

Orofacial Complex Regional Pain Syndrome: A Review of Case Reports

- 

Aila Malik, MD, MSPH
Department of PM&R, University of Texas Health Science Center at Houston, Houston, TX.
- 

Ryan D' Souza, MD
Department of Anesthesiology and Perioperative Medicine, Mayo Clinic, Rochester, MN.
- 

Jay Karri, MD
Department of Orthopedic Surgery, University of Maryland School of Medicine, Baltimore, MD.
- 

Saba Javed, MD
Department of Pain Medicine, University of Texas MD Anderson Cancer Center, Houston, TX.

OBJECTIVE

- We present an evidence-based narrative comprised of prior reports of complex regional pain syndrome (CRPS) with face and neck involvement with an additional focus on evaluation and treatment.

METHODS

- A comprehensive search of relevant literature was conducted in the MEDLINE (PubMed) database using the keywords “face”, “facial pain”, “complex regional pain syndrome” and “trigeminal neuralgia”. Case reports of CRPS affecting the orofacial region published between January 1, 2010, and December 31, 2023, were reviewed and analyzed with regard to disease presentation, diagnosis, and management.

RESULTS

- Our literature review identified ten cases of CRPS with face and neck involvement (Table 1).
- Orofacial CRPS typically followed head and neck trauma and dental procedures, with presumed injury to the trigeminal nerve. In most cases, the predominant complaints of burning pain, hyperalgesia, and allodynia in the facial region were accompanied by skin color, temperature, and trophic changes.
- In some patients, ocular manifestations including periorbital edema, ptosis, persistent eye pain, and visual abnormalities were described.
- Conservative management with physiotherapy and desensitization techniques provided favorable outcomes in three patients.
- A series of stellate ganglion blocks were most frequently utilized to treat orofacial CRPS with significant pain relief noted in four patients lasting from three months to two years.

DISCUSSION

- While the authors of these case reports have designated a diagnosis of orofacial CRPS, it should be noted the International Classification of Orofacial Pain published by the International Headache Society does not recognize CRPS with face and neck involvement as an orofacial pain syndrome.¹
- Similar to CRPS presentations following injury to an extremity, patients with head and neck involvement report neuropathic pain accompanied by signs and symptoms of autonomic dysfunction.
- Sympathetic hyperactivity has been implicated in the pathogenesis of CRPS, postulated to result from increased expression of sympathetic receptors on nociceptive nerves following tissue damage.²
- The positive response to sympathetic blockade with local anesthetic and sympatholytic agents such as clonidine further suggests a component of sympathetically mediated pain.^{3,4}
- The Budapest Criteria, a widely recognized diagnostic criteria for this condition has not been validated for CRPS affecting the orofacial region.⁵
- Given the location of orofacial CRPS, a comprehensive diagnostic workup may be required to differentiate between dental, musculoskeletal, neurological, vascular, auditory, and ocular pathology.
- A comprehensive multidisciplinary treatment approach should be initiated focusing on improving functional outcomes and inclusive of frequent patient education and reassurance.

REFERENCES

- Pigg M, Nixdorf DR, Law AS, et al. New International Classification of Orofacial Pain: What Is in It For Endodontists? Journal of Endodontics. 2021;47(3):345-357. doi:10.1016/j.joen.2020.12.002
- Misidou C, Papagoras C. Complex Regional Pain Syndrome: An update. Mediterranean Journal of Rheumatology. 2019;30(1):16-25.
- Gungor S, Aiyer R, Baykoca B. Sympathetic blocks for the treatment of complex regional pain syndrome. Medicine (Baltimore). 2018;97(19):e0705. doi:10.1097/MD.00000000000010705
- Basha AKMM, Simry HAM, Abdelbar AE, Sabry H, Raslan AM. Outcome of Surgical Treatments of Chronic Pain Caused by Trigeminal Neuropathy. World neurosurgery. 2023;170:e57-e69. doi:10.1016/j.wneu.2022.10.057
- Harden RN, Bruhl S, Perez RSGM, et al. Validation of proposed diagnostic criteria (the “Budapest Criteria”) for Complex Regional Pain Syndrome. Pain. 2010;150(2):268-274. doi:10.1016/j.pain.2010.04.030



Table 1. Case Reports of Complex Regional Pain Syndrome affecting the Orofacial Region

Author, Year Published	Patient Age, Sex	Etiology	Symptoms	Signs	Treatment	Results
Sakamoto et al, 2010	68, M	Ipsilateral trigeminal nerve block for trigeminal neuralgia	Right-sided facial pain with vasomotor dysfunction	Right-sided facial tactile allodynia, trophic changes of right face and ipsilateral tongue	Ketamine infusions every two to eight wk.	Pain gradually diminished, ipsilateral tongue remained atrophied
Heir et al, 2012	Case 1: 56, F Case 2: 36, F	Case 1: Apicoectomy Case 2: Endodontic procedure of a maxillary left molar tooth	Case 1: Pain in the right CN-V2 division with spread to the right upper extremity, skin color changes of right ear and hand Case 2: Pain in left maxillary teeth with spread to ipsilateral face, worse with exposure to cold	Case 1: Hypoesthesia of right CN-V2 division, right hand cooler to touch and darker appearing compared to left side Case 2: Left face hyperalgesia and skin color changes	Case 1: Two consecutive stellate ganglion blocks one month apart Case 2: Stellate ganglion block	Case 1: 65%-70% reduction in facial pain, at six mos. follow-up, VAS score improved from 9/10 to 3/10 Case 2: Complete resolution of pain, patient pain free at two yr. follow-up
Goenka et al, 2014	13, F	Snowball injury	Left-sided facial pain and swelling with skin color and temperature changes	Left-sided facial tactile allodynia, trophic skin and hair changes, partial ptosis	Physiotherapy with a focus on desensitization, behavioral therapy, frequent patient reassurance	Significant clinical improvement over a one-yr. period
Parkitny et al, 2016	23, F	Left eyelid laceration	Left-sided facial pain and tactile allodynia with associated social phobia	Left-sided facial tactile allodynia, swelling, increased skin temperature, hyperhidrosis, impaired hemifacial motor control and hemi spatial neglect	Graded motor imagery and desensitization program	At four mos. patient no longer met diagnostic criteria for CRPS. At 12 mos. ongoing pain but restoration of normal eyelid and ocular function
Arjmand et al, 2017	25, F	No inciting factor	Right periorbital edema, pain with extraocular movements	Right periorbital area hyperalgesia, skin discoloration, and warmth	IV methylprednisone followed by daily oral prednisone	Significant improvement at one wk., complete resolution of symptoms at six mos. follow-up
Lee et al, 2017	50, F	Spread from bilateral upper and lower extremity CRPS	Pain in all teeth and bilateral orofacial hypersensitivity	Bilateral (left>right) facial tactile allodynia, hyperalgesia, and increased sweating	Series of three stellate ganglion blocks combined with gabapentin	Significant improvement in orofacial symptoms over several mos.
González-Cantero et al, 2018	13, M	Tennis ball injury	Left-sided facial pain and swelling with redness and burning sensation	Left-sided facial erythema, swelling, and allodynia, impairment of left musculus frontalis	Outpatient PT and home exercise program focused on desensitization and use of affected area	Complete resolution of symptoms at three wk., no recurrence in nine mos.
Kalladka et al, 2020	39, F	Dental extraction	Left-sided facial pain worse with emotional stress and cold exposure, allodynia, skin color and temperature changes	Diffuse swelling, redness, and increased skin pigmentation in the left maxillary region	Two consecutive stellate ganglion blocks combined with clonidine (0.1 mg daily)	90% pain relief, sustained at three mos. follow up
Soifer et al, 2021	18, F	Five-year history right lower extremity CRPS with spread to bilateral upper extremities and right face	Recurrent facial pain in right eye, cheek, and mouth	Reduced visual acuity in right eye with corneal opacification and development of persistent neurotrophic keratopathy (NK)	CRPS treatment: pregabalin, amitriptyline, oxcarbazepine, lumbar sympathetic blockades NK treatment: superficial keratectomy with placement of an amniotic membrane graft, cenegermin	No recurrence of neurotrophic manifestations or neuropathic facial symptoms following the procedure



Orofacial CRPS remains an under-treated condition, and further recognition is needed to standardize management protocol ultimately leading to its inclusion in societal guidelines for facial pain and headaches.