

Conservative therapy for Complex Regional Pain Syndrome Type I in a paediatric patient: a case study

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Complex regional pain syndrome (CRPS) is a term that describes a variety of chronic pain conditions that are believed to result from dysfunction in the central or peripheral nervous systems. Typical features include dramatic changes in the colour and temperature of the skin over the affected limb or body part, accompanied by an intense pain which is out of proportion to the injury thought responsible. Skin sensitivity, sweating, and swelling are also commonly involved. This case study presents subjective reports of changes in pain and extremity weight bearing capacity in an 8 year-old child with Chronic Region Pain Syndrome Type I. The changes reported occurred over a 12 week conservative course of treatment which included manipulation, nutritional supplementation and rehabilitation. The patient was able to regain full control of her legs and full weight bearing after 3 weeks of treatment.

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KEY WORDS: Regional Pain Syndrome, nervous system dysfunction, manipulation

Introduction

Complex regional pain syndrome (CRPS) is a term that describes a variety of chronic pain conditions that are believed to result from dysfunction in the central or peripheral nervous systems.^{1,2,3} Typical features include dramatic changes in the colour and temperature of the

Le Syndrome douloureux régional complexe (CRPS) est un vocable qui décrit une variété de douleurs chroniques qu'on croie être le résultat d'un dysfonctionnement du système nerveux central ou périphérique. Les caractéristiques typiques comprennent des changements dramatiques dans la couleur de la peau sur la partie affectée du membre ou du corps, accompagnés d'une douleur intense, disproportionnée par rapport à la blessure qui en est à l'origine. La sensibilité de la peau, les sueurs et la tuméfaction sont en général présentes. Cette étude de cas présente des rapports subjectifs de changements dans la douleur et de la capacité extrême de supporter un poids chez un enfant de huit (8) ans souffrant du Syndrome douloureux régional complexe de type I. Les changements rapportés sont survenus sur une période de 12 sessions consécutives de traitements, qui ont compris la manipulation, un régime alimentaire enrichi et une réadaptation. Le malade a été capable de reprendre le contrôle complet de ses jambes et de se tenir debout après trois semaines de traitement.

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MOTS CLÉS : Syndrome douloureux régional complexe, dysfonctionnement du système nerveux, manipulation

skin over the affected limb or body part, accompanied by an intense pain which is out of proportion to the injury responsible.^{4,5} Allodynia, hypoalgesia, sudomotor and vasomotor disturbances such as skin sensitivity, sweating, and swelling are also commonly reported.^{6,7} Other theories propose that post-injury CRPS is caused by a triggering

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of the immune response, which leads to the characteristic inflammatory symptoms of redness, warmth, and swelling in the affected area.⁸

Reports of CRPS in chiropractic patients are very rare. A literature search revealed only two reported cases in adults^{9,10} with no reported cases in children. This seems surprising since population studies in school children indicate chronic or recurrent musculoskeletal pain is quite common in presentations to paediatricians and paediatric rheumatologists.¹¹ The most commonly reported presenting complaints in children to the above specialties are back pain (20%), limb pain (16%) and fibromyalgia (6%).^{12,13}

The unreported nature of CRPS by chiropractors may be related to the fact that in the early stages of the condition there are no specific laboratory tests available to the chiropractor that effectively help diagnose the condition. In addition, in children the radiological and bone scan findings commonly present in adults are often absent.^{11,14,15,16} In all likelihood, CRPS does not have a single cause, but is rather the result of multiple causes that produce similar symptoms.

There are no scientifically validated treatment for CRPS, however manipulation, mobilization¹⁷ and vitamin supplementation with antioxidants¹⁸ has shown the greatest promise to date. This case presents the changes in subjective pain and weight bearing during a 12 week course of conservative management of an 8 year old female patient with CRPS type I.

Presentation History

The patient, an eight year-old female, underwent removal of ear grommets with bilateral myringoplasties under general anaesthesia. She spent approximately 25 minutes in recovery before being transferred to the ward. The patient received paracetamol as a premedication and inhalation anaesthetic with nitrous oxide, oxygen and sevoflurane during the procedure. The recovery nurse noted that a rash which was present preoperatively on her arms and neck had become more florid during the recovery period. During discharge procedures she complained of mild weakness in her legs but was discharged regardless.

Two days later, the patient was seen in the emergency room (ER) with loss of power and a diagnosis of viral myalgia. The following day she again presented to the ER following a hyperventilation episode, and painful

joints. She was admitted to the hospital and was discharged one month later following an extensive investigation, including MRI on both legs and her low back, multiple blood and neurological testing with the diagnosis of complex regional pain syndrome type 1. Several weeks later the patient presented for evaluation of unbearable left leg and foot pain with increased sensation of touch and pressure.

On presentation, the patient was not bearing weight on either of her legs, and was being carried by her mother. The patient experienced pain on both passive and active flexion of her legs and in all ranges of motion of her left ankle with associated limbic concomitants (moaning/groining and crying). The pain had also spread to her arms bilaterally at the time of presentation. The patient described the pain as a burning type pain, occurring in both legs but worse from the knee to the foot on the left leg. When asked to outline the area that she felt the pain the patient pointed to her entire lower legs, both anterior and posterior, bilaterally. It was noted that the area outlined by the patient did not follow the distribution of any single nerve or the common dermatomal patterns normally associated with nerve root involvement.

Past History

The patient had always been a healthy active child. She had been training for cross-country competition prior to the recent illness. She had received a vaccination for meningitis 3 months prior to the present illness with no reactions noted. Her mother was not aware of any allergies to medications, pollen or food. Prior medical testing for diabetes was negative.

Physical Examination

Visual acuity was normal in both eyes. Pupil sizes were equal bilaterally. Pupil light reflexes were fast and fatiguing on the left. Visual fields were tested by confrontation and were unremarkable. Convergence was unremarkable. Eye movements in four quadrants were normal. She could maintain eyes closed against opening pressure. Ophthalmoscopic examination revealed normal fundi bilaterally with an increased V/A ratio bilaterally. No nystagmus was noted. Otoscopic examination revealed a right swollen external auditory meatus with white exudates. The left ear canal was clear but scar tissue in posterior aspect of the ear canal was visualised. Hearing was normal in both

ears. The patient denied any ringing sensations in the ears. Facial nerve activity including smile, frown and grimace was normal. Palate elevation was equal and normal bilaterally. Tongue movements were normal and controlled. Tongue protrusion was equal and centred. Opticokinetic (OPK) testing revealed slowing of eye movements and fatigue moving from the patient's right to left. Vestibular/ocular and painful stimuli on the left side of the face in all divisions (VI, V2 and V3). Corneal reflexes were normal. Glabella tapping showed a normal habituation response. Facial nerve responses were unremarkable. Sterocleidomastoid palpation was painful bilaterally with radiations into the forehead region. Auscultation of the heart revealed a racing rhythm at 136 bpm. Respiration was shallow but within the high range of normal at 23 per minute. Examination of the lungs and abdomen were both unremarkable. Reflexes of the upper limbs and right lower limb were all within normal limits. The S1 and L4 reflexes of the left lower limb were reduced. Plantar responses were unremarkable bilaterally.

Dermatographia (sustained wheal and flare reaction following mild pressure to the skin) was present and remained exaggerated through-out the examination on the lower limbs but worse on the left lower limb. Passive movement of both lower limbs was painful but tolerated. Active movement of both lower limbs was refused due to pain. The patient described both lower limbs as feeling cold but they were normal temperature to touch. Sensation of hot and cold was within normal limits. Several areas in the patient's spine and extremities displaying altered joint motion were identified utilizing motion and static palpation, and joint play analysis.

Differential Diagnosis

It is essential to distinguish CRPS from other neuropathic and pain syndromes. Many patients with post traumatic neuropathy have pain but they do not present with the full clinical picture of CRPS. In non-CRPS neuropathy the post traumatic pain is usually contained within the territory of the involved nerve.¹⁹ Patients with post traumatic neuropathy commonly report burning pain similar to those patients with CRPS but do not display marked oedema or progressive spread of symptoms beyond the territory of the involved nerve.²² The spontaneous pain of non-CRPS neuralgia is typically superficial rather than located in the deep tissues of the involved limb. The in-

tensity of both types of pain is not dependant on the position of the extremity, but occurs spontaneously in all positions.

