Case Report Annals of Dermatology 2022;34(5):378-381 • https://doi.org/10.5021/ad.20.145

Postherpetic Trigeminal Trophic Syndrome: A Case Report

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Received June 8, 2020
Revised October 22, 2020
Accepted October 31, 2020

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Trigeminal trophic syndrome (TTS) is a rare condition characterized by anesthesia, paresthesia, and facial ulceration involving the trigeminal dermatome secondary to self-manipulation of the skin after a peripheral or central injury to the trigeminal nerve or its branches. Differential diagnosis of TTS includes conditions presenting with chronic facial ulceration, such as various infectious diseases, malignancy, vasculitis, pyoderma gangrenosum and dermatitis artefacta. We report a case of postherpetic TTS and highlight the importance of early diagnosis and prompt treatment of this condition, which may commonly be misdiagnosed.

Keywords: Dermatitis artefacta, Herpes zoster, Trigeminal trophic syndrome, Ulcer

INTRODUCTION

Trigeminal trophic syndrome (TTS) is a rare condition characterized by anesthesia, paresthesia, and facial ulceration involving the trigeminal dermatome secondary to trigeminal nerve injury. Trigeminal nerve injury causes numbness, burning, tingling, or an itching sensation across the trigeminal dermatome. Patients indulge in repetitive self-manipulation to alleviate these sensations, and such compulsive self-mutilation results in chronic ulceration of the trigeminal dermatome[1]. Iatrogenic nerve ablation and ischemic injury of the trigeminal ganglion constitute the most common causes of trigeminal nerve injury, followed by tumors, trauma, surgical complications, and infections such as herpes zoster, syphilis, and leprosy, which are known to affect this nerve[1]. TTS is a diagnosis of exclusion and should be differentiated from other conditions presenting with facial ulceration. We report a rare case of post herpes zoster TTS.

CASE REPORT

A 72-year-old female presented with paresthesia and a wide area of ulceration on her left frontal scalp (Fig. 1B). She initially developed painful grouped vesicles and was diagnosed with herpes zoster ophthalmicus affecting the left ophthalmic or first division of the trigeminal nerve, 1 month prior to presentation (Fig. 1A). She was treated with antiviral drugs and analgesics. While the lesions improved, she experienced numbness, itching and tingling sensations in the same area, resulting in repetitive self-mutilation in an attempt to alleviate these symptoms. Subsequently, she developed ulceration with an alopecic patch that expanded despite antihistamine treatment. She had a history of hypertension, diabetes mellitus, heart failure and dyslipidemia. Laboratory investigations showed no abnormalities in her complete blood count, liver and kidney function and serological tests (including rapid plasma reagin, human immunodeficiency virus antibody, hepatitis B virus surface antigen and antibody and hepatitis

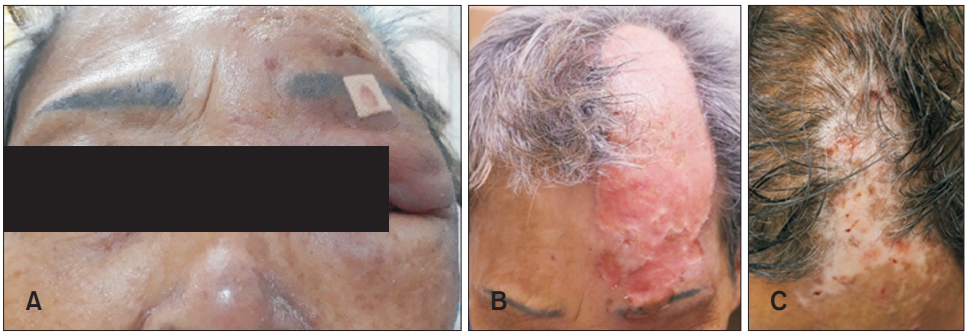

Fig. 1. Clinical pictures of the case. (A) Erythematous papules, patches develop with swelling due to herpes zoster on the left V1 dermatome. (B) Ulceration with wide fibrous alopecic patch on the left frontal scalp. (C) Healed lesion after 6 months of treatment. We received the patient's consent form about publishing all photographic materials.

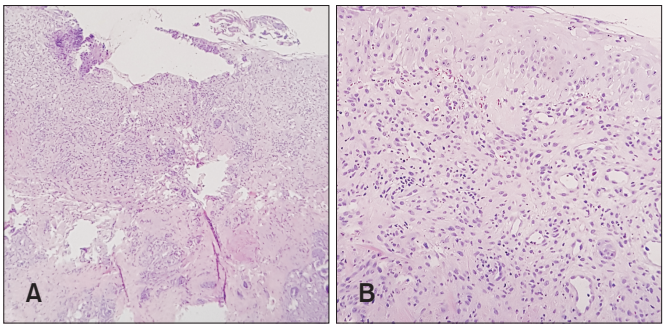

Fig. 2. Histological pictures of the case. (A) Crust and ulcer with epidermal atrophy in epidermis and interstitial inflammatory infiltration in dermis (H&E, ×40). (B) Lymphocytes and neutrophils are observed with liquefactive degeneration in dermis (H&E, ×200).

