

Single Case

Nail Lichen Planus in a Patient with Cogan Syndrome: Report of a Case and Discussion

Carine Houriet Eckart Haneke

Department of Dermatology, Inselspital, Bern University Hospital, University of Bern, Bern, Switzerland

Keywords

Nail lichen planus · Cogan syndrome

Abstract

Cogan syndrome and lichen planus represent two autoimmune disorders. Cogan syndrome is a very rare type of ANCA-negative vasculitis affecting the eyes and vestibulocochlear system. It has been associated with other autoimmune disorders, none of them showing any lichenoid inflammation. We herein report the first case of a patient that suffered from Cogan disease and developed isolated lichen planus on all nails a few years after the first diagnosis. The combination of two autoimmune disorders is not unusual and raises the question of common immunogenetic pathomechanisms.

© 2019 The Author(s)
Published by S. Karger AG, Basel

Introduction

Cogan syndrome (CS) and nail lichen planus represent two autoimmune disorders. Both have been associated with other autoimmune disorders. So far only two cutaneous diseases, none of them lichenoid, were described in patients with CS.

Case Report

We report on a 49-year-old patient that was referred for nail consultation for evaluation of a progressive onychodystrophy involving most of his fingernails. The patient was treated for CS, which had been diagnosed 15 years prior to this consultation, with auditory and ocular involvement. At that time, the therapy included systemic corticosteroids, azathioprine, and cyclophosphamide. Cessation of the acute inflammation was achieved, but the hearing impairment persisted. There was no personal or family history of inflammatory skin diseases or other autoimmune disorders.

Clinical examination revealed thinned nail plates with a rough surface, longitudinal fissuring, and distal onycholysis of eight fingernails. Both the thumb and big toenails showed a thickened nail plate with xanthonychia (Fig. 1). Otherwise, examination of the whole body including the hair and mucous membranes was unremarkable. A lateral longitudinal nail biopsy was performed, and light microscopy studies showed a characteristic dense band-like lichenoid lymphocytic infiltrate in the proximal nail matrix and undersurface of the proximal nail fold (Fig. 2). With this clinicopathological correlation, the diagnosis of nail lichen planus was confirmed.

Discussion

CS is a rare type of ANCA-negative vasculitis with predominant involvement of the eyes and vestibulocochlear system. Typical symptoms include keratitis and a Ménière-like triad of vertigo, tinnitus, and sensorineural hearing loss that often persists after remission [1]. Systemic manifestations of vasculitis are present in up to 50% of the patients, with complaints such as fever, arthralgia, and myalgia [1–4]. Aortitis can evolve as a serious complication of CS [4, 5]. Systemic corticotherapy associated with disease-modifying antirheumatic drugs and TNF inhibitors currently represents the main treatment of CS. Infliximab showed superior effects in vestibuloauditory outcomes in a retrospective study [6].

In the current literature, pyoderma gangrenosum and vasculitis were described as the only cutaneous manifestations in patients with CS [7]. So far, no lichenoid reactions were reported.

Nail involvement of lichen planus may occur as an isolated finding but more often in association with other mucocutaneous manifestations [8]. Involvement of the nail matrix characteristically leads to thinning of the nail and roughening of the nail surface, called trachyonychia, as well as longitudinal ridging and fissuring. Subungual hyperkeratosis and distal onycholysis as signs of nail bed involvement are less frequent [9].

Immune dysregulation with a predominantly CD8+ T-cell reaction plays an essential role in the pathophysiology of the disease [10].

The combination of two or even more autoimmune disorders is not unusual. It is often explained by immunogenetic relations between these diseases. However, until now, no such immunogenetic links between CS and nail lichen planus have been known. Thus, it remains speculative whether this association is just fortuitous or based on a common pathomechanism.

Statement of Ethics

Informed consent was given by the patient. All procedures of this study were in accordance with the standards for human experimentation and with the Helsinki Declaration of 1975, as revised in 1983.

Disclosure Statement

The authors have no conflict of interest to disclose.