In acute stages (less than 6 months) of CRPS-I the affected limb is often warmer and the perfusion higher than the contralateral limb. This reverses in the chronic situation (greater than 6 months) which may lead to confusion of the diagnosis.²³ Neuropathies such as diabetic polyneuropathy can also present with spontaneous pain, skin colour changes and motor deficits. These conditions can be distinguished from CRPS by the symmetric distribution of the symptoms, blood testing and the patient history. Unilateral arterial or venous occlusive diseases or conditions can also mimic the symptoms of CRPS and should be excluded by Doppler ultrasound or on clinical grounds.

Working Diagnosis

Previous medical testing had ruled out meningitis, diabetes or tumour as differential diagnostic considerations. The working diagnosis of complex regional pain syndrome was established which was consistent with the previous medical diagnosis.

Treatment

The patient received a variety of manipulations to the joints of her lower limbs and spine. Spinal motion units and extremity joints were adjusted to correct altered motion as determined by motion palpation, static palpation and joint play analysis. Mobilization was initially utilized on her lower limbs and was gradually increased in force, speed and intensity ultimately evolving to manipulation as tolerated by the patient. Manipulations of the spine were standard diversified technique and included side posture lumbar and pelvis manipulations, anterior thoracic spine manipulations and cervical manipulations. The patient was manipulated as determined by need which initially resulted in manipulations once or twice per week for a period of 8 weeks, and every two weeks for the remaining 4 weeks of the trial. Nutritional supplementation was also instituted during the first week of treatment including omega 3 fish oils (1 gram/day equivalent), essential amino acids (recommended daily allowance), and standard multivitamins including vitamin C (recommended daily allowances).

Breathing exercises were introduced in the first week to create a breathing pattern in which inhalation exceeded

exhalation in duration and reduced her respiration rate. Rehabilitation strategies included stretching to increase function and ROM of the involved joints, education aimed at increasing both the patient’s and her mother’s understanding of her condition and endurance activities such as walking in a pool and stationary bike riding with very little tension were instituted in week two and increased as tolerated by the patient throughout the remaining course of her treatment.

Clinical Outcome At 12 Weeks

The patient regained full control of her legs and full weight bearing after 3 weeks of treatment. A one year verbal follow up revealed that the patient has returned to a virtually normal life, attending school and playing soccer. She still experiences occasional bouts of pain that may last 2–3 hours in duration following high periods of stress.

Discussion

Clinical Characteristics

In the mid 1990s, the International Association for the Study of Pain (IASP) published guidelines for the diagnosis of CRPS type I and Type II.¹⁹ CRPS type I, frequently results following tissue injury. CRPS-I is characterized by the following features: pain, which may take the form of spontaneous pain, hyperalgesia and allodynia; movement disorders, both active and passive; abnormal sympathetic regulation in the form of blood flow and sweating abnormalities; edema and trophic changes to the involved tissues (Table 1).

The term CRPS type 1 describes all patients with the above symptoms but with no underlying nerve injury. CRPS type II develops following injury to a peripheral nerve.²⁰ Patients with CRPS II experience the same symptoms as described above but their symptoms are clearly associated with a nerve injury.

Diagnostic Criteria

The current diagnosis of CRPS-I and CRPS-II are mainly based on the patient’s history and a careful and complete physical examination. There are is no diagnostic gold standard or objective test for CRPS. The International Association for the Study of Pain (IASP) diagnostic criteria describes the diagnosis of CRPS-I as follows; “*Type I*

Table 1 *Characteristic Features of CRPS Type I*

| | |
|---------------------------------|--|
| Pain | Spontaneous Not limited to any single nerve distribution Disproportionate to instigating injury Hyperalgesia Allodynia Both active and passive movement may initiate pain |
| Movement Disorders | Both active and passive movement can be affected |
| Abnormal Sympathetic Regulation | Blood flow abnormalities Inappropriate sweating Edema Trophic changes |

is a syndrome that develops after an initial noxious event. Spontaneous pain or allodynia / hyperalgesia occurs, is not limited to the territory of a single peripheral nerve, and is disproportionate to the original inciting event. There is or has been evidence of edema, skin blood flow abnormality, or abnormal sudomotor activity in the region of the pain since the inciting event. The diagnosis is excluded by the existence of conditions that would otherwise account for the degree of pain and dysfunction.”²¹

