# Ulcerative C2 neurocutaneous dysesthesia (trigeminal trophic syndrome in an alternative distribution)

Dylan Haynes MCR, Christina Topham BS, Tamar Hajar MD, Alex G Ortega-Loayza MD, Kevin P White MD, Jesse J Keller MD

Affiliations: Department of Dermatology, Oregon Health & Science University, Portland, Oregon, USA

*Corresponding Author: Name: Jesse Keller MD, OHSU Dermatology, 3303 SW Bond Avenue, Portland, OR, 97239, Tel: 503-418-3376, Fax: 503-494-6907, Email: kellerje@ohsu.edu*

## Abstract

Trigeminal trophic syndrome is an uncommon condition characterized by paresthesia, itch, and self-inflicted wounds following the trigeminal dermatome(s). Similar processes adhering to cervical nerve distributions have been reported, calling into question the specificity of trigeminal trophic syndrome for the trigeminal network. Herein, we report patient with trigeminal trophic syndrome adhering to the C2 dermatome, a previously unreported distribution.

*Keywords: dysesthesia, neurocutaneous, trigeminal trophic syndrome*

## Introduction

Trigeminal trophic syndrome is a rare and poorly understood condition which characteristically results in self-inflicted wounds following dermatomal distributions of the terminal trigeminal branches [1]. Most commonly, the syndrome arises in the setting of cerebrovascular accidents or trigeminal nerve ablations, though craniofacial surgery may be responsible for up to 21% of cases [2]. The ensuing lesions are suspected to occur from insidious trauma to patches of intractable paresthesia [1-3]. We present a case of a trigeminal trophic-like syndrome adopting a previously unreported sensory distribution.

## Case Synopsis

A 98-year old woman with longstanding cardiovascular disease and non-insulin-dependent diabetes mellitus was admitted for evaluation of a chronic, non-healing, post-auricular ulceration. History revealed an episode of right-sided unilateral tinnitus approximately two years prior, which prompted otolaryngologic evaluation and external auditory canal debridement. Within weeks of the procedure, a small, pruritic ulcer developed on the right posterolateral scalp which rapidly expanded in size over the proceeding months before eventually stabilizing. Supervised dressing changes proved unsuccessful in managing the ulceration, as did repeated surgical debridement, multiple courses of systemic antibiotics, and an extensive hyperbaric oxygen regimen (greater than 40 treatments). Ten months into the disease course, wound care was complicated by a persistent skin and soft tissue infection requiring hospitalization. Prior to discharge a dermatological consult was requested.

On examination, the patient was noted to have a 15cm by 20cm, sharply demarcated, geometric ulcer with central granulation tissue and fibrinous exudate overlying the projection of the right temporal bone extending to, but sparing, the pinna (**Figure 1**). Laboratory testing, including a comprehensive metabolic panel, complete blood count, anti-nuclear antibody screening, erythrocyte sedimentation rate, C-reactive protein level, and basic nutritional studies were unremarkable apart from borderline pre-albumin and zinc levels. Peri-lesional skin biopsy

