Trigeminal trophic syndrome mimicking squamous cell carcinoma.

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Abstract

Trigeminal trophic syndrome is a rare cause of persistent secondary facial ulceration due to trigeminal nerve damage. Lesions often mimic squamous cell carcinoma but can be differentiated histologically. We present an unusual case of trigeminal trophic syndrome with both clinical and histologic features of squamous cell carcinoma.

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INTRODUCTION

Trigeminal trophic syndrome (TTS) is a rare cause of persistent facial ulceration. Lesions arise from repetitive self-inflicted skin trauma after insult to the trigeminal sensory system.[1] Facial dysesthesia and anesthesia result from damage to the trigeminal nerve or sensory nuclei and provoke excessive skin-picking.[2]Clinically, TTS resembles other primary cutaneous diseases and is often misdiagnosed due to its rarity.[3,4] Thorough

neurologic history and examination are essential to accurately diagnose the disorder. Herein we present a unique case of TTS clinically and histologically mimicking cutaneous squamous cell carcinoma (SCC).

CASE REPORT

A 56-year-old man with history notable for recent ischemic stroke presented to an outpatient dermatology clinic for evaluation of progressive ulcerations on the face that initially started on the right nasal ala and spread to involve the right nasolabial fold and right upper and lower cutaneous lip (Figure 1). The patient reportedly had a nevus on the right side of his nose, which he picked off prior to symptom onset. He was treated with multiple courses of antibiotics for suspected soft tissue infection but his symptoms persisted and he developed progressive tissue loss and retraction over the right alar rim. Multiple tangential biopsies were obtained and revealed atypical squamous proliferation. The patient was referred for Mohs micrographic surgery for a presumed diagnosis of well-differentiated SCC.

Punch biopsies of the lesions at 3-mm thickness were performed to confirm the diagnosis and determine depth of involvement. Repeat histopathology demonstrated findings consistent with lichen simplex chronicus/prurigo-like changes including hyperkeratosis, patchy parakeratosis, marked irregular acanthosis, and hypergranulosis. Focal ulceration was also seen. The dermis showed fibrosis and chronic inflammation (Figure 2). No significant keratinocyte atypia was observed. Given the history of self-manipulation and likely neurologic mechanism involving trigeminal anesthesia in the setting of a recent ischemic stroke, a final diagnosis of TTS was made. The patient was educated on the self-inflicted cause of the ulcers and advised to avoid manipulating the sites.

DISCUSSION

TTS is a rare cutaneous dysesthesia that results in chronic facial ulceration due to repeated self-mutilation and excoriation of the skin. The syndrome is most frequently reported as an iatrogenic consequence of trigeminal ablation for trigeminal neuralgia.[1] Less common causes include tumors, herpes zoster, Hansen disease, and posterior circulation stroke, as seen in our patient. Treatment consists primarily of behavioural modification to reduce skin trauma. Medications such as carbamazepine and amitriptyline can reportedly reduce paresthesia to prevent further skin manipulation.[1,5]

Diagnosis of TTS relies on thorough neurologic history and physical examination to reveal evidence of damage to the trigeminal nerves or its nuclei. Paresthesia and anesthesia ipsilateral to the skin lesion are classic in TTS, although bilateral TTS has been reported.[6] Paresthesia promotes skin-picking to relieve abnormal cutaneous sensations while anesthesia permits prolonged painless skin trauma.[2] Our patient demonstrated an atypical mechanism as he lacked a cutaneous paresthesia trigger and instead self-manipulated due to the visual perception of skin blemishes. He also reported experiencing a sensation of nasopharyngeal drainage, which has been previously described in association with trigeminal anesthesia and may serve as a diagnostic clue to TTS.[1]

TTS may resemble neoplasms, granulomatous diseases, and various infections including leprosy, cutaneous tuberculosis, and primary syphilis. Patients are often misdiagnosed and delayed care because the condition's rarity and mimicry allow it to elude the differential diagnosis for persistent skin ulceration.[3,7] Lesions tend to present as well-demarcated, sickle-shaped ulcers, most frequently around the nasal ala.[1] Histopathology can help differentiate TTS from other primary cutaneous disorders and is expected to show nonspecific inflammatory changes consistent with chronic mechanical trauma, including dermal fibrosis, hyperkeratosis, hypergranulosis, and lichenification.[1,2,7] As demonstrated in our case, TTS can be histologically identical to nodular prurigo, another secondary skin disorder caused by chronic rubbing.

Superficial biopsies of our patient's lesions initially demonstrated atypical squamous proliferation that led to early suspicion for well-differentiated cutaneous SCC. While TTS has been known to clinically resemble SCC, there are no previous reports of this syndrome containing histologic features of SCC.[8,9] Atypical squamous proliferation is most commonly associated with cutaneous SCC, however it can also appear in actinic keratosis, warts, and benign inflammatory dermatoses like prurigo nodularis.[10] As a nonspecific

histologic description, the finding of atypical squamous proliferation alone likely indicates an insufficient or shallow sample, and it would be responsible practice to obtain a deeper repeat biopsy if possible to confirm the diagnosis. Our unique case confirms that atypical keratinocyte proliferation may also present in TTS, likely due to chronic inflammation caused by repeated skin-manipulation. Dermatologists should be aware of this unusual histopathological manifestation of TTS to avoid misdiagnosis, unnecessary surgery, and delay in treatment.