The Pathophysiology of CRPS-I

Patients with CRPS-I generally report a burning, spontaneous pain felt most prominently in the deep tissues of the distal part of the affected extremity. Experimental investigations have mainly focused on superficial pain mechanisms, sympathetically mediated or maintained pain, and abnormalities of the skin. Yet clinical observation tends to suggest that the pain of this condition is projected into the deep tissues and many patients show no change in the symptoms following sympathetic block.²⁴ As well, up to 5% of patients do not experience spontaneous pain, but rather discrete evoked pathological pain which would suggest that other underlying mechanisms are responsible for the persistence of the symptoms.²⁴ Another feature of CRPS type-I that cannot be over emphasized is that the severity of the symptoms are disproportionate to the severity of the trauma, and the symp-

toms have the tendency to spread through out the involved limb without regard to the distribution of innervation of any individual nerve. The progression of symptoms just described may occur irrespective of the type of the preceding lesion.^{25,26,27}

Typically, the pain experienced by patients with CRPS type-1 can be elicited by movements of the involved joints (deep somatic allodynia) even when these joints are not directly involved in the initial inciting injury.²¹ The spontaneous pain and various forms of allodynia experienced are thought to be generated by processes of peripheral and central sensitization.²⁸ About 50% of patients with CRPS-I also develop hypoesthesia and hypoalgesia on the affected half of the body or in the upper quadrant, ipsilateral to the effected extremity. These patients that exhibit extended sensory involvement have longer illness duration, greater pain intensity, higher frequencies of mechanical allodynia and a greater tendency to develop somatomotor system involvement than do patients that only exhibit spatially restricted sensory involvement.^{29,30} The anatomical distribution of these symptoms suggest that they may be due to changes in the functionality of the CNS that have resulted in alterations in the perception of painful as well as non-painful stimuli. Support for the existence of these changes in the CNS in both the thalamus and the cortex has been found utilizing both positron emission tomography (PET) and magnetoencephalography (MEG).^{1,2,31}

Some evidence and several authors have proposed that a certain population of patients suffering from CRPS-I also experience a degree of sympathetically maintained pain (SMP). This statement is supported by investigations that demonstrate that a certain percentage of the population of patients with CRPS will obtain pain relief with the application of sympathetic blockade drugs such as phentolamine.^{32,33} This potentially implicates sympathetic involvement in at least some portion of the pain experienced by CRPS patients.

Evidenced Based Therapy for CRPS Type I

A review of the literature of outcome studies in CRPS revealed very little consistent evidence for a therapeutic approach to treatment of CRPS in general and even less evidence when the condition occurs in children.^{6,34,35} Forouzanfar⁶, in a systematic review found limited to no evidence for the efficacy of sympathetic stellate ganglion

blockade, prednisolone administration, acupuncture and manual lymph drainage in the treatment of CRPS. The absence of an evidenced based approach to treatment of CRPS leaves clinicians in the position of having to utilize treatment approaches that;

1. have been used on other neurological conditions and may not be effective in CRPS,
2. rely on clinical experience,
3. utilise an experimental approach.

The most promising approaches to treatment in the literature include nerve stimulation techniques that activate central disinhibitory processes (TENS, Epidural spinal cord stimulation),^{36,37} deep brain stimulation of the lateral thalamus and medial lemniscus^{38,39} and functional return of joint motion of the involved limb (physical and occupational therapy, manipulation). In children, early restoration of functional joint motion seems to be particularly important in the long term control of the condition.⁴⁰ Some evidence exists that Vitamin C administration may prevent the onset of CRPS type 1.⁴¹

Summary

The pathophysiology of CRPS remains controversial. Various treatment approaches have been attempted but none have been documented as consistent or reliable in the literature. This case study documents a reduction of pain and return of functional weight-bearing in a child with CRPS-I following the application of a conservative treatment approach to therapy including manipulation and nutritional modalities. Obviously more robust studies need to be undertaken before consistent and reliable conclusions can be drawn about this approach to treatment, however, the success of symptom alleviation in this case suggests that a conservative approach to CRPS type-I may show promise in the treatment of this condition.

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