Diffuse Complex Regional Pain Syndrome in an Adolescent

A Novel Treatment Approach

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Objectives: Late diagnosis and management of complex regional pain syndrome (CRPS) can have severe consequences. We report the case of a young adolescent who failed several months of medical and physical therapy for CRPS, which was initially limited to one extremity but then spread to the remaining extremities.

Methods: At the time of admission to our hospital, she had been unable to independently perform activities of daily living for several months. We placed an epidural catheter and 2 infraclavicular catheters under general anesthesia and ran continuous infusions of local anesthetic and morphine in the epidural catheter (ropivacaine 0.1% and preservative-free morphine [20 μg/mL] at 8 mL/h) and ropivacaine 0.1% 6 mL/h in each infraclavicular catheter.

Results: Patients were started intense physical, occupational, and psychotherapeutic treatments the following day. Color and temperature of extremities normalized within 10 minutes from the time of the bolus of local anesthetic through the catheters. The patient was able to walk and use her hands 48 hours after the placement of the catheters. She was weaned off opioids during her hospital stay and was discharged home 9 days after her initial admission, and was able to walk and attend to her daily living activities.

Discussion: The extensive use of regional anesthesia techniques can greatly benefit patients with CRPS during the acute phase of the rehabilitation process, which includes appropriate physical, and occupational therapy and psychological interventions. It is critical to continue physical therapy and psychological support after discharge from the hospital.

Key Words: CRPS, regional anesthesia, pain, pediatric

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Complex regional pain syndrome (CRPS) is characterized by the presence of persistent pain that is often disproportionate to the injury, and not limited to 1 nerve distribution.[1] CRPS is usually localized in one of the extremities. Contrary to the adult population, where there is no significant clinical difference between involvements of the lower versus upper extremities or the left versus the right side of the body,[1,2] lower extremities are affected much more frequently in adolescents.[3,4] The relative rarity of this condition and the lack of familiarity of general practitioners with CRPS symptoms often lead to late diagnosis and inappropriate management with the possibility of CRPS spreading beyond the original extremity.[5]

Regional anesthesia has been used to treat pain in patients with CRPS and the results have been inconclusive.[6] Better outcomes have been demonstrated when regional anesthesia is combined with physical therapy interventions.[7] The spread of CRPS from one limb to others is not unusual[8,9] and the management of these patients can be challenging.[8] There are no reports in the literature on the prolonged use of multiple central and peripheral nerve catheters in children with diffuse CRPS to facilitate the rehabilitation process. In this case report we describe the use of multiple catheters for the management of a child with CRPS in both upper and lower extremities.

CASE PRESENTATION

This case review was approved by the Human Subjects Protection Committee at Children's Hospital Los Angeles and the informed consent was waived. A 13-year-old girl weighing 74 kg was admitted to the hospital with a diagnosis of CRPS involving her 4 extremities.

Presenting History

The patient experienced a whiplash injury 8 months before admission that resulted in severe neck pain. She was treated conservatively with physical therapy, nonsteroidal anti-inflammatory medications, and oral opioids resulting in a complete resolution of her symptoms and was taken off pain medications. Four months later she sustained another injury to her neck and left shoulder during a confrontation at school that resulted in painful swelling and cyanosis of the left hand and arm. The patient was unable to use the extremity despite taking gabapentin, ketorolac, and ibuprofen. She reported burning pain with occasional episodes of shooting pain from her elbow to the hand. Because of persistent pain, 2 weeks later she underwent a left stellate ganglion block that provided relief only for 24 hours. Within a few days both legs became swollen and purple. A month later, because of the persistent disability and lack of response to the recommended treatments, she was admitted to another facility with the diagnosis of severe CRPS of the 4 extremities. She was prescribed antidepressants and because of her lack of cooperation with physical therapy (PT) she was quickly transferred to an inpatient psychiatry unit with a diagnosis of conversion disorder. She was prescribed antidepressant, antipsychotic medications along with opioids and baclofen. None of these medications were effective. Two months after her initial admission she was discharged home on duloxetine, methadone, clonidine patch, and gabapentin. She was then followed for a few weeks in an outpatient

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pain facility for management of her medications and psychotherapy. She was referred to our program for lack of improvements.

Initial Evaluation

At the time of admission in the Chronic Rehabilitation Unit, which is located within the main hospital, she was wheelchair bound with extensions to support her 4 extremities and she needed assistance with activities of daily living. Her hands and feet were swollen and cold, with purple discoloration. She was unable to move her 4 extremities and the parents had to feed her, lift her from the chair to take her to the bathroom, or put her in bed. She was wearing shorts and a short sleeves shirt because she could not tolerate any type of clothing covering her extremities. She had significant allodynia on both hands and feet. Her medications included gabapentin 900 mg tid, clonidine 0.1 mg qD, baclofen 5 mg tid, methadone 5 mg PO bid, quetiapine 20 mg PO bid, duloxetine 40 mg qD, and lidocaine 5% patches randomly applied to her extremities.

The PT and occupational therapists (OTs) reported that the patient was able to actively grip with a light grasp bilaterally. The patient allowed some active-assisted range of motion of both legs and was able to actively flex and extend hips and knees through partial range, as well as adduct and abduct both hips. She exhibited little active movement of ankles, feet, and toes. Passive range of motion seemed limited in bilateral ankles. The psychological evaluation highlighted the presence of severe depression and anxiety as well potential family conflicts.

Interventions

The day of admission, she was taken to the operating room for placement of an epidural catheter and 2 infraclavicular catheters under general anesthesia. The epidural catheter was placed under sterile conditions at L1-L2 level. An 18 G Tuohy needle was used to identify the epidural space and a stimulating catheter (Pajunk Tsui Set; Pajunk Medical System LP, Norcross, GA) was advanced 5 cm into the space. After a negative test dose (bupivacaine 0.25% + epinephrine 1:200,000, 3 mL) the catheter was bolused with 8 mL of bupivacaine 0.1% + epinephrine 1:200,000. Two infraclavicular catheters were then placed under ultrasound guidance, in combination with nerve stimulation. An 18 G insulated needle (Pajunk, Pajunk Medical Systems LP) was used. A stimulating catheter was advanced and placed in proximity of the posterior cord of the brachial plexus. The correct placement of the catheter, distally from the axillary artery and vein was confirmed by ultrasound. Contractions were lost at 0.48 and 0.62 mA on the right and left side, respectively. The catheters were bolused with 15 mL of ropivacaine 0.15%.

An infusion of ropivacaine 0.1% and preservative-free morphine (20 mcg/mL) at 8 mL/h was started through the epidural catheter. Each infraclavicular catheter was connected to an On-Q C-bloc pump (I-Flow, Lake Forest, CA) to deliver ropivacaine (0.1%, 6 mL/h). An intravenous infusion of naloxone 0.5 mcg/kg/h, a dose shown not to reverse analgesia,[9] was started to minimize the potential side-effects of epidural morphine, including nausea, vomiting and pruritus. The color and temperature of hands and feet normalized within 10 minutes from the bolus of local anesthetic and remained normal for the rest of the hospitalization.

The patient started an intense rehabilitation program the next day, which included individual and group OT, individual and group PT, psychotherapy, and daily evaluation by the psychiatrist (Table 1). The family was allowed to spend only 2 hours a day with the patient at dinner time. This was done to limit the negative impact of potentially complex family dynamics on the patient's recuperation. The patient also had no access to a cell phone during the day or the internet.

The PT and OT interventions included jumping, running, stairs, weight-bearing exercises, writing desensitization using massage, and contrast baths with hydrotherapy. Patient functional disability was assessed at the time of admission and discharge using the WeeFIM (Functional Independence Measure for Children) tool.[10] The psychologist conducted individual counseling sessions every day for 60 minutes, which were focused on addressing the patient anxiety disorder and developing coping skills to better manage stressful life events. During these sessions the patient learned pain self-management techniques, which included relaxation, deep breathing, and guided imagery. The psychiatrist managed her psychotropic medications.

TABLE 1. Daily Schedule for Inpatient Management of Children With CRPS

7:00 AM	Wake up, get ready for the day
7:45 AM	Breakfast in the dome (common area)
8:00-9:00 AM	Psychotherapy
9:00-10:00 AM	OT
10:00-11:00 AM	PT
11:00-12:00 PM	PT group
12:00-1:00 PM	Lunch in the dome
1:00-1:30 PM	Patient practices OT exercises in his/her room
1:30-2:00 PM	OT
2:00-2:30 PM	PT
2:30-3:30 PM	Patient practices PT exercises in his/her room
3:30-4:30 PM	School homework or playroom
Tuesday	Music therapy
Thursday	Art therapy
4:30-5:00 PM	Patient practices OT exercises in his/her room
5:00 PM	Dinner in the dome
6:00-7:30 PM	School homework
7:30-8:30 PM	Activity of your choice, out of bed
8:30 PM	Shower
9:00 PM	Lights out

CRPS indicates complex regional pain syndrome; OT, occupational therapy; PT, physical therapy.

