Trigeminal Trophic Syndrome: Case Report

Trigeminal Trofik Sendrom

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Geliş Tarihi/*Received:* 08.01.2008
Kabul Tarihi/*Accepted:* 18.04.2008

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ABSTRACT Trigeminal trophic syndrome (TTS) is an unusual complication of peripheral or central damage to the trigeminal nerve and characterized by a triad of trigeminal anesthesia, paresthesia and ala nasi ulceration. Patients spontaneously refer repeated manipulation like picking, rubbing and/or scratching at the affected areas because of anaesthesia, paraesthesia and/or pain. One or several crescentic ulcerations unilaterally may appear on a trigeminal dermatome, especially on the ala nasi. We report a TTS case that shows the characteristic clinical and histological features of this disorder as a sequela of cerebrovascular diseases. To our knowledge, this case is the first report of TTS from Turkey.

Key Words: Trigeminal nerve diseases; nose deformities, acquired

ÖZET Trigeminal trofik sendrom (TTS), trigeminal sinirin periferik veya santral bağlantılarının hasarlanmasıyla ortaya çıkan, trigeminal anestezi, parestezi ve burun kanadında ülserasyon triadıyla karakterize nadir görülen bir durumdur. Trigeminal sinirin hasarlanmasına bağlı oluşan anestezi, parestezi veya ağrı hastalarda farkında olmadan etkilenen bölgeyi koparma, ovuşturma ve kaşıma gibi tekrarlayan manüplasyonları tetiklemektedir. Buna bağlı olarak da sıklıkla burun kanadında olmak üzere trigeminal sinir dermatomunda bir veya bir kaç tane, tek taraflı, yarım ay şeklinde ülserasyonlar oluşmaktadır. Biz burada bu hastalığın karakteristik klinik ve histolojik özelliklerini gösteren ve serebrovasküler hastalık sonrası sekel olarak oluşan bir TTS olgusu bildirdik. Bizim bilgilerimize göre bu, Türkiye'den bildirilen ilk TTS olgusudur.

Anahtar Kelimeler: Trigeminal sinir hastalıkları, kazanılmış nazal deformite

Turkiye Klinikleri J Neur 2008;3:74-76

Trigeminal trophic syndrome (TTS) is an unusual complication of peripheral or central damage to the trigeminal nerve.[1] One or several crecentic ulcerations unilaterally may appear on a trigeminal dermatome, especially on the ala nasi.[1-3] Differentiation of TTS from malignancy, infection, vasculitis, factitial dermatitis and granulomatous diseases is possible on the basis of clinical history, tissue biopsy, and serologic evaluation.[1]

Here, we report a 31-year-old female with TTS.

CASE REPORT

A 31-year-old woman presented with progressive ulceration of the left ala nasi. Three years previously, the patient had experienced a stroke after ovarian hyperstimulation therapy. The patient reported that an ulceration had

developed on her left nostril after nasogastric tube insertion when she was in the intensive care unit. Despite the cleaning and using topical bacitracin and fucidic acid, the wound continued to enlarge and bled intermittently after being discharged from the hospital. She denied any picking or scratching of the wound. Physical examination demonstrated a cresentic approximately 0.5X0.6 cm, sharply demarcated, crusted ulcer on the left ala nasi and an atrofic scar on the left part of the upper lip (Figure 1). Neurologic examination confirmed trigeminal anesthesia. Her magnetic resonance scan showed left cerebellar atrophy and right cerebellar infarct. Laboratory studies for autoimmun diseases were within normal limits. A recent tuberculin skin test was negative. Cultures and special stains were negative for bacteria, fungi, acid-fast bacteria, and viruses. Biopsy specimen revealed ulceration and nonspesific inflammatory changes without vasculitis, granulomas or neoplasia. Topical wet dressing with serum physiologic and mupirocin ointment were applied. The ulcer healed with a scar formation at the end of two weeks (Figure 2).

Trigeminal trophic syndrome (TTS) was diagnosed based on the histopathology and the triad of trigeminal anesthesia, facial paresthesias, and crescentic ala nasi ulceration.

DISCUSSION

TTS, is a rare consequence of damage to the trigeminal nerve or its central sensory connections.[1-3]

FIGURE 1: A cresentic 0.5X0.6 cm, sharply demarcated, crusted ulcer on the left ala nasi and an atrofic scar on the left part of the upper lip.

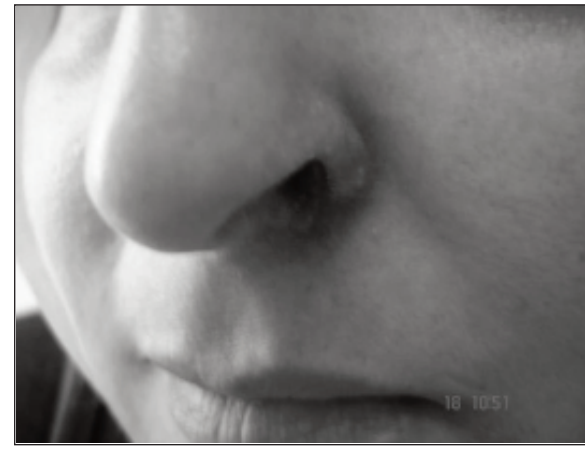

FIGURE 2: Scar formation on the ala nasi at the end of two weeks.

One-third of the patients had undergone trigeminal ablation for the treatment of trigeminal neuralgia and another third had a history of stroke[1]. Other causes of trigeminal sensory nerve damage implicated in TTS are listed in Table 1.

In all cases, patients spontaneously refer paraesthesias (burning, itching, crawling and/or tickling sensations) and repeated manipulation (picking, rubbing and/or scratching) at the affected areas.[12] The mechanism of ulceration is primarily traumatic, usually self-induced. The self-mutilating behavior may become compulsive; patients are sometimes unaware of their actions and deny wound manipulation.[1] Self-induced nasal lesions that occur in factitious disorder are primarly distinguished from those in TTS by the presence of normal trigeminal nerve function and frequent patient denial of lesion manipulation.[13] The period of latency between trigeminal injury and ulceration ranges from weeks to decades. The syndrome is more common in women and the elderly.[3,6,14] The ulcer appears throughout the trigeminal distribution, especially on the ala nasi . The tip of nose is spared, as a result of innervation by the medial branch of the anterior ethmoidal nerve. Alar ulceration occurs in noncartilagenous areas, adopting a cresentic shape that helps differentiate it from ulcers secondary to neoplastic or other inflammatory processes.[1]

The differential diagnosis of TTS is broad and a definitive diagnosis relies on excluding other causes of nasal/facial ulceration (Table 2). The diagnosis of TTS is made clinically, but histologic examination helps differentiate it from other causes of nasal/facial ulceration. Histologically, chronic ulceration in identified with minimal inflamatory infiltrate (in absence of a secondary superinfection) and no giant cells, granulomas, vasculitic lesions, or neoplastic cells.[1]

Treatment of TTS is difficult and involves prevention of the compulsive behaviors in response to facial paresthesias[1,7]. Educating the patient about the self-induced nature of the ulceration is important. Protective devices or prostheses to cover the affected area, fingernail care, finger bandages and protective cotton gloves may help.[6,13] Systemic and topical antibiotics should be applied to treat secondary infections.[1,15] There are numereous previously reported treatment options for diminishing paresthesias and compulsive behaviors, including carbamazepine,[16] chlorpromazine,[17] and clonazepam.[18] Other interventions include transcutaneous electrical stimulation,[19] iontopheresis and nerve blockade.[10] Reconstructive surgery, autologous cultivated epidermal cells[20] and thermoplastic dressing[21] has been successfully performed.

In conclusion, TTS is an extremely rare condition characterized by a triad of trigeminal anesthesia, paresthesia and ala nasi ulceration. It is important to differantiate this syndrome from the autoimmun disorders, infectious diseases, and neoplastic process.

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