C virus antibody) and basic chemistry profile (including sodium, potassium, chloride, magnesium, phosphorus, calcium, protein and albumin). Antinuclear antibody test results were positive with a 1:160 titer (nucleolar pattern). However, additional laboratory test results, including anti-double stranded DNA antibody, anti-centromere antibody, anti-RNP antibody, anti Scl-70 antibody, and anti-Smith antibody were unremarkable. Histopathological evaluation of a biopsy specimen showed epidermal atrophy, liquefactive degeneration and interstitial infiltration of lymphocytes and neutrophils in the dermis without evidence of vasculitis or malignancy (Fig. 2). The patient was diagnosed with TTS and was treated with gabapentin (100 mg twice daily), amitriptyline, antihistamine, topical antibiotics, topical steroid, and dressing changes once a week. The patient was instructed not to scratch the lesion. The ulcer and alopecic patch showed clear improvement with treatment (Fig. 1C).

DISCUSSION

Various conditions that cause of injury to the trigeminal nerve and/or its branches can result in TTS. The exact pathogenesis of TTS and persistent ulcer formation remains unclear; however, previous studies have suggested that self-mutilation secondary to anesthesia and dysesthesia of the affected trigeminal dermatome is the main contributor to TTS[2]. Other studies attribute ulceration to vasomotor nerve injury[3,4]. Trigeminal nerve ablation and cerebrovascular disease are the most common causes of TTS; tumors, surgical complications, and infections are other likely contributors[5]. A few reports in the literature have described meningioma, maxillofacial surgery, and synthetic marijuana use as rare causes of TTS[4-6]. Our patient developed anesthesia, paresthesia and itching sensation after improvement of herpes zoster. Self-manipulation of the skin in an attempt to reduce these uncomfortable sensations resulted in ulceration in the affected dermatome. A few cases of postherpetic TTS are reported overseas; however, TTS has not been previously reported in Korea[2,7,8]. Reactivation of the varicella zoster virus in the dorsal ganglion following herpes zoster infection precipitates a cellular immune response with neuritis, which results in TTS, as observed in our patient[7].

Facial ulceration in patients with TTS usually presents as unilateral crescent lesions that affect the trigeminal dermatome innervated by the second division of the trigeminal nerve (the nasal alar area is most commonly involved) in approximately 80% of cases[9]. Our patient presented with an ulcer and a wide fibrous alopecic patch on the left frontal scalp. Scalp involvement (as observed in our patient) occurs in less than 10% of patients with TTS[10]. A few patients with TTS show involvement of the cornea, bone, and the paranasal sinuses[8]. Previous studies have reported that the latent period between nerve injury and ulcer formation ranges from 2 weeks to 20 years. The mean latent period was approximately a year, and ulcers developed within 3 months after nerve injury in most (33%) cases of TTS[1,7,11]. The latent period for the development

of TTS after herpes zoster infection ranged from 3 months to 3 years, and the mean period was approximately a year, which is longer than that observed with typical TTS[2,7,8,12-14]. In our patient, the ulcer developed after 2 weeks of the latent period. Approximately 50% of patients with post herpes zoster TTS show lesions involving the trigeminal dermatome innervated by the first division of the trigeminal nerve (similar to our patient). However, this presentation differs from that of typical TTS in which patients present with lesions affecting the trigeminal dermatome innervated by the second division of the trigeminal nerve.

TTS should be differentiated from other conditions that present with skin ulceration, including malignancy, vasculitis, infections, granulomatous disease, and pyoderma gangrenosum[1]. Biopsy examination of specimens obtained from patients with TTS shows nonspecific inflammatory changes; however, biopsy and laboratory evaluation including autoantibody testing are important to exclude other diseases[1]. We performed histopathological examination to exclude malignancy, vasculitis, and granulomatous disease in our patient. Differentiating TTS from dermatitis artefacta is challenging; however, patients with TTS typically acknowledge scratching behavior and anesthesia over the affected area[1,9]. Our patient also presented with anesthesia and acknowledged self-manipulation of the V1 dermatome.

Currently, no standard treatment has been established for TTS; however, behavioral modifications, such as patient education regarding the development of self-inflicted lesions, cutting nails short, and the use of gloves while sleeping are a few useful strategies. Gabapentin and carbamazepine are also commonly prescribed to control anesthesia and paresthesia[1]. Surgical intervention is also effective treatment in case the patient can restrain self-manipulation[1]. Our patient was instructed not to scratch the lesions and was treated with oral gabapentin, topical agents, and regular dressing changes. Most cases reported in the available literature describe treatment with oral gabapentin, topical steroids and antibiotics, as well as regular dressing changes[11]. Several case reports have described significant improvement in patients with behavioral modification, and use of topical agents and dressings without pharmaceutical interventions; therefore, behavioral modification is an indispensable component of therapy in patients with TTS[2,7,14]. Previous studies have reported that approximately 60% of patients showed significant or complete improvement within 6 months; however, 30% of patients showed recurrence within a mean period of 15 months[11]. Our patient showed significant improvement with 6 months of treatment, and no recurrence has occurred until the time of writing this report.

TTS is a rare condition and can be easily misdiagnosed; therefore, it should be distinguished from other conditions with a similar clinical presentation of ulcerative lesions. Delayed diagnosis and inappropriate management without behavioral modification tend to result in chronic ulcers[9]. To our knowledge, this is the first report that describes a rare case of TTS (which remains clinically challenging) in Korea.

CONFLICTS OF INTEREST

The authors have nothing to disclose.

FUNDING SOURCE

None.

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