Author Contributions

Both authors had full access to all the data in the study and take responsibility for the integrity of the data and the accuracy of the data analysis. Drafting of the manuscript: both authors. Critical revision of the manuscript for important intellectual content: E. Haneke.

References

- 1 Kessel A, Vadasz Z, Toubi E. Cogan syndrome – pathogenesis, clinical variants and treatment approaches. *Autoimmun Rev*. 2014 Apr-May;13(4-5):351–4.
- 2 Angiletta D, Wiesel P, Pulli R, Marinazzo D, Bortone AS, Regina G. Endovascular treatment of multiple aneurysms complicating Cogan syndrome. *Ann Vasc Surg*. 2015 Feb;29(2):361.e9–361.e12.
- 3 Sevgi DD, Sobrin L, Papalioidis GN. Cogan syndrome with severe medium and large vessel vasculitis. *Digit J Ophthalmol*. 2015 Feb;22(1):32–4.
- 4 Gluth MB, Baratz KH, Matteson EL, Driscoll CL. Cogan syndrome: a retrospective review of 60 patients throughout a half century. *Mayo Clin Proc*. 2006 Apr;81(4):483–8.
- 5 Kaya M, Erkanlı K, Kılınc F, Sar M, Bakır İ. Surgical Treatment in a Case of Cogan's Syndrome Complicated with Proximal Aortic Vasculitis. *Ann Thorac Surg*. 2015 Oct;100(4):1467–9.
- 6 Durtette C, Hachulla E, Resche-Rigon M, Papo T, Zénone T, Lioger B, et al.; SNFMI and CRI. Cogan syndrome: characteristics, outcome and treatment in a French nationwide retrospective study and literature review. *Autoimmun Rev*. 2017 Dec;16(12):1219–23.
- 7 Chua EP, Mallett RB, Dahiya S. Cogan's syndrome with pyoderma gangrenosum: management of two uncommon disorders with aggressive presentation in a patient. *BMJ Case Rep*. 2018;2018:bcr-2017-223876.
- 8 Haneke E. Non-infectious inflammatory disorders of the nail apparatus. *J Dtsch Dermatol Ges*. 2009 Sep;7(9):787–97.
- 9 Goettmann S, Zarea I, Moulouguet I. Nail lichen planus: epidemiological, clinical, pathological, therapeutic and prognosis study of 67 cases. *J Eur Acad Dermatol Venereol*. 2012 Oct;26(10):1304–9.
- 10 Tziotzios C, Lee JY, Brier T, Saito R, Hsu CK, Bhargava K, et al. Lichen planus and lichenoid dermatoses: clinical overview and molecular basis. *J Am Acad Dermatol*. 2018 Nov;79(5):789–804.



Fig. 1. a–d The clinical features involve distal onycholysis of almost all fingernails (a–c) and the big toenails (d) due to nail bed involvement. The majority of the nails show thinning of the nail plate and xanthonychia.

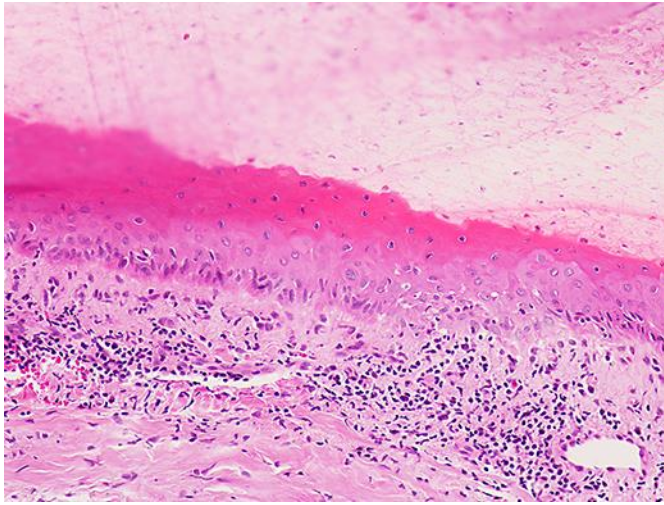


Fig. 2. Lateral longitudinal nail biopsy showing a dense band-like lymphocytic infiltrate in the matrix typical of nail lichen planus.