Case Report

CRPS of Lower Extremity– A Case Report and Review

Mrinal Joshi¹, Mahima Agrawal²

Abstract

Complex regional pain syndrome (CRPS) of the lower limb is a relatively uncommon entity as compared to CRPS of the upper extremity. Literature search has revealed only 2 retrospective case series and a single case report of lower extremity CRPS type I from 1975 to 2014 on Pubmed, isolated cases of CRPS type I of lower extremity have also been reported following knee surgeries and arthroscopies. This report presents a case of lower limb CRPS type I following blunt trauma to right foot, treatment of which was directed towards management of allodynia, vasomotor symptoms and surgical correction of deformity which had developed because of the disease, coping mechanisms were also reinforced through counselling and relaxation training. The individual responded well to treatment with a reported 75% reduction in the disabling symptoms and improvement in ambulatory status.

Key words: Complex regional pain syndrome.

Introduction:

CRPS is a complex and poorly understood condition which was known previously by varying names, most commonly by the term reflex sympathetic dystrophy (RSD) and causalgia. Until late, the name was finally changed to complex regional pain syndrome at a consensus meeting in Orlando, Florida in 1994¹². Specific diagnostic criteria were codified by the International Association for the Study of Pain (IASP) as depicted in Table I ³. CRPS may affect either upper or lower limb, may be localised, or may involve the whole extremity. Rarely it affects more than one limb⁴⁻⁵. CRPS involving the lower extremity presents a diagnostic and a therapeutic challenge to the physicians, it is an uncommon disease with a prevalence of <2% in most retrospective series ⁶. A study⁷ from Netherlands reported an incidence of 26.2 cases per 100,000 person years, whereas a study⁸ from the United States estimated the incidence at 5.5 cases per 100,000 person years. A higher incidence of CRPS is reported in patients between the ages of 40 and 49 with a female preponderance (76%)⁹. The upper extremity is affected twice as commonly as the lower limb, and fracture is the most common trigger (46%).

The success of pharmacologic and other treatment modalities differs in the upper and lower limb with the lower limbs being more refractory to treatment¹⁰⁻¹⁵. This differential response has led some to recognise the CRPS of lower limbs as a distinct clinical entity ¹⁰,¹⁶⁻¹⁹. CRPS in the lower limbs is a recognised complication of knee surgery including tibial osteotomy ²⁰,²¹, and arthroplasty²² where it is associated with a poor prognosis. It has also been reported following crush injury to foot ²³, ankle arthrodesis ²⁴, amputation ²⁵, and hip arthroplasty ²⁶. One multicentre study quotes a frequency of 2.3% following knee arthroscopy ²⁷ and less frequently following ankle arthroscopy ²⁸,²⁹.

The clinical course of CRPS passes through three stages and earlier the intervention is done better are the outcomes, but the same might not be true for lower extremity affection ¹⁰,¹⁶⁻¹⁹.

Here, we report a person who developed clinical findings suggestive of CRPS type I, nearly one month after a traumatic event in the left lower extremity. The patient did not do any weight bearing on the affected limb due to which he also developed a fixed equines deformity of the same extremity.

Authors’ affiliation:

¹ MBBS, M.D., DNB PMR, Professor
² MBBS, M.D., DNB PMR, Senior Resident
Department of Physical medicine and rehabilitation, S.M.S.M.C and hospital, Jaipur, Rajasthan, India

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Correspondence:

Dr. Mahima Agarwal, Professor and head of the department, Department of Physical medicine and rehabilitation, S.M.S.M.C and hospital, Jaipur, Rajasthan, India. Phone: 09829757969, Fax: 011-23463203, Email id: drmahima@yahoo.com
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Case Report:

Mr (R) an 18 years old, Indian male was referred to the Rehabilitation Research Centre, Sawai Man Singh Hospital, Jaipur, India for chronic right lower extremity pain. He used to work as an iron cutter and while working, one of the rod pieces fell on his foot following which he experienced excruciating pain. He was taken to local hospital where a below knee plaster cast was applied for few weeks and was advised immobilisation. When the cast was removed, he was not able to bear weight due to pain and also started experiencing allodynia. It was not relieved by local or systemic medication and slowly he stopped weight bearing on the affected leg, which soon became stiff and also developed a fixed equinovarus deformity (Fig 1).

The chief complaint was severe, excruciating pain even with non-painful stimuli like touch or blowing over the skin, stiffness in the entire left lower limb with inability to bend the leg at hip, knee, ankle and foot. Pain was aggravated by motion, dependency and touch and there was intolerance to cold due to which he used to keep his ankle and foot covered with a soft cotton cloth. His pain was severe burning type not relieved with any treatment.

He was extensively investigated elsewhere but his haematology and biochemistry investigation reports were normal. MRI of brain and spine were also normal. X-rays of left ankle, knee and hip which revealed osteopenia which was confirmed on a DEXA scan. A triple phase bone scan was done which revealed increased flow of the tracer in perfusion phase, increased soft tissue pooling in blood pool phase and increased tracer uptake in the peri-articular regions of the left lower limb in delayed skeletal phase. These findings were consistent with the diagnosis of CRPS. Initially, nerve conduction study (NCS) was not possible because of pain, but after the subsidence of symptoms NCS revealed pure axonal neuropathy of common peroneal and tibial nerves. This can be seen in Table 2.

