Case Report

Complex regional pain syndrome—A forgotten entity

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Abstract

Complex Regional Pain Syndrome (CRPS) is a painful condition characterized by regional pain that is disproportionate temporality and severity to the usual course of any known cause. The pain is regional which is not in a specific nerve territory or dermatomal distribution. Judicious clinical suspicion and targeted investigations are necessary for correct diagnosis and appropriate treatment. We hereby report a case of a young lady with CRPS who presented to us with unilateral upper limb pain, swelling, discoloration and muscle atrophy without any history of inciting injury.

Keywords: Bone scan, causalgia, complex regional pain syndrome, stellate ganglion blockade

Introduction

CRPS is a clinical syndrome characterized by searing pain disproportionate to any inciting event, hyperalgesia, edema, allodynia, as well as sudomotor, vasomotor, and skin changes.[1] CRPS Type I is the most common and involves a limb without a direct nerve injury. CRPS Type II, previously referred to as causalgia, involves a defined nervous pathway and is associated with a direct nerve injury.[1]

Case Report

A 25-year-old female, teacher by profession, with complaints of weakness and pain in left hand involving 3rd, 4th, and 5th digits associated with pain in wrist, elbow, and shoulder joint on the same side for 2 months. Over the next 15 days she started developing swelling and redness involving last three digits along with blackish mottling of the overlying skin on the dorsal aspect. She also had increased pain sensation in those digits to the slightest of touch such that she would keep her fingers flexed and avoid movements. She sought medical consultation at the dermatology clinic before consulting various other specialties and ultimately being referred to our rheumatology clinic.

At dermatology clinic, on clinical examination, ulnar nerve was found to be minimally thickened and palpable along with skin changes including hyperpigmented patches and hypoaesthetic plaques. There was reduced sensation to cold with preserved fine touch and pain sensations. Ulnar nerve biopsy was ordered which came out to be normal. Skin biopsy showed hyperkeratotic epidermis with large number of budding yeasts in stratum corneum and was prescribed fluconazole along with NSAID'S and topical ointments but to no relief.

Patient visited neurology clinic for her complaints of excessive pain apparently in the ulnar nerve distribution. A possibility of mononeuritis multiplex was considered clinically. She underwent nerve conduction studies of all four limbs and MRI of cervical spine were revealed normal. Patient was started on Tramadol 50 mg twice daily, Gabapentin 150 mg twice daily, duloxetine 30 mg once daily, and Vitamin B-12 supplements but without any relief in her symptoms.

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Meanwhile, the patient consulted many orthopaedicians and physical medicine and rehabilitation centers but to no relief. With every passing day, her pain increased to the extent that she stopped venturing out in public places to avoid any touch on her fingers. Pain became so debilitating that she used to cry and could not sleep for weeks altogether.

In our rheumatology clinic, she denied history of recent or remote trauma to the left upper limb or any history of previous surgeries. On examination, there was flexion deformity at proximal and distal interