Nucleus caudalis dorsal root entry zone lesioning for the treatment of anesthesia dolorosa

Case report

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While many chronic pain conditions are challenging to treat, facial anesthesia dolorosa is among the most difficult. Patients experience numbness in facial areas that also have constant severe pain. The condition results from deafferentation injuries of the first-order trigeminal nerve. These injuries can be traumatic or surgical. Anesthesia dolorosa occurs among 2%–4% of patients who have undergone trigeminal rhizotomy.

Deafferentation releases second-order neurons along the trigeminal pain pathway to generate spontaneous pain signals, without a nociceptive stimulus.

Medications used for neuropathic pain, such as gabapentin, have been recommended. Unfortunately, pharmaceutical therapy is often insufficient. As second-line approaches, surgical procedures have been proposed to either modulate or ablate neural pathways. Neuro modulation by motor cortex stimulation is often effective for some neuropathic facial pain conditions, but it has little success for anesthesia dolorosa. Deep brain stimulation has also been attempted for the treatment of anesthesia dolorosa, with mixed reports of efficacy.

Neuroablation by open surgical trigeminal nucleus caudalis DREZ lesioning has shown efficacy at bringing pain relief. The procedure is also effective for postherpetic neuralgia as well as other craniofacial pain conditions that are not responsive to less invasive therapies. The surgery is based on the understanding that pain-carrying first-order trigeminal neurons separate from motor and touch-sensation neurons at the pons. These first-order pain-carrying neurons descend to the cervicomedullary junction as the spinal trigeminal tract and then synapse to deeper second-order neurons within the trigeminal nucleus caudalis (Fig. 1). Lesions to the nucleus caudalis prevent spontaneous pain generation at this level.

Case Report

History and Examination. This 69-year-old man presented with a 7-year history of anesthesia dolorosa affecting the right V1 and V2 distributions. At onset, the patient...
had a right retroorbital headache without facial numbness. He sought care at an outside practice and was found to have a right trigeminal neuroma (Fig. 2). He underwent open resection, followed shortly afterward by Gamma Knife surgery for a portion of tumor that was not resectable. After surgery, he had new-onset numbness in the V1 and V2 distributions but experienced worsened constant pain in the same distribution. On a 0–10 pain scale, his pain averaged 8–9. Additionally, he had hypersensitivity in the rest of the right of his face to light touch and wind. For 7 years, he sought help from multiple clinics. He tried multiple medications, including gabapentin and pregabalin. He tried taking antidepressants and oxycodone, without relief of his severe pain.

After conservative measures failed, the patient was referred to the Lake Cumberland Neurosurgical Clinic for evaluation and consideration for nucleus caudalis DREZ lesioning. On examination, trigeminal motor function was intact, but sensation to pinprick and light touch was lost in the right V1 and V2 distributions and was decreased in the right V3 distribution. Extraocular movements were intact, but the patient reported some diminished vision in the right eye following his previous surgery and radiation treatment. After an interim neuropsychological evaluation, surgical options were discussed with the patient, and the decision was made to proceed with nucleus caudalis DREZ lesioning on the right side.
Operation. The patient was positioned prone in 3-point head fixation. A curvilinear suboccipital skin incision was made 1 cm to the right of midline, starting at the spinous process of C-2 and curving laterally at the level of the occipital bone. The greater occipital nerve was identified between fibers of the semispinalis muscles, which were reflected laterally. The rectus capitis posterior major muscle was dissected laterally, and the rectus capitis posterior minor muscle was detached from the posterior tubercle of C-1. Both lesser and greater occipital nerves were protected. The right C-1 hemilamina was removed, exposing the dura. A limited suboccipital craniectomy was performed. The dura was incised using a blade under direct microscopic supervision.

The arachnoid was opened to expose the medulla, C-2 dorsal roots, cranial nerve XI, dentate ligament, and vertebral artery. An 18-mm distance was measured from the level of the obex to the C-2 dorsal nerve rootlets. El-Naggar-Nashold nucleus caudalis DREZ electrodes (Cosman Medical, Inc.) were used in sequence to make a row of serial lesions along this length of the nucleus caudalis (Fig. 3). The electrodes are designed with an active tip that extends beyond a length of insulated electrode. The lengths of both components were designed to match the depth of the nucleus caudalis along the lesion path.

A total of 18 lesions were created along a path 1 mm posterior to the exiting position of the accessory nerve rootlets. The thermocoupled radiofrequency electrodes were brought to 80°C for 15 seconds per lesion. The first electrode (0.8-mm-long active tip, 0.6 mm of insulation) was used to make 4 lesions, ascending from the level of the C-2 dorsal nerve roots. The second electrode (1-mm-long active tip, 0.5 mm of insulation) was used to make 3 more ascending lesions. Continuing upward, a third electrode (1.2-mm-long active tip, 0.6 mm of insulation) was used to make 2 paired side-by-side lesions to create a slightly wider lesion segment. A fourth electrode (1.5-mm-long active tip, 0.6 mm of insulation) was used to make 5 more ascending lesions along the path. A fifth electrode (1.8-mm-long active tip, 0.6 mm of insulation) was used to make the 5 most superior lesions, finishing the row at a level 1 mm below the obex. After the lesions were made, the dura was closed with silk suture and covered with fibrin glue. The muscle layers were closed with suture in standard fashion.

Postoperative Course. The patient had an immediate and complete resolution of his V1 and V2 pain and V3 hypersensitivity. The distribution of facial numbness was unchanged. The patient recovered in a neurosurgical ICU and denied poor coordination of arms or legs. His gait was mildly unsteady when discharged to home on postoperative Day 3, but this improved by his 10-day postoperative follow-up appointment. The patient was weaned from oxycodone to hydrocodone at that appointment, and he was able to stop taking any pain medication within 7 weeks of his surgery. He continued taking gabapentin,
which he was taking prior to surgery. At the 8-week follow-up appointment, he had no facial pain at all on the right side and no gait ataxia. At the 6-month follow-up appointment, he continued to be without right-sided facial pain, only reporting continued numbness to light touch in the right V1 and V2 distributions. One year after surgery, the patient continued to be free of facial pain. He reported an active lifestyle and had no limitations in activities of daily living. When asked if, in hindsight, he would have decided to have this surgery, he emphatically said that he would choose this surgery.

Discussion

Among patients with chronic neuropathic pain, it is estimated that 60%–70% obtain less than 50% relief from their pain by using medications. The literature regarding second-line surgical interventions for facial anesthesia dolorosa is sparse. Neurmodulation by motor cortex stimulation is advantageous because it is less invasive than other surgical options. Brown and Pilitsis reported that 8 of 10 patients with neuropathic facial pain had more than a 50% reduction in their pain initially after placement of a motor cortex stimulator; however, their study specifically excluded patients with documented anesthesia dolorosa. Raslan et al. reported that 3 of 3 patients with trigeminal deafferentation pain responded poorly to motor cortex stimulation, 1 of whom was diagnosed with anesthesia dolorosa.

Another neuromodulatory surgery considered for neuropathic facial pain is deep brain stimulation. Rasche et al. reported little or no relief of pain in 3 of 6 patients treated with ventral posterior medial nucleus/periventricular gray deep brain stimulation for facial dysesthesia dolorosa. Broggi et al. reported failed relief of atypical facial pain after posterior hypothalamus deep brain stimulation in 3 of 3 patients, among whom 1 patient was described in a way consistent with a diagnosis of anesthesia dolorosa following a history of transmandibular tumor resection.

Nucleus caudalis lesioning is a neuroablative approach that has shown greater efficacy for facial anesthesia dolorosa pain relief than that obtained by neuromodulatory implants. Nashold and Rossitch reported that 2 of 3 patients with trigeminal anesthesia dolorosa and 7 of 8 patients with postherpetic pain resembling anesthesia dolorosa had excellent or good pain relief following nucleus caudalis DREZ lesioning. Gorecki and Nashold summarized the surgical results at Duke University. With follow-up data only available for 11 of 14 patients treated for anesthesia dolorosa, 7 reported excellent or good relief. Bullard and Nashold reported that 6 of 8 patients with postsurgical trigeminal dysesthesia had excellent or good pain relief following this surgery. Single-lesion percutaneous trigeminal tractotomy-nucleotomy was reported by Kanpolat et al. for 1 patient with anesthesia dolorosa, but pain relief was not achieved.

It is important to consider the complications that can occur with this invasive procedure. In early use of the nucleus caudalis DREZ surgery, straight, uninsulated electrodes were used, and lesions were made both above and below the level of the obex. This surgical technique resulted in a 90% incidence of ataxia involving either the ipsilateral arm or leg. This complication was attributed to injury of the dorsal spinocerebellar tract, which courses superficially to the anterior aspect of the spinal trigeminal tract at the level of the obex and above. It is also possible that the straight, uninsulated electrode was placed through the fasciculus cuneatus, injuring fibers of the cuneocerebellar tract (which travels within the fasciculus cuneatus) and resulting in arm ataxia. A third possibility is that the electrode was placed too deep, causing injury to corticospinal fibers and resulting in weakness perceived as ataxia. Young et al. described using insulated electrodes, and Nashold et al. described using angled insulated electrodes. Both advances resulted in a decreased incidence of ataxia, with a 33% incidence of ataxia reported in the latter study.

Technique was also adjusted so that lesions were only made below the obex, avoiding direct injury to the dorsal spinocerebellar tract. In current practice, if ataxia develops, it generally resolves within 10 days of surgery. Care is taken to use short electrodes to make small focused lesions in the nucleus caudalis that match cadaveric measurements of the nucleus (Fig. 4). Small lesions may predispose patients to pain recurrence due to incomplete lesioning of the nucleus, but it is our belief that small, precisely placed lesions afford the patient safety from injury of neighboring brainstem structures. Other poten-
tial complications of this surgery are those related to the exposure, including the potential for CSF leakage and wound infection.  

Conclusions

This case report describes the complete and lasting pain relief brought to a patient with facial anesthesia dolorosa who underwent nucleus caudalis DREZ surgery. The surgery has rare indications, among which is facial anesthesia dolorosa. Because of the potential risk for serious injury to neighboring structures within the brainstem, it should only be performed by neurosurgeons who have special training for this procedure. Because of the rarity of facial anesthesia dolorosa, the current evidence base supporting the use of this surgery for this indication is limited to case reports and case series. The literature on anesthesia dolorosa would certainly benefit from larger prospective studies of this procedure that use validated measures of pain intensity and quality of life.

Disclosure

The authors have no financial disclosures related to the surgery. Both authors assisted with the dimension design of the manufactured electrodes used in this surgery, but neither author holds patent rights or receives royalties for their sales.

Author contributions to the study and manuscript preparation include the following. Conception and design: both authors. Acquisition of data: both authors. Analysis and interpretation of data: both authors. Drafting the article: Sandwell. Critically revising the article: both authors. Reviewed submitted version of manuscript: both authors. Approved the final version of the manuscript on behalf of both authors: Sandwell. Administrative/technical/material support: Sandwell. Study supervision: Sandwell.

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