ABBREVIATIONS AND ACRONYMS

TTS: trigeminal trophic syndrome

SCC: squamous cell carcinoma

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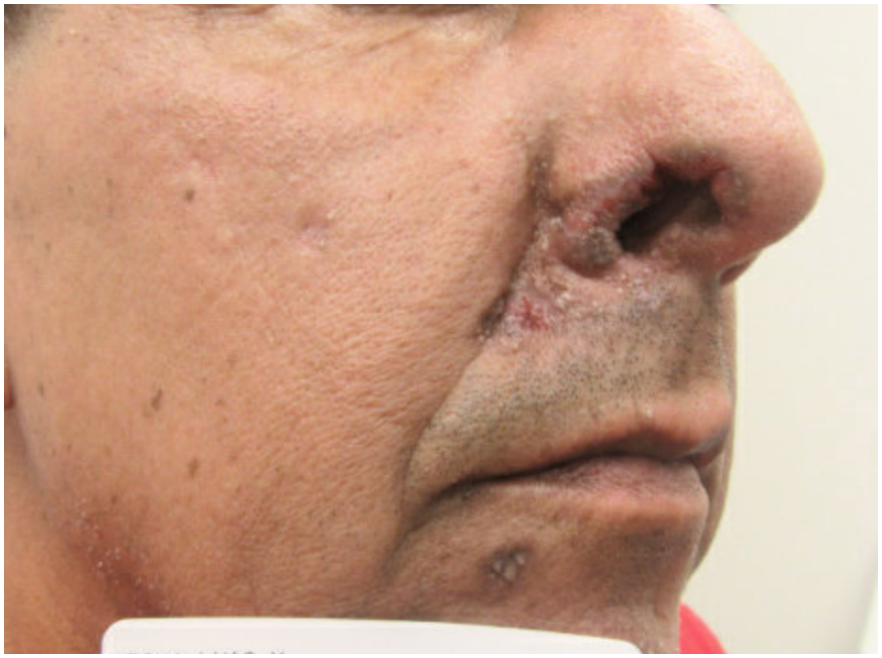
Figure 1. Unilateral erosions, ulcerations, and alar rim retraction upon initial presentation (A) and at subsequent visit (B).



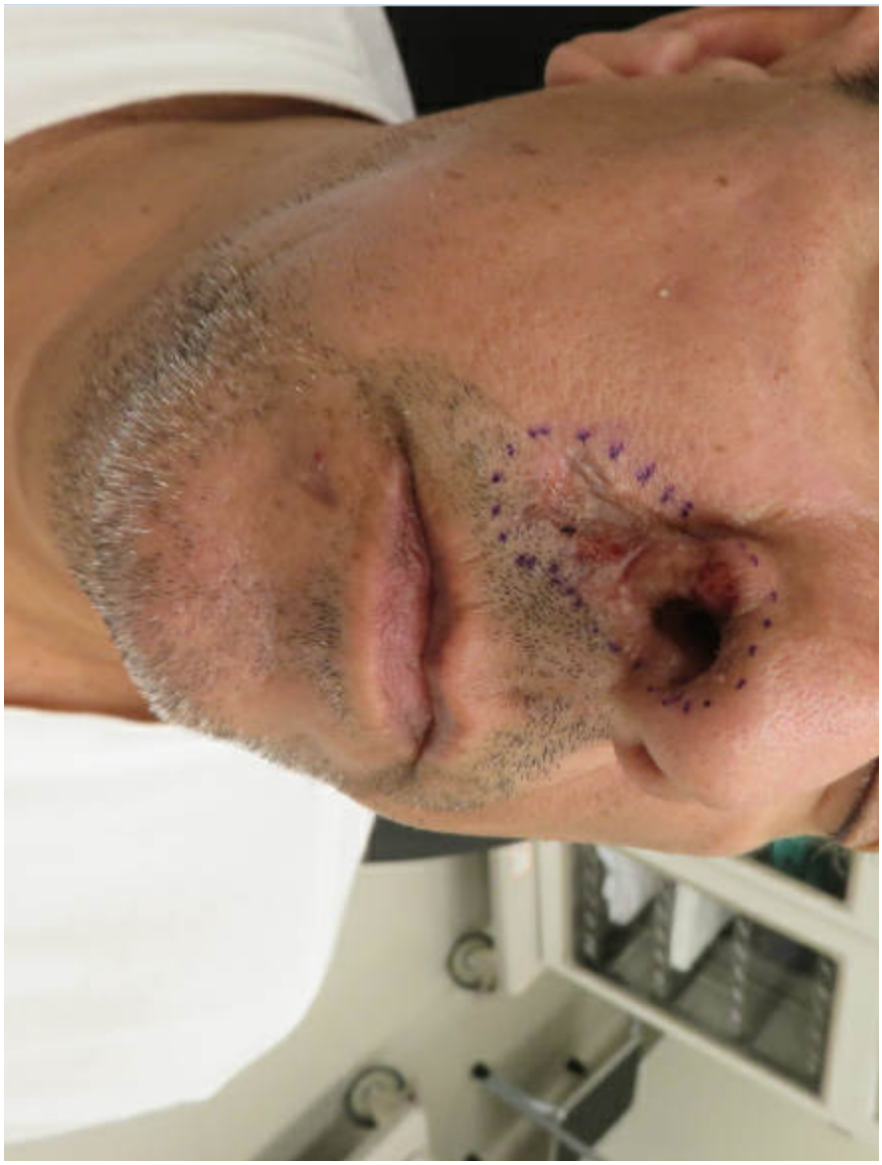


Figure 2. Histologic features of TTS on punch biopsy. Specimen of right nasal ala (A) at 10X magnification shows hyperkeratosis, marked irregular acanthosis and hypergranulosis without significant keratinocytic atypia. Dermal fibrosis and chronic inflammation are present. Specimen of right upper cutaneous lip (B) at 4X magnification shows ulceration with irregular acanthosis and no significant keratinocytic atypia. Dermal fibrosis and chronic inflammation are present.