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**Photo vignette**

# A rare case of trigeminal trophic syndrome with an extensive scalp, forehead, and upper eyelid ulceration in a patient with undiagnosed Alzheimer disease

**Pinar Incel Uysal, Refika Ferda Artuz, Basak Arslan Yalcın**

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**Department of Dermatology, Ankara Numune Training and Research Hospital, Ankara, Turkey**

**Correspondence:**

Pinar Incel Uysal, MD
Address: Kızılırmak Mah. 1424. Cad. Pembekosk apt. 25/30 Cukurambar, ANKARA, 06520
E-mail: incelpinar@gmail.com
Phone : 00903125084040
Fax: 00903123114340

## Abstract

**Background:** Trigeminal Trophic Syndrome (TTS) is a rare presentation of facial ulceration, which is characterized by the triad of anesthesia, paraesthesia, and damage of trigeminal sensory branches.

**Main observations:** We report a unique case of TTS as an extensive forehead and scalp ulceration in a patient with undiagnosed Alzheimer disease.

**Conclusions:** Treatment options for trigeminal trophic syndrome are limited and disappointing especially in older patients with dementia. Family education and behavioral modification therapies may be well tolerated option in this population.

**Keywords: Alzheimer Disease, facial ulceration, trigeminal trophic syndrome**

## Introduction

Trigeminal trophic syndrome (TTS) occurs as a result of damage to the trigeminal nerve. The classical triad of TTS includes intractable ulceration with anesthesia and paresthesia. Patients frequently rub or scratch the affected zones when they try to alleviate these uncomfortable sensations and this eventually causes persistent ulceration or tissue loss. TTS was reported to be rare by Sadeghi et al [1].

## Case synopsis

An 81-year-old man had developed persistent left-sided forehead and scalp skin ulceration for 1 year. Ten months prior to his presentation his family noticed his repetitive manipulation of the forehead and frontal scalp skin. His medical history included hypertension, congestive cardiac failure, and aortic aneurism. He had prior surgeries for femur fracture and gastric and renal cancer. He had no history of skin disease. His family reported his cognitive decline and dementia for about 2 years. However, he had not been referred to a neurologist previously.

On dermatologic examination, well demarcated shallow ulceration with clean borders and eythematous background was present at the left upper lid, brow, forehead, and frontal scalp along the distribution of ophthalmic ($V_1$) branch of the trigeminal nerve (Figure 1a-b). Cicatricial alopecia was observed on left sided frontal scalp skin, eye-brow, and upper eyelashes. There was a hypopigmented cicatricial zone all around the ulceration. A few islands of re-epithelization were observed by the side of the left margin. Neurological examination revealed decreased pain, temperature, and light touch

sensations overlying the left sided ulcerated area. Initially tissue culture and punch biopsy were performed. At presentation the differential diagnosis included trigeminal trophic syndrome, factitious disorder, squamous cell carcinoma, and erosive pustular dermatosis. Pending culture and biopsy results conservative therapy with topical rifomycin and occlusive dressing was initiated. He was evaluated and diagnosed as having Alzheimer disease by the neurology and psychiatry departments. Treatment with Vitamin B (1000mcg/day, orally) and donepezil (5 mg/day, orally) was started. Ophthalmologic examination revealed neurotrophic keratitis and blepharitis. Appropriate ophthalmologic therapy was initiated.