Initially, he was started on carbamazepine and baclofen, dose was increased after 7 days as there was no relief. Patient was then shifted on gabapentin 800 mg and nortryptaline 20 mg per day in divided doses, after which he showed 50% improvement in his symptoms.

He was also initiated on alendronate and propranolol. The symptoms kept on improving slowly and he was then progressed from non-weight bearing range of motion exercises to partial weight bearing. At the end of two months, he was able to freely bear weight and was not having any sensory symptoms.

Then, a surgical correction of equinovarus deformity was performed and was given a solid ankle foot orthosis for ambulation (Fig 2). After 6 weeks of immobilisation in the cast patient had some pain on removal of cast, which was again managed by increasing the dose of Nortryptaline. He has developed good pain relief and is now ambulating with ankle foot orthosis without any walking aid and is currently on Nortryptaline 10mg only.

Table 1: IASP Diagnostic Criteria for Complex Regional Pain Syndrome (CRPS)³

1. The presence of an inciting noxious event, or a cause of immobilization ^{1}
2. Continuing pain, allodynia, or hyperalgesia in which the pain is disproportionate to any known inciting event.
3. Evidence at some time of edema, changes in skin blood flow, or abnormal sudomotor activity in the region of pain (can be sign or symptom).
4. This diagnosis is excluded by the existence of other conditions that would otherwise account for the degree of pain and dysfunction.

If seen without “major nerve damage” diagnose CRPS I; if seen in the presence of “major nerve damage” diagnose CRPS II.

³ Not required for diagnosis; 5-10% of patients will not have this
Table 2: Nerve Conduction Study of Reported Case.

<table>
<thead>
<tr>
<th>Nerve: Right Peroneal</th>
<th>Site</th>
<th>Lat1 (ms)</th>
<th>Dur (ms)</th>
<th>Amp</th>
<th>NCV (m/s)</th>
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</thead>
<tbody>
<tr>
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<td>8.44</td>
<td>0.7 mV</td>
<td>29.03</td>
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<tr>
<td>Knee</td>
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<td>6.25</td>
<td>65.7 μV</td>
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<th>Site</th>
<th>Lat1 (ms)</th>
<th>Dur (ms)</th>
<th>Amp</th>
<th>NCV (m/s)</th>
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<td>14.58</td>
<td>0.3 mV</td>
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<tr>
<td>Knee</td>
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<td>10.10</td>
<td>0.3 μV</td>
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<th>Site</th>
<th>Lat1 (ms)</th>
<th>Dur (ms)</th>
<th>Amp</th>
<th>NCV (m/s)</th>
</tr>
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<tbody>
<tr>
<td>Ankle</td>
<td>8.23</td>
<td>10.00</td>
<td>0.5 mV</td>
<td>25.85</td>
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<tr>
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<th>Site</th>
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<th>Dur (ms)</th>
<th>Amp</th>
<th>NCV (m/s)</th>
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<td>34.11</td>
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<tr>
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<td>8.44</td>
<td>1.4 μV</td>
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</table>

**Discussion:**

Treatment of CRPS has not yet achieved precise standard guidelines and doctors as well as patients face difficulties with the diagnosis and management. Treatment should be started as early as possible to avoid secondary complications of chronicity of pain and fear, anxiety of living with undiagnosed pain. Because of the variation in treatment approach, an integrated interdisciplinary treatment approach is recommended, tailored to the individual patient. The primary aims are to reduce pain, preserve or restore function, and enable patients to manage their condition and improve their quality of life.

CRPS has a variable presentation. Nearly, 95% cases of CRPS elicit a history of trauma or surgery. In a classic case, within the first six weeks or so after injury, the sympathetic overreaction causes a swollen, immobile, and painful limb. Pain is in a non-anatomic distribution i.e. it does not follow the distribution of a peripheral nerve, and is often described as burning in nature. There is increased sweating, with colour changes in the extremity in most of the patients. Frequently, there is allodynia i.e. pain elicited with a non-noxious normal stimulus such as light touch of a bed sheet, blowing of air, etc. Late in the course of the disease, there are trophic changes (sudomotor): Dystrophic, smooth, shiny skin; osteoporosis; fast growing and brittle nails; hypertrichosis; and muscular and subcutaneous atrophy. All these symptoms may be found along with joint swelling and contractures. In the dystrophic stage, ankle involvement may lead to a fixed equinovarus deformity of the hind foot with firm induration around the tibiotalar joint which was also present in this case.
In acute phase of CRPS there is neither a radiographic change nor an evidence of osteoporosis. Technitium 99m bone scans are commonly used to confirm the diagnosis of CRPS, but they have a specificity of 75% to 98% and a sensitivity of only 50% for this condition [34,35]. In the later stage, x-rays may reveal patchy demineralisation of the affected part [36]. A comparison of x-rays an bone scintigraphy [36] reveals bone scans are more sensitive than x-rays, especially in the early phase of this condition. If the knee is involved, osteopenia of the patella and medial femoral condyle on the sunrise view characteristically is present [16,37-39]. MRI has more recently gained recognition in diagnosis [40, 41], especially when radiology is contraindicated. Infrared thermography is sensitive and specific in the diagnosis, but it may have a role in monitoring progress [41]. But combining clinical evaluation and bone scan, give a better direction to diagnosis than singly.

