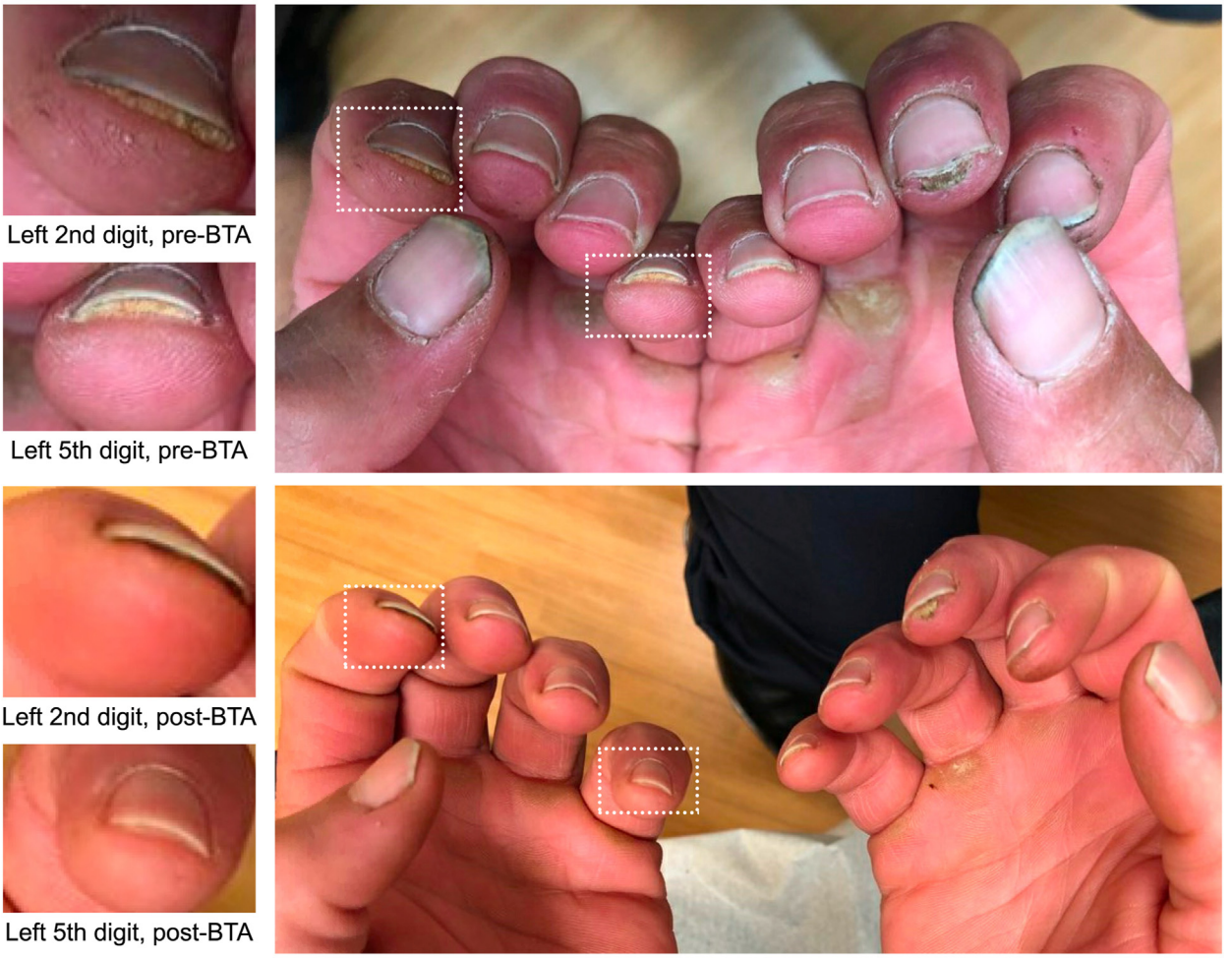

Fig 1. Photos of fingertips before (*top*) and 4 months after (*bottom*) treatment with interdigital BTA. Magnifications of left second and fifth digits (*white boxes*) are shown on the left. *BTA*, Botulinum toxin A.

DISCUSSION

Treatment of SSc-associated RP with BTA is a promising treatment approach that has shown improvement in other important aspects of hand function, including skin ulceration, pain, and pinch and power grip.[3,4] SSc-associated PIU, in contrast, has few treatment options, with previous reports using nail lacquer containing hydroxypropyl chitosan[5] and electrodessication.[6] Through treatment of our patient's SSc-SLE-associated RP with BTA, we discovered concurrent improvement in his PIU. Pathogenesis of PIU has been postulated to be from reactive hyperkeratosis secondary to impaired vascular perfusion,[7] which in our patient would be due to his frequent RP attacks. By successfully controlling his RP, his reactive hyperkeratosis improved as well. In sum, we present a case of refractory SSc-SLE-associated RP treated with BTA which showed improved PIU along with pain symptoms and quality of life.

Conflicts of interest

None disclosed.

REFERENCES

1. Caputo R, Cappio F, Rigoni C, et al. Pterygium inversum unguis. Report of 19 cases and review of the literature. *Arch Dermatol*. 1993;129:1307-1309.
2. Khanna D, Furst DE, Clements PJ, et al. Standardization of the modified Rodnan skin score for use in clinical trials of systemic sclerosis. *J Scleroderma Relat Disord*. 2017;2:11-18.
3. Dhaliwal K, Griffin MF, Salinas S, Howell K, Denton CP, Butler PEM. Optimisation of botulinum toxin type a treatment for the management of Raynaud's phenomenon using a dorsal approach: a prospective case series. *Clin Rheumatol*. 2019;38:3669-3676.
4. Uppal L, Dhaliwal K, Butler PE. A prospective study of the use of botulinum toxin injections in the treatment of Raynaud's syndrome associated with scleroderma. *J Hand Surg Eur*. 2014;39:876-880.
5. Marinho Falcao Gondim R, Bezerra da Trindade Neto P, Baran R. Pterygium inversum unguis: report of an extensive case with good therapeutic response to hydroxypropyl chitosan and review of the literature. *J Drugs Dermatol*. 2013;12:344-346.

6. Kang JH, Kim M, Cho BK, Park HJ. Pterygium inversum unguis: treated by electrodessication. *J Am Acad Dermatol*. 2015;72: AB116.

7. Vadmal M, Reyter I, Oshtory S, Hensley B, Woodley DT. Pterygium inversum unguis associated with stroke. *J Am Acad Dermatol*. 2005;53:501-503.