Case Report

Complex regional pain syndrome Type II (causalgia) in head neck region: A case report

Sandip Ghose, Debasish Pramanick

From Department of Oral Pathology, Dr. R. Ahmed Dental College and Hospital, Kolkata, India

Correspondence to: Sandip Ghose, Assistant Professor, Departmentt of Oral Pathology, Dr. R. Ahmed Dental College and Hospital, Kolkata - 700091, India. E-mail: sanindra1967@gmail.com

Received – 13 May 2017 Initial Review – 18 June 2017 Published Online – 19 July 2017

ABSTRACT

Complex regional pain syndrome (CRPS) with localized erythema and hyperthermia of skin can be a perplexing clinical situation and has created varied diagnostic dilemmas for clinicians. Various cases of CRPS in the extremities have been documented, but in the head neck region, it is quite uncommon and only 13 cases have been reported since 1947 and here lies the importance of reporting the case. A 30-year-old female reported to the OPD with complain of severe pain, occasional swelling, change in overlying skin temperature, and color in left lower one-third region of face near mandibular angle since 2½ years. She had a trauma to that region 3 years back. The history and clinical examination revealed that the patient met "Budapest Clinical Criteria" for diagnosis of CRPS Type II and was treated with amitriptyline, pregabalin, methylcobalamine, steroids, and other supportive treatments. The patient showed marked improvement in pain control. Therefore, history and clinical examination and early diagnosis are crucial for the better treatment outcome in CRPS Type II.

Key words: *Causalgia, Complex regional pain syndrome, Head-neck region*

Complex regional pain syndrome (CRPS) is one of the rare causes of deep-seated facial pain encountered in clinical dental practice. CRPS Type II, previously termed as causalgia, was first demonstrated by Mitchell et al., [1] and Mitchell [2] in 1867 while reporting cases of severe burning pain associated with erythema and localized hyperthermia over the skin of the soldiers who had sustained nerve injuries in those areas previously during the American Civil War. "International Association for Study of Pain" describes CRPS as a syndrome characterized by a continuing (spontaneous and/or evoked) regional pain that is seemingly disproportionate in time or degree to the usual course of pain after trauma or other lesion. CRPS are two variants - CRPS Type I, which is not associated with nerve injury and CRPS Type II, which is associated with nerve injury.

Clinical findings in CRPS Type II include the combination of different signs and symptoms such as continuing regional pain, temperature asymmetry, skin color asymmetry, hyperalgesia, sweating asymmetry, edema, and decreased range of motion. Pathophysiology of CRPS is not fully understood, but various attempts have been made to explain the disease process. Damage to the peripheral nerve following trauma causes neurobiological changes in various components of central and peripheral nervous systems which are considered as a cause of nociception stimulation without any actual noxious stimuli. In the zone of previous neural injury, nerve rewiring may also create an altered neurophysiological state which interprets non-nociceptive inputs as noxious stimuli. This may partly explain the hyperalgesia in CRPS. Besides these, there may be a short-circuiting phenomenon following trauma due to synapse formation between collateral sympathetic efferent with afferent sensory fibers. Apart from pain, other clinical signs such as inflammation and increased blood flow have been explained in the light of alteration of sympathetic system [3]. There is also evidence of abnormal immune activation in CRPS [4].

As far as the diagnosis of CRPS is concerned, it is a challenging task as no imaging or diagnostic modalities are specific for CRPS. The most often accepted diagnostic criteria are based on the Bruehl and Harden 1999 criteria, modified at a consensus meeting in 2003 and subsequently validated and termed the Budapest Criteria [5]. CRPS Type II is mostly reported to have been occurred in the extremities, and available reports in the head neck region are rare. Marcello explained about 13 reported cases of causalgia in the head neck region in his review [3].

CASE REPORT

A 30-year-old female patient reported to the OPD of the Department of Oral Pathology with complaints of pain, skin color changes, and occasional swelling over left lower one-third region of the face often extending into ear and neck for the past 3 years. Pain was spontaneous in onset and persistent all along the day with certain episodes of acute exacerbation. The patient also reported about the local increase in temperature and appearance of redness in the skin overlying the left angle of the mandible (Figure 1). The temperature and erythema increases with acute exacerbation of pain which leads to decreased movement in left

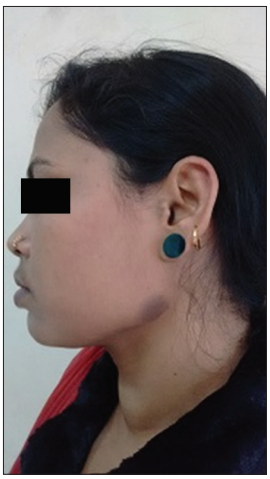

Figure 1: Lateral profile of the patient showing erythema near left mandibular angle region

side of lower one-third of face and neck region. All these above symptoms started only after a traumatic injury that the patient had sustained in left side of face and neck region 3 years back. The medical history was nil of note.

Before performing local examination, generalized systemic examination was performed which included examination of cardiovascular system, respiratory system, gastrointestinal system, and neurological system. Accepting some locally altered neurological findings (discussed later), no abnormality was detected during generalized systemic examination. Vitals were recorded and found to be within normal limits.

Local clinical examination revealed tenderness with increased temperature of the overlying skin near left angle of the mandible. An erythematous area was also detected near the skin over the aforesaid area. Quantitative sensory test was done to measure subjective responses to superficial stimulation and collect information regarding peripheral nerve function of myelinated and unmyelinated afferent fibers. During performing the test, tactile, pressure, thermal, and noxious stimuli were given to the said area and response was found to be altered when compared with its normal counterpart. Sympathetic nerve block test by injecting local anesthetic in the sympathetic ganglion was avoided because of its adverse effects ranging from Horner's syndrome to recurrent laryngeal nerve injury (resulting in hoarseness) and iatrogenic blockade of phrenic nerve (resulting in shortness of breath).

Radiological investigations in the form of orthopantomogram of jaws as well as intraoral periapical radiograph of 36 were done to rule out any bony lesion and secondary caries on 36, respectively. After ruling out any odontogenic infections, intrabony lesions, neuralgic pain, and temporomandibular joint disorders, the diagnosis of CRPS Type II was established using "Budapest Clinical Criteria" in the lower one-third of face near left mandibular angle region. Diagnosis of CRPS was established as the patient demonstrated disproportionate pain, hyperalgesia; temperature asymmetry, skin changes, edema, and restricted movement following a traumatic injury.

The patient was treated with amitriptyline 20 mg [6] once daily along with the combination drug of (pregabalin 75 mg + methylcobalamine 750 mg) [7] for 2 weeks. At 2-week follow-up, the pain subsided to some extent as well as the erythematous area of skin faded. The patient was asked to continue the same medications for next 2 weeks. At a recall visit on 4th week, the patient is still complaining of pain. Hence, oral steroids [8] (prednisolone 10 mg thrice daily) were started together with the continuation of previously advised drugs with an aim for complete relief of pain. In the next recall visits at 15 days interval, the patient was doing well with the significant decrease in pain.

DISCUSSION

As described earlier, diagnosis of CRPS is quiet cumbersome for the doctors because no laboratory tests are specific for its diagnosis. Therefore, Budapest clinical criteria [5] are widely accepted for the diagnosis of the disease which includes:

i. Continuing pain disproportionate to any inciting event.
ii. Must report at least one symptom in three of the four following categories:
    (a) Sensory: Hyperalgesia and/or allodynia.
    (b) Vasomotor: Temperature asymmetry and/or skin color changes and/or skin color asymmetry.
    (c) Sudomotor or edema: Edema and/or sweating asymmetry.
    (d) Motor or trophic: Decreased range of motion and/or motor dysfunction (weakness, tremor, dystonia) and/or trophic changes (nail, hair, and skin).
iii. Must display at least one sign at the time of evaluation in two or more of the following categories:
    (a) Sensory: Hyperalgesia (to pin prick) and/or allodynia (to light touch, deep somatic pressure, and joint movement),
    (b) Vasomotor: Temperature asymmetry and/or skin color changes and/or skin color asymmetry,
    (c) Sudomotor or edema: Edema and/or sweating changes and/or sweating asymmetry,
    (d) Motor or trophic: Decreased range of motion and/or motor dysfunction (weakness, tremor, and dystonia) and/or trophic changes (nail, hair, and skin).
iv. There is no other diagnosis that better explains the signs and symptoms.

Although CRPS is very uncommon in the head neck region and only 13 cases have been reported since 1947 [3], still the nature of the disease process needs to be understood to prevent the misdiagnosis. In fact, the exact pathophysiology of CRPS is still to be understood, but it can be interpreted as exaggeration of physiological response due to misprocessing as well as misinterpretation of sensory information [9]. Trauma is the most common precipitating factor accounting for almost 80% cases [10]. Regional pain following a trauma with hyperesthesia, temperature asymmetry, changes in skin color, restricted movement, edema, and hyperhidrosis are the features suggestive CRPS Type II. It can occur in all age group and mostly involves the extremities.

A wide range of treatment measures have been documented for treatment of CRPS with an aim to relieve pain and allow early mobilization. CRPS patients who are diagnosed early responds to treatment well in comparison to those having long standing duration of the disease [11]. Some patients with early CRPS may, however, have spontaneous resolution of their disease [12]. Treatment includes the use of various drugs such as amitriptyline, pregabalin, methylcobalamine, steroids and lastly, stellate ganglion block. It has already been documented that these drugs provide a good result to the patients in controlling pain and hypersensitivity.

As far as the findings of the previously reported cases of causalgia in the head and neck region are concerned, it has been found that most of the patients with symptoms of causalgia had a definite history of trauma. Earlier patients were treated with cervical sympathetectomy; later sympathetic ganglion block was given. These treatments measure resulted in disease-free state for a period of more than 3 months. However, some patients who were treated with methylprednisolone had complete relief of symptoms within 6 days and were disease-free for more than 3 months [3].

CONCLUSION

Sound knowledge about the disease helps in early diagnosis of the case and management should be rendered as soon as possible to have good treatment outcome. In the differential diagnosis of orofacial pain, the possibility of CRPS should be included while managing the patients of atypical facial pain.

REFERENCES

1. Mitchell S, Morehouse C, Keane W. Gunshot Wounds and Other Injuries of the Nerves. Philadelphia, PA: Lippincott; 1864.
2. Mitchell S. Injuries of Nerves and Their Consequences. 2nd ed. New York: Dover; 1965.
3. Melis M, Zawawi K, al-Badawi E, Lobo Lobo S, Mehta N. Complex regional pain syndrome in the head and neck: A review of the literature. J Orofac Pain. 2002;16(2):93-104.
4. Calvo M, Dawes JM, Bennett DL. The role of the immune system in the generation of neuropathic pain. Lancet Neurol. 2012;11(7):629-42.
5. Harden RN, Bruehl S, Perez RS, Birklein F, Marinus J, Maihofner C, et al. Validation of proposed diagnostic criteria (the 'Budapest Criteria') for complex regional pain syndrome. Pain. 2010;150(2):268-74.
6. Wheeler AH, Berman SA. Complex regional pain syndromes treatment and management: Medscape. Drug Dis. 2016.
7. Yabuki K, Onda A, Hamba M. Complex Regional Pain Syndrome Type I after Lymph Node Biopsy of the Neck: A Case Report; Research Gate, January; 2010. DOI: 10.5631/jibirin.103.65.
8. Glick EN. Reflex dystrophy (algoneurodystrophy): Results of treatment by corticosteroids. Rheumatol Rehabil. 1973;12(2):84-8.
9. Lindelfeld TN, Bach BR Jr, Wojtys EM. Reflex sympathetic dystrophy and pain dysfunction in the lower extremity. J Bone Joint Sur Am. 1996;78:1936-44.
10. Smith DL, Campbell SM. Reflex sympathetic dystrophy syndrome. Diagnosis and management. West J Med. 1987;147(3):342-5.
11. Kurvers HA. Reflex sympathetic dystrophy: Facts and hypotheses. Vasc Med. 1998;3(3):207-14.
12. Tietjen R. Reflex sympathetic dystrophy of the knee. Clin Orthop Relat Res. 1986;209:234-43.

Funding: None; Conflict of Interest: None Stated.

How to cite this article: Ghose S, Pramanick D. Complex regional pain syndrome Type II (causalgia) in head neck region: A case report. Indian J Case Reports. 2017;3(3):161-163.