Akkoc et al [44] reported a case with bilateral lower extremity CRPS in a patient with paraplegia occurring following spinal disc herniation surgery, who was treated with anticonvulsants and opioids for 6 months. She remained well but had a recurrence of symptoms for which she was treated successfully with pulsed radiofrequency (PRF) lumbar sympatholysis.

Harris et al [45] reported the characteristic trends in 64 lower extremity CRPS patients in a retrospective study where they examined 26 variables broadly classified under demographic characteristics, CRPS characteristics and healthcare utilisation. The sample had predominantly white, middle aged women with CRPS I. Subjective complaints consisted of burning, sharp, throbbing, or aching pain with shooting symptoms. Most common cause was blunt trauma to foot with or without fracture and ankle sprain.

Saranita et al [46] reported a case of CRPS who developed the same after ankle injury and surgery highlighting the treatment options for the same including spinal cord stimulation (SCS). They reported the clinical evidence of SCS to be of level 4. Manjunath et al [47] reported 20 cases of lower extremity CRPS and compared the efficacy of percutaneous radiofrequency lumbar sympathectomy and phenol lumbar sympathetic neurolysis and found the two to be comparable.

Majumdar et al [48] reported 4 cases with fixed dystonia who developed CRPS of the involved extremity, one of whom had an identifiable traumatic precipitant. They stated that treatment with multi-disciplinary approach including psychological measures and physiotherapy is more likely to be successful and may prevent unnecessary physical measures.

McCrorry and Westerling [49] reported a 73 years old female of polycythemia vera presenting as CRPS of lower extremity. She was treated successfully with repeated venesection, mild analgesics and amitryptaline. Unlu Sakaci et al [50] reported a 70 years old female who developed simultaneous upper and lower limb CRPS of the hemiplegic side after stroke. She was treated successfully within 5 weeks with a combination of medical and physical therapy.

Gallo and Codispoti [51] reported a case of 25 years old soldier who sustained a blast injury causing multiple spinal fractures, extremity fractures and developed lower extremity CRPS. Electrophysiological studies showed lumbosacral plexopathy and bone scan findings were consistent with the diagnosis of CRPS. He was treated successfully with conservative measures.

Several studies report treatment modalities based on their results in upper extremity CRPS. Literature reports no difference between intravenous regional anaesthesia with guanethidine, reserpine and saline control [52-54] based on which intravenous regional anaesthesia was not used as a treatment modality in this case.

Continuous epidural anaesthesia or intrathecal narcotic pump allows the oral narcotics to be used in a lesser dosage and have been found to be effective [55,56]. Despite this success the requirement for long term hospitalisation, risks of urinary retention, hypotension and excessive cost of treatment precludes the use of this modality in a public hospital. Some of the alpha adrenergic blocking agents like phenoxybenzamine and prazocin have been used successfully but all the treated patients required long term management [57-59]. And for this reason these drugs were not tried in the hereby reported case.

There is no published data regarding the efficacy of clonidine in the treatment of CRPS. There is some clinical evidence that oral beta blockers like propranolol may be of some benefit. A total of five patients with acute CRPS treated with oral Propranolol have been reported. Three of them were free of symptoms at their last follow-up [60,61] and two had no relief [62]. There was significant pain relief as the patient was started initially on propranolol 20mg which was tapered later on depending the amount of pain relief. Some of the calcium channel blockers were used with great success in CRPS [43] but Prough et al reported that three patients had relief with nifedipine while 2 required maintenance doses [63].

Newer treatments supporting intrathecal baclofen [64], pulsed radiofrequency along with spinal cord stimulation [65] have been shown to have some evidence in literature.
Adami et al.\textsuperscript{66} reported that alendronate was effective in treating patients with long standing CRPS. Alendronate was started initially in this case for first two months and after starting weight bearing it was withdrawn. Rehman et al.\textsuperscript{67} showed that these patients have lower than normal bone density and that this relative osteoporosis improves with pamidronate. Many review articles have studied the use of anticonvulsants in treating lower extremity CRPS, but none have substantiated their effectiveness\textsuperscript{34}. The only anticonvulsant studied clinically and reported in the literature to date is gabapentin which is helpful in relieving pain in CRPS\textsuperscript{68}. Use of intravenous corticosteroids is well established, but none of the studies have reported their efficacy alone in lower extremity CRPS\textsuperscript{34}. Use of gentle physical therapy and electrotherapy has been recommended all throughout the course of the disease.

Conclusions:

For diagnosing CRPS, along with IASP diagnostic criteria, bone scan results can give a more conclusive picture. Early treatment and multidisciplinary approach can give equivocal results in comparison to an interventionist approach.

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References:


