Successful Treatment of Trigeminal Trophic Syndrome in a 6-Year-Old Boy With Negative Pressure Wound Therapy

Arden E. Fredeking, MD; Robert A. Silverman, MD; Department of Dermatology, Washington Hospital Center (Dr Fredeking), and Division of Dermatology, Georgetown University Hospital (Dr Silverman), Washington, DC

The Cutting Edge: Challenges in Medical and Surgical Therapeutics

REPORT OF A CASE

A 6-year-old boy presented with 4 areas of ulceration on the right side of his face and scalp. Medical history was significant for a right-sided exophytic juvenile pilocytic astrocytoma that was diagnosed at 5 years of age after he developed a persistent head tilt to the left. Postoperative scans disclosed minimal residual tumor in the lateral brainstem.

Within a year, the patient noticed the absence of sensation on his right cheek and lip and no taste on the right side of his tongue. He began drooling from the right side of his mouth, and his parents noticed that his smile was uneven as well. A residual tumor had grown to impinge on cranial nerves V and VII, and the patient underwent a second craniotomy in an attempt to restore function. The entire tumor was removed with this second surgery, but 3 months later, the patient’s right eye became edematous and an absence of tear production was discovered. Ocular lubricant was prescribed, and the eye was patched daily for 2 weeks. Within a short time, the skin on his right forehead began to break down owing to the adhesive bandage used for his eye protection. He then developed a similar ulceration on his right nasal ala and at other locations that were not occluded with tape or dressings (Figure 1).

THERAPEUTIC CHALLENGE

These areas were unresponsive to occlusive dressings and mupirocin. Treatment with carbamazepine was started by a neurologist who assumed that the ulcerations were factitial. It was believed that the new areas of deterioration on the scalp and the medial canthus of the right eye were also self-induced in response to paresthesias created by damaged nerves.

When the patient presented to us, the results of the physical examination revealed a 1×1-cm ulcer on the right ala, a 2.5×2-cm ulcer on the anterior right scalp, a 2.8×2.5-cm ulcer on the right forehead, and a 3×1.4-cm ulcer in the right eyebrow. All 4 of these areas were similar in appearance with a glistening erythematous base and a thin serosanguineous crust.

SOLUTION

After failed attempts toward a cure with antibiotic ointment, occlusive dressing, psychotropic medication, and behavior modification, the patient was successfully treated with a vacuum device (V.A.C. GranuFoam; KCI, San Antonio, Texas) (Figure 2) made to deliver negative press-
sure wound therapy (NPWT). The ulcers on the forehead and scalp were the first to be treated. The device was worn on the ulcer on the forehead 24 hours a day for an entire week, with dressing changes on Tuesday, Thursday, and Saturday. The next week, the same was done for the ulcer on the scalp. Not only had the ulcers re-epithelialized during that time but the ulcer on the right alar had began to heal as well. After ophthalmologic consultation, the final wound on the right medial canthus was treated and healed in less than 1 week, and has remained healed for 5 months despite persistent sensory deficits (Figure 3).

COMMENT

Trigeminal trophic syndrome (TTS) (also called trigeminal neurotrophic ulceration, or trigeminal neuropathy with nasal ulceration) is a rare cause of chronic ulceration on the face that was first described in 1933 by Loveman and McKenzie. It is usually observed in elderly patients, especially in older women who have undergone ablative treatment for trigeminal neuralgia. Trigeminal trophic syndrome manifests itself as a unilateral crescent-shaped ulceration around the nasal ala, but in severe cases, there may be involvement of the jaw, forehead, cheek, and lip. We know of only 3 previously reported pediatric cases, and to our knowledge, TTS has never been successfully treated with a vacuum-dressing device.

Trigeminal trophic syndrome is a rare cause of chronic facial ulceration. Parasitic, fungal, viral, and tuberculous infections, neoplasms (squamous and basal cell carcinomas), vasculitis (Wegener granulomatosis), pyoderma gangrenosum, and factitial dermatitis must be considered. While biopsies are frequently necessary for diagnosis, this was not done in our pediatric patient. The diagnosis of TTS is made by findings in the patient’s medical history and from physical examination. Damage to the trigeminal ganglion or of the nerve from herpes infection, brainstem tumors, stroke, or trauma has been reported. Patients often describe odd paresthesias such as tingling, burning, or shooting pains, and it is thought that ulceration is unknowingly self-induced from repeatedly touching the face because of these sensations. It has been reported that the unilateral ulceration caused by TTS occurs more on the right side of the face than on the left.

Because the patient is often unaware of the part they play in their own disease, treatment is especially difficult. A great deal of damage from self-manipulation may occur during sleep when the patient has no control over his or her actions. Treatment options have included attempts at behavior modification, oral psychotropic medications to decrease the paresthesias, occlusive dressings to block mechanical trauma, and even arm splints in an attempt to stop the trauma altogether. There have been conflicting reports as to the efficacy of surgical repair. Sadeghi et al report that innervated flaps from the contralateral nasal ala or from the forehead produce good long-term results, whereas Setyadi et al report that surgical repair should only be considered if partial resolution has occurred with conservative wound care and digital manipulation has stopped completely.

Our patient was treated for TTS with a regimen of oral carbamazepine, occlusive dressings, and mupirocin that did not result in complete resolution. Rapid re-epithelialization occurred with NPWT that used the vacuum device. The reason for the concomitant healing of the nontreated alar site remains speculative. It is hypothesized that direct healing of one area supplied by a nerve may cause indirect healing in a different area supplied by the same nerve.

Our patient had suffered from these ulcers for 8 months. The chronicity of the ulcers of TTS plays a sub-

Figure 2. V.A.C. GranuFoam device (KCI, San Antonio, Texas). Special wound dressing attached to subatmospheric pressure that intermittently or continuously conveys negative pressure to help promote wound healing.

Figure 3. After treatment with negative pressure wound therapy, the ulceration on the forehead was treated first in 1 week followed by the ulceration on the scalp. All 4 ulcerations healed within 6 weeks.
substantial role in diagnosis and treatment. Chronic wounds with fibrotic changes are often more difficult to treat with conventional methods than are acute wounds. A study by Armstrong et al.\textsuperscript{10} has proven NPWT superior to standard wound therapy in both chronic and acute wounds.

The few randomized controlled trials of chronic wound treatment with NPWT have produced conflicting results. Therefore, our case is significant due to the rapid, successful treatment using this modality in a young child with chronic wounds. Simple occlusion to block the digital manipulations by our patient was not enough to promote healing. In addition to providing a secure barrier, NPWT is successful because it also removes chronic edematous fluid containing matrix metalloproteinases that degrade extracellular matrix proteins\textsuperscript{11} and it decreases the afterload for blood flow, resulting in better tissue perfusion. These factors greatly enhance the formation of granulation tissue formation.\textsuperscript{12}

Another study conducted with pig models by Morykwas et al.\textsuperscript{12} demonstrated 4-fold-higher peak flow levels over baseline values using NPWT and significantly decreased bacterial colony counts after 4 to 5 days of using the technique. A moist environment is essential for faster wound healing because it creates a hypoxic environment to stimulate angiogenesis, to cause the proliferation of normal growth factors, and to increase fibrolysis.\textsuperscript{13} Although a simple bandage with mupirocin could provide this beneficial moist environment, it cannot remove the chronic wound fluid thought to inhibit wound healing. Negative pressure wound therapy removes the glucose, albumin, and protein-poor chronic-wound fluid thought to inhibit keratinocyte, endothelial cell, and fibroblast proliferation.

In addition, it is suggested that the cells within the wound are able to respond to the mechanical forces placed on them from the vacuum device and to upregulate specific genes and induce cellular programs.\textsuperscript{14} Healing is demonstrated even in studies in which very little fluid was extracted from the wound, suggesting that these forces promote capillary sprouts and cellular growth within the wound.

We describe this patient because of very few pediatric cases of TTS published in the past and to highlight the successful treatment of chronic ulcers with NPWT delivered by the vacuum device to ulcers otherwise resistant to treatment.

Accepted for Publication: October 29, 2007.
Correspondence: Arden E. Fredeking, MD, Department of Dermatology, Washington Hospital Center, 110 Irving St NW, Washington, DC 20010 (arden04@gmail.com).

Author Contributions: Ms Fredeking had full access to all of the data in the study and takes responsibility for the integrity of the data and the accuracy of the data analysis. Study concept and design: Fredeking and Silverman. Acquisition of data: Fredeking and Silverman. Analysis and interpretation of data: Fredeking and Silverman. Drafting of the manuscript: Fredeking. Critical revision of the manuscript for important intellectual content: Fredeking and Silverman. Administrative, technical, and material support: Fredeking. Study supervision: Silverman.

Financial Disclosure: None reported.

Additional Contributions: We acknowledge Wendy Fiddler, a wound care nurse, for her help in gathering information on the treatment regimen used in this case.