Outcomes

The patient was able to use her hands and was able to walk with the help of a walker, within 48 hours from the placement of the catheters. The infraclavicular and epidural catheters were removed 4 and 7 days, respectively, after the placement. The decision to remove the catheters was taken in conjunction with the PT and OT and was based on how rapidly the patient recovered the function of her upper and lower extremities. The clonidine, baclofen, and quetiapine were stopped the day of admission and the dose of duloxetine was increased to 60 mg. The methadone was weaned off in 6 days. The WeeFIM scores at the time of admission and discharge were 11 and 45, respectively (max 63).

Nine days after admission, the patient was discharged home, able to walk, climb stairs, eat, shower independently, and write. Her medications at discharge included gabapentin 900 mg tid and duloxetine 60 mg. She was referred to PT and OT for biweekly sessions and to a team psychiatrist for psychotherapy and medication management. The psychiatrist diagnosed her with pain disorder complicated by anxiety and depression. A few days after her discharge home, the patient had a set back with increasing leg pain and the need for a walker. It took the patient 3 additional months of intense psychotherapy and PT as an outpatient to be functional and back in school. The psychiatrist, a member of our team, followed up with the patient providing individual and family therapy.

A year after her discharge from the hospital the patient is still participating in individual and family psychotherapy and she is still on psychotropic medications. She was never readmitted to the hospital for recurrence of CRPS or other pain symptoms.

DISCUSSION

This case report supports the role of regional analgesia in children with CRPS. Although PT has been shown to provide sustained benefits in most children with CRPS,[11] there are patients, like the one described in this case report, in which even prolonged sessions of PT are ineffective. The use of single or multiple catheters should be considered in the

treatment of adolescents with CRPS who have failed non-interventional techniques, because of the ability to lower pain intensity and allow for an active participation in a rehabilitation process.

The use of multiple catheters has been documented for management of adult patients with acute postsurgical or posttrauma pain.[12] Similarly, the use of single or double catheters for treatment of CRPS has been documented in the literature.[13] However, there are no reports of 3 catheters used simultaneously to control diffuse pain in children with CRPS.[7,14] There are 2 caveats when using regional anesthesia techniques in children to facilitate PT and OT interventions: local anesthetic toxicity and motor blocks. The maximal recommended dose of amide local anesthetic used for continuous infusion is 0.4 mg/kg/h.[15] In this case the combined amount of local anesthetic given was 0.3 mg/kg/h. The peripheral nerve catheter infusion of ropivacaine 0.1% has been shown to provide less analgesia after extremity surgery compared with ropivacaine 0.15% to 0.3%.[16,17] The pathophysiology of pain after surgery is different from pain secondary to CRPS[18] and this may account for the effectiveness of the ropivacaine 0.1%, in our case. The decision to use ropivacaine for the continuous infusion in the catheters was based on the reduced CNS and cardio toxic potential and its lower propensity for motor block compared with bupivacaine.[19] The presence of a motor block would have affected the patient's ability to participate in the rehabilitation sessions.

A general consensus among pain specialists has emerged indicating that the cornerstone treatment of CRPS in adults is based on early PT and OT interventions.[20,21] It has been more difficult to establish the value of physical therapy in children.[22] A recent review of the literature shows that the combination of PT and interventions such as psychotherapy and medications is successful in resolving patients' impairments. The reported average median time for recovery was 8 weeks for patients treated in outpatient settings.[22] This case report seems to support an inpatient rehabilitation approach for managing severe cases of CRPS. The time of resolution of physical impairments in this case was significantly lower than those reported in the literature, including the studies where patients were enrolled in an inpatient or day-hospital rehabilitation program, and regional anesthesia techniques were not used. In this case series the patients' recovery time was 2.8 to 4 weeks on average.[23–25] Pain is the major factor impeding a fast recovery. There is insufficient evidence for efficacy of systemic opioids in CRPS patients.[26,27] There is weak scientific evidence for using antiepileptic drugs and antidepressants and yet they are commonly used in patients with CRPS.[26] However, the literature documenting their efficacy in CRPS patients is lacking.

This case report emphasizes the role of regional anesthesia pain in children with CRPS during the acute phase of the rehabilitation program. The use of multiple catheters was effective in controlling pain and allowing this patient to actively participate in the rehabilitation program. It also eliminated the need for pharmacological interventions, in particular systemic opioids. Insurance companies usually impose significant time constraints with respect to the allowed duration of the hospital stay. This patient was discharged home as soon as she was able to walk and attend to her daily living activities. However, as shown in this case, it is paramount to establish a rigorous follow-up before patients are discharged from the hospital to prevent relapse and to assure prompt and adequate interventions should the patients experience new symptoms. The type, frequency, and duration of specific interventions should be individualized given the complexity of physical, social, and psychological conditions that characterize young patients with CRPS.

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