

Case Report

A Curative Treatment Option for Complex Regional Pain Syndrome (CRPS) Type I: Dorsal Root Entry Zone Operation (Report of Two Cases)

Running Title: A Curative Treatment Option for CRPS-I

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Abstract

Background: Complex Regional Pain Syndrome Type I (CRPS-I) is a debated health problem concerning its pathophysiology and treatment strategies. Two cases of CRPS-I are presented herein. Various procedures had been performed in both cases with unsatisfactory outcomes, and they were finally treated with Dorsal Root Entry Zone (DREZ) lesioning.

Clinical Presentation: A 12-year-old boy and a 35-year-old woman were diagnosed with CRPS-I at different times. They had previously undergone various types of interventions with no success. After one year of follow-up and observation, DREZ lesioning operation was performed. Afterwards, both cases had transient lower extremity ataxia. The first case was followed for 60 months with no recurrence and total cure. The second case was pain-free until the 6th month, when she required psychological support; she was followed for 33 months with partial satisfactory outcome.

Conclusion: Although not a first-line option, DREZ lesioning procedure can be chosen and may be a curative option in selected cases of CRPS-I who are unresponsive to conventional therapies.

Keywords: CRPS-I, DREZ, pain, reflex sympathetic dystrophy

Introduction

Complex Regional Pain Syndrome (CRPS) is a group of disorders presenting with regional pain, edema, skin alterations, vascular changes, and other symptoms associated with impaired neural and vasomotor activity.

Ambroise Pare first mentioned a likelihood disorder in 1634. Subsequently, Claude Bernard mentioned an association of pain with sympathetic nervous system involvement, and Weir Mitchell, one of Bernard's students, reported the first description of the syndrome in the 1860's (18). Following this description, several nomenclatures were used (6,29). Finally, the International Association for the Study of Pain (IASP) concluded this clinical syndrome's name as CRPS and classified it into two groups (Type I and II) according to the existence or not of nerve injury (9,17,28).

Although the potential causes of this clinical syndrome are well recognized, the precise pathophysiology has not been identified comprehensibly. As a result of this uncertainty regarding the pathology, a curative treatment strategy for this debilitating disease is lacking (13,20). Available treatment options include conventional pharmacological and physical therapy, nerve blocks, transcutaneous electrical nerve stimulation, both chemical and surgical sympathectomies, and spinal cord stimulation (SCS). Destructive procedures such as dorsal root entry zone (DREZ) lesioning should be considered as a treatment option in intractable cases (10).

We report two cases of CRPS-I who underwent various treatment procedures with unsatisfactory outcome who were finally treated with DREZ lesioning.

Clinical Presentation

Patient 1

A 12-year-old boy presented with a 19-month history of severe pain in his right hand after a blunt trauma to his hand (caused by a ball). The pain was sharp and sometimes throbbing. Light touch and minimal temperature changes increased the pain as expected. The pain also restricted limb movements and limited the patient's daily activities. The visual analogue scale (VAS) score of the patient was 9/10. The Karnofsky performance scale was 50, and the Beck Depression Inventory score was 38. The physical examination revealed allodynia and hyperalgesia of the affected limb. When compared with the left side, the limb was colder, edematous and had a restricted range of motion (ROM). Peripheral pulses were palpable and no trophic changes were observed. Radiological interventions and nerve conduction studies revealed no abnormalities. Various types of procedures had been performed before his current admission, including stellate ganglion block, transcutaneous electrical nerve stimulation, morphine pump combined with oral opioids, and two orthopedic interventions. Because of the health insurance subsidization policy, the SCS procedure could not be done. Therefore, spinal cord DREZ operation was selected to achieve the pain relief. While preparing the patient for the surgical procedure, we obtained the diametric measurements of the affected spinal cord by computed tomographic scan. With the help of these dimensions, the depth of the electrodes was arranged. The patient was placed in the prone position under general anesthesia. A median vertical skin incision was made. C3-T1 laminoplasty was performed. The dura was opened vertically just in the line of the roots of the affected side with the help of the surgical microscope. The

arachnoid was opened directly over the DREZ with the help of micro nerve hooks or micro scissors. Approximately 25 DREZ lesions were made using the DREZ electrodes, limited sulcotomy, and bipolar coagulation. Standard radiofrequency (RF) lesions were made, with the parameters of 75° to 80°C and 15 seconds for each lesion. The dura was then sutured watertight with atraumatic silk, laminas were replaced, and closure was completed in order of anatomic layers. Immediately after the operation, the VAS score decreased to 0/10. The Karnofsky performance scale improved to 100, and the Beck Depression Inventory score had reduced to 2 at the 6th month after the procedure. The patient experienced transient ipsilateral lower extremity ataxia, which improved in 2 months. He was able to move his arm and hand easily and was able to use his fingers. He was discharged on the 3rd day and returned to school, which he had been unable to attend for nearly 1 year because of pain. He has been followed for 60 months with no recurrence and no complications.

Patient 2

A 35-year-old female presented with a 14-month history of severe pain in her right hand after a blunt trauma to the hand. The pain was sharp and pulsating. Even light touch and minimal temperature changes increased the pain. The pain also restricted limb movements and limited her ability to conduct daily activities; in fact, the patient was completely incapacitated. The VAS score of the patient was 10/10. The Karnofsky performance scale was 60, and the Beck Depression Inventory score was 40. The physical examination revealed allodynia and hyperalgesia of the affected limb. When

compared with the left side, the limb was colder and had a restricted ROM. Peripheral pulses were palpable, but there were visible trophic changes and muscle atrophies in her right arm. Radiological interventions and nerve conduction studies revealed no abnormalities. Before her current admission, the patient had undergone various types of procedures, including stellate ganglion block (twice), morphine pump combined with oral opioids, axillary block, and peripheral nerve stimulation. Because of the health insurance subsidization policy, SCS could not be performed. C5-C7 partial laminectomy was performed, and microsurgical DREZ lesioning was performed to the C5-C6 and C7 segments as described previously. Approximately 25 lesions were made. Immediately after the operation, the VAS score decreased to 1/10 and this level was sustained for 6 months. The Karnofsky performance scale improved to 90 and the Beck Depression Inventory score had reduced to 20 at the 6th month after the procedure. Thereafter, her pain recurred, with a VAS score of 6. The patient was referred to a psychiatry clinic, and anti-depressive treatment was administered for both the intractable pain and depression. She also experienced transient ipsilateral lower extremity ataxia, which improved in 3 months. Postoperative 8th month axial T2-weighted magnetic resonance image of the patient is demonstrated in Figure 1. She has been followed for 33 months with partial satisfactory pain relief (VAS score: 4). Currently, her complaints are partially under control with Pregabalin (Lyrica®, 150 mg) and Risperidone (Rileptid®, 2 mg).

Discussion

The pathophysiology of CRPS has not been well identified in the literature, and controversies remain in terms of the management strategies for this troublesome syndrome. A survey of CRPS patients shows that more than 70% of patients report "constant" pain, 74% report having to stop daily activities due to pain, more than 84% report sleep disturbance due to pain, more than 30% report that pain interferes with their marriage and family life, and 33% report that pain interferes with their sexual activity (1). It has been estimated that CRPS costs over \$100,000 annually in the United States per one case, and the social and economic cost of CRPS may be substantially higher than this because of the associated long-term morbidity (24). Different treatment modalities, including medications, transcutaneous electrical nerve stimulations, SCS, peripheral nerve blocks, and sympathectomies have been unsatisfactory in relieving the pain totally or in regaining limb functions and acceptable quality of life (8,11,13,15,21). Additionally, there are a lack of randomized controlled trials concerning the efficacy of the aforementioned treatment strategies. Thus, while dealing with this syndrome, treatment usually depends on the clinician's experience.

Gabapentin should be considered as a first-line therapy for treating CRPS. Mellick et al., in a study of 6 patients, demonstrated the efficacy of gabapentin for treating CRPS (16). Verbal pain scores were decreased approximately 60-100%. Moreover, Tan et al. achieved similar improvement in pain scores with gabapentin in a study enrolling 21 patients (30). Although the benefit of tricyclic antidepressants (TCAs) could not be documented in contrast to other neuropathic pain syndromes, these medications should be considered as an adjuvant therapy for treating CRPS. Non-steroidal anti-inflammatory drugs, free radical scavengers such as dimethylsulfoxide (DMSO) and N-

acetylcysteine (NAC), bisphosphonates, oxycodone, and morphine should also be evaluated as a medical treatment option (4,22,25,31). Zuniga et al. demonstrated the efficacy of chronic intrathecal infusion of baclofen in a study of two medically intractable CRPS patients (32).

Sympathetic nerve blockade should be considered as a treatment option for treating CRPS. This method can be done both chemically and surgically. It could also assist as a diagnostic tool. Nevertheless, the potency of this intervention is still uncertain (3,7).

Since SCS is safe, reversible and minimally invasive, it should be recommended as a possible treatment for CRPS. Although the potential effect of SCS for reducing pain when used for CRPS is well described, the outcome in terms of improving the quality of life remains arguable (5,12,21).

CRPS is associated with both psychological and psychosocial negative outcomes, but there is no direct relationship between psychological diseases and the development and maintenance of CRPS-I (2,14). The first case did not present with any psychiatric condition but the second case was affected seriously; thus, in patients with CRPS-I, the frequency and intensity of psychotherapy can be adjusted individually according to the progression of CRPS (8).

The DREZ operation is a destruction-based lesioning operation concerning the dorsal root entry region. The operation was first described as posterior selective rhizotomy by Sindou et al. and was first performed using RF electrode systems by Nashold et al (19,26,27). Although it is an irreversible destructive procedure, the DREZ operation should be considered as a treatment option for CRPS-I in selected cases. Prestor

reported two cases who underwent DREZ coagulation for treatment of CRPS-I, with excellent results (23). Selection criteria should be taken into consideration, such as medically refractory cases or cases unresponsive to neuro-augmentative procedures such as SCS therapy. Cases should also be followed at least 12 months before deciding the surgery (23). In our report, Case 2 was impaired six months after the surgery; the patient improved in terms of VAS score and daily activities after neuro-psychiatric evaluation and treatment. The psychiatric condition of the patients should be examined in detail and appropriate treatment should be administered before and after the surgical intervention. We want to emphasize the necessity of supporting a procedure such as the DREZ lesioning procedure in patients suffering CRPS-I, as it may be an option after a sufficient time period of observation and especially when other methods fail to satisfactorily manage the situation.

Conclusion

Since DREZ lesioning is a destructive procedure, requires surgical experience and is associated with potential serious surgical complications such as ataxia and limb weakness, the procedure should not be considered as a first-line treatment option for CRPS-I. However, DREZ lesioning should be considered in selected cases that are unresponsive to conventional therapies and surgical therapies, including nerve blocks, morphine pumps and SCS.

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Figure Legends

Figure 1: Axial T2W MR image of the second case in the 8th postoperative month.