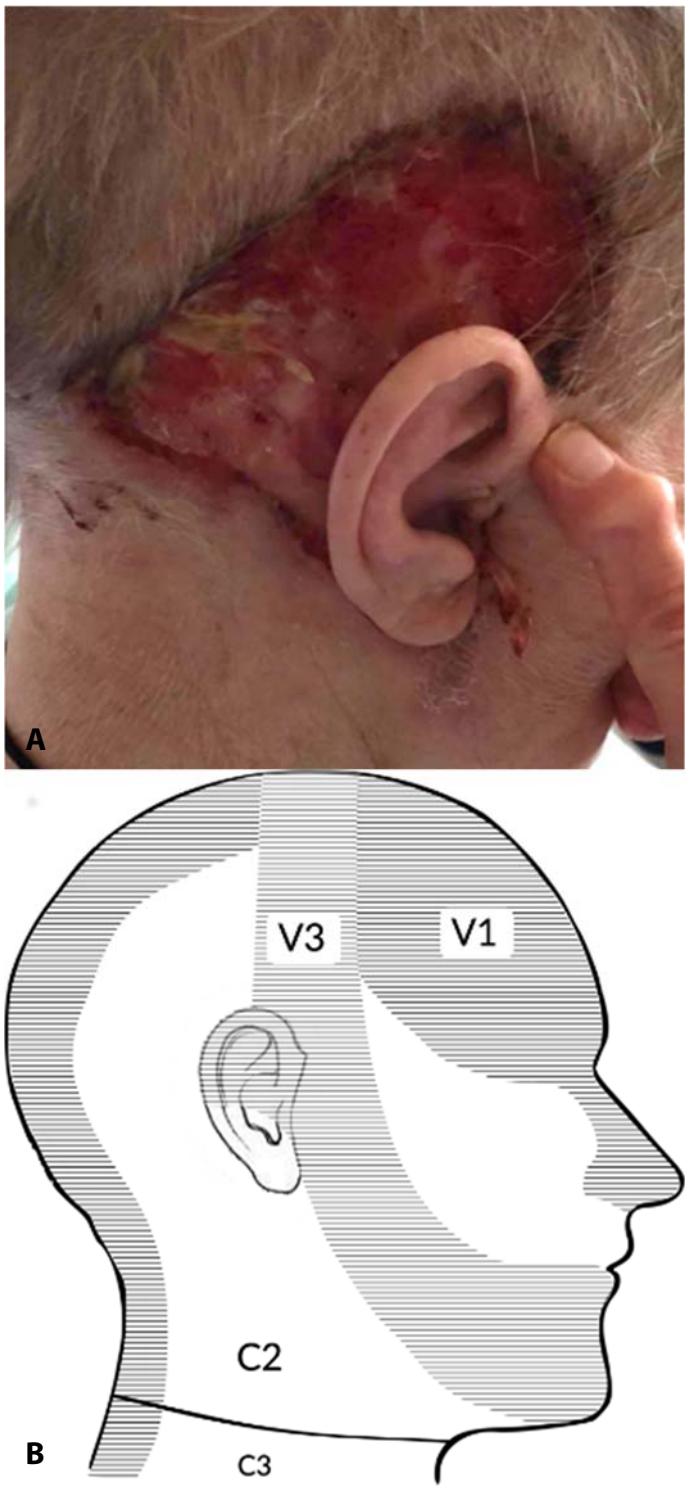


**Figure 1**. *Lesion morphology and distribution.* ***A)*** *Clinical photography demonstrating ulcer adherence to C2 dermatome by* ***B)*** *dermatomal mapping.*

revealed perifolliculitis and a nonspecific nodular, suppurative, and granulomatous dermatitis (**Figure 2**). Given the lesion's pruritic nature, geometric shape, lack of treatment response, and strict adherence to the C2 dermatome, a diagnosis of cervical variant trigeminal trophic syndrome was made.

## Case Discussion

The differential diagnosis for trigeminal trophic syndrome is broad and made only after exclusion of other infectious, neoplastic, or inflammatory conditions. Clinically, trigeminal trophic syndrome presents with crusted, painless, crescent-shaped ulcerations with a granulating base. Tissue biopsy, although generally non-specific, is essential to exclude other potential etiologies [1]. In the present case, particular consideration was given for erosive pustular dermatosis of the scalp (following development of a solitary pustule along the posterior wound edge), pyoderma gangrenosum, basal cell carcinoma, and temporal arteritis.

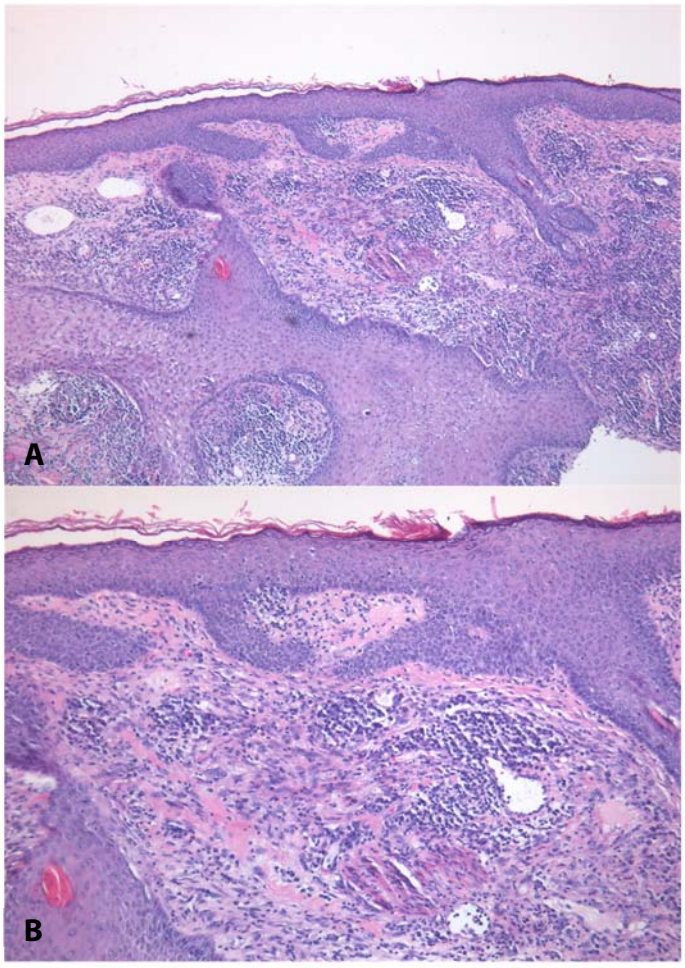


**Figure 2**. *Lesional biopsy. Perifolliculitis with non-specific granulomatous dermatitis by hematoxylin and eosin staining at* ***A)*** *5×,* ***B)*** *10× magnifications.*

Ultimately, the lack of vertex scalp involvement, minimal crusting, and equivocal response to corticosteroids disfavored erosive pustular dermatosis of the scalp. Pyoderma gangrenosum was believed less likely in the absence of frank pathergy to wound debridement and sparse neutrophilic infiltrate on histopathology. Histologic findings were also inconsistent with neoplastic change, ruling out basal cell carcinoma. Scalp necrosis from temporal arteritis, although rare, has been reported [4]. However, the patient's normal inflammatory markers were incompatible with an underlying temporal arteritis.

Classically, trigeminal trophic syndrome affects the nasal ala or cheek with less than 10% of reported cases involving the scalp [2]. A C2 distribution has not yet been described, but two reports of trigeminal trophic syndrome following the C3-C5 dermatomes call into question the specificity of trigeminal trophic syndrome for the trigeminal network [5, 6]. Indeed, seemingly similar clinical syndromes have acquired distinct names when localized to the C5-C6 (brachioradial pruritus) or T2-T6 dermatomes (notalgia paresthetica), [7]. With growing evidence for high cervical neurocutaneous dysesthesia patterns, we assert the need for a standardized naming convention. These authors propose using dermatomal distribution followed by neurocutaneous dysesthesia with optional specification of overlying change, such as ulcerative C2 neurocutaneous dysesthesia.

## Conclusion

Neurocutaneous dysesthesias can result in chronic, recalcitrant ulcerations following dermatomal distributions. Moreover, they may occur in the setting of planned, incidental, or intrinsic damage to cutaneous nerve supplies. Given the widely disparate naming conventions associated with these syndromes despite considerable overlap in clinicopathologic features, we propose a new standardized nomenclature which recognizes the unified spectrum of disease.

## Potential conflicts of interest

The authors declare no conflicts of interests.

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