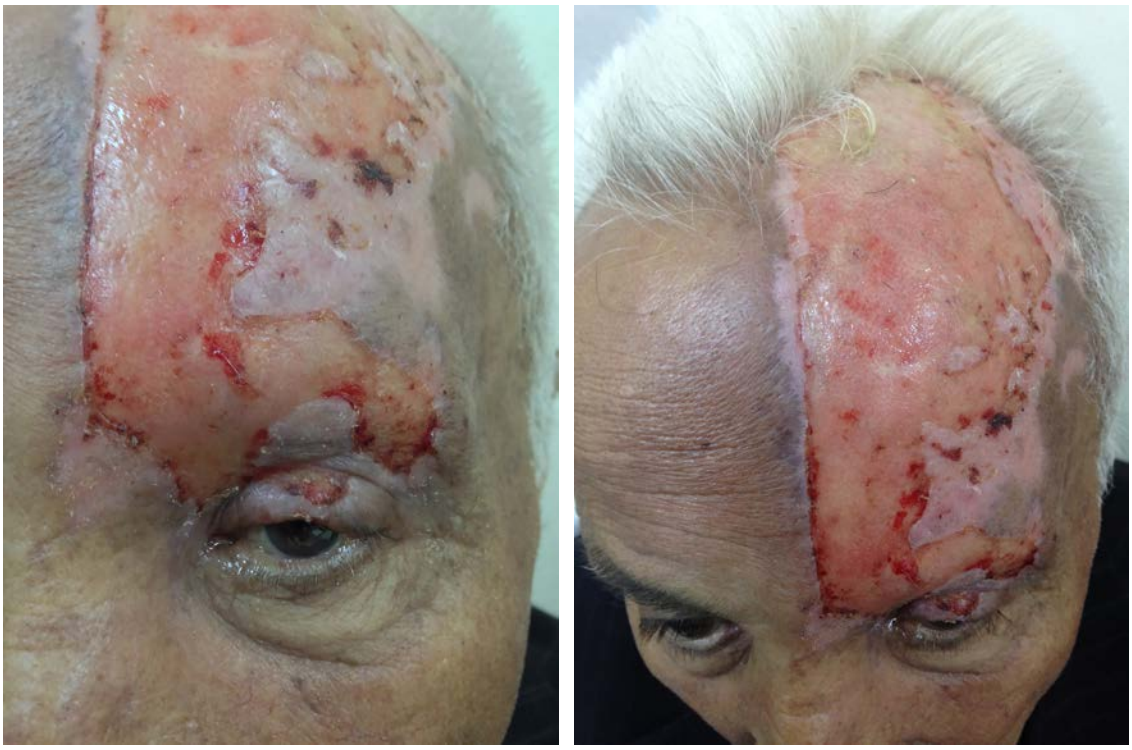

**Figure 1.** (a-b) Well demarcated left-sided ulceration along the distribution of V1 branch of trigeminal nerve.

Tissue culture was negative for bacteria and fungi. Histopathological examination revealed reactive nonspecific chronic inflammatory infiltrate rich in neutrophils within the epidermis and periadnexial dermis. There was not any evidence of vasculitis, granuloma, neoplasia, or infection. Laboratory tests including complement levels and anti-nuclear antibodies (ANA) were normal. Anti HBsAg, HIV RNA, HCV RNA, and syphilis serologies were negative. Computed tomography showed atrophy of cerebrum and cerebellum and bilateral calcified aterom plaques in internal carotid arteries. Magnetic resonance imaging was unremarkable.

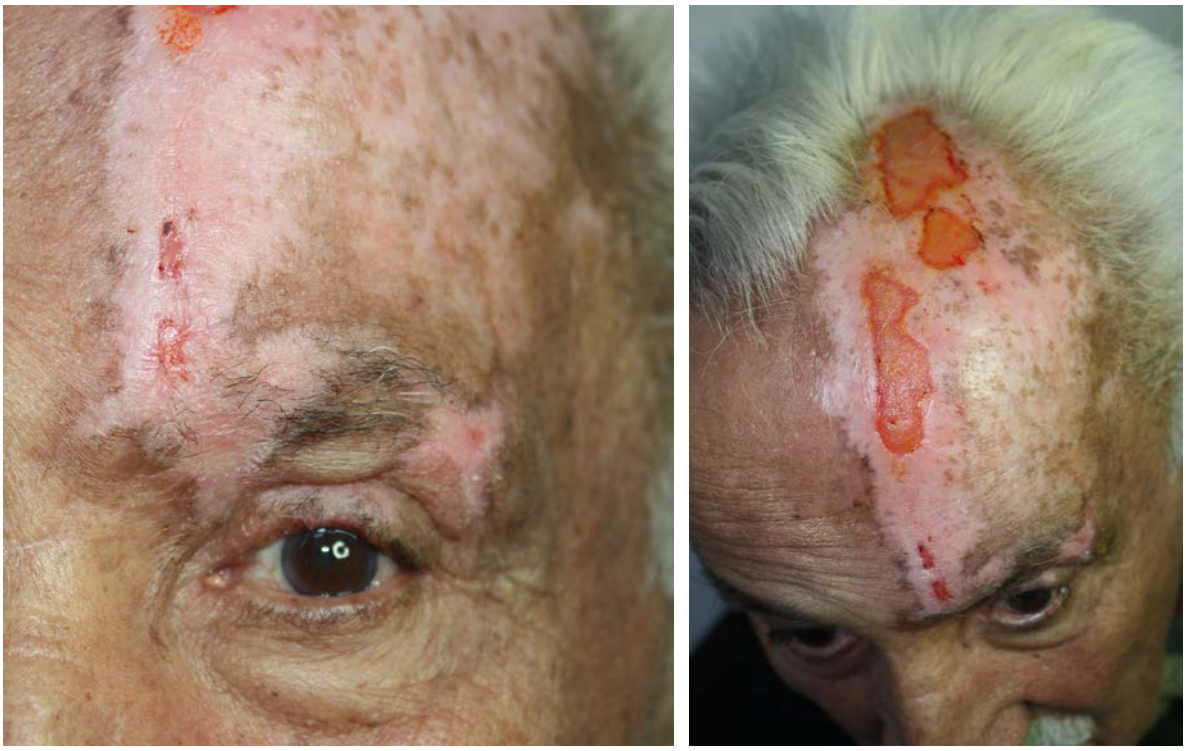

**Figure 2.** (a-b) Improvement of the ulceration 1 month after initial presentation.

On the basis of the patient's history, clinical appearance, location, and progression of ulceration and nonspecific histological findings, trigeminal trophic syndrome affecting distribution of first branch of trigeminal neuron was diagnosed. On examination 1 month after the patient's initial evaluation the ulcer was almost completely resolved (Figure 2a-b). Furthermore, his family reported recovery of his manipulative behaviors. Treatment with donepezil (10mg/day, orally) and topical occlusive dressing were recommended until he could return for follow-up examination. Unfortunately, he failed to return for follow-up but his family reported further improvement of ulceration by the phone.

# Discussion

Trigeminal trophic syndrome is a rare cause non-healing facial or scalp ulceration, which develops from self-manipulation. Although the exact pathologic process is unknown, trigeminal trophic syndrome is an unusual complication developing after peripheral or central damage to trigeminal nerve including trigeminal nerve ablation, cerebrovascular accidents, meningioma, acoustic neuromas, postencephalitis, leprosy, or syphilis. Dementia and mental impairment have been suggested to prevent self-inhibition of habitual manipulation and non-intentional self-injury [1,2]. Harper et al. reported that mental abnormality was described %19 of trigeminal trophic syndrome cases in literatüre [3]. In our opinion, cerebrovascular microocclusive process may cause ischemic damage of trigeminal ganglion and coexisting Alzheimer disease- as a behavioral component- may provoke habitual manipulation and forming such an extensive ulceration.

Ulceration classically develops in areas that are supplied by specific sensory branches of the three major trigeminal divisions. Ala nasi and nose are the most frequent sites of ulceration with only small percentages of lesions involve other areas of skin [4,5]. Our case is an interesting example of trigeminal trophic syndrome with diffuse, sharp demarcated ulceration with eyelid, eyebrow, forehead, and scalp involvement located on the left sided $V_1$ dermatome. Other potential causes of scalp and forehead ulceration including malignancy -squamous cell carcinoma-, factitious disorder, infectious and granulomatous diseases, and erosive pustular dermatosis of scalp should be ruled out. Notably, it is important to rule out delusional parasitosis, which can occur without an organic cause. Both conditions present with same concerns. However, trigeminal trophic syndrome tends to be restricted to a specific dermatome, as in our case.

In our opinion trigeminal trophic syndrome may be a more common entity and not a rare phenomenon. Even if it is correctly diagnosed, it is difficult to manage. Patient and also family education is crucial to promote healing. Conservative management may be the initial approach. Reports are available on medical therapy including gabapentin, carbamazepine, pimozide, amytriptiline, and diazepam, with variable responses [6,8]. Thermoplastic dressings were shown to be useful via blocking the cycle of perceived irritation and secondary compulsive rubbing in two cases [5]. Treatment options including cultured epidermal cells have been shown to be effective with the aim of tissue regeneration [9]. Behavioral modification therapies should be combined with surgical methods to prevent recurrence of ulcer. Our patient who is a poor candidate for surgical repair was controlled with combination pharmacological and behavioral modification therapy. Donepezil, which is an acethylcolinesterase inhibitor may help to improve cognitive impairment and control habitual skin picking.

Overall, it is important to emphasize that patient and family education is crucial for maintenance of tissue integrity. Of note, corneal lesions were reported to be noted in 18% of cases by Sawada et al [5]. In concordance with these findings, our case supports the importance of ophthalmologic evaluation. We hope awareness of this syndrome will lead to further reports and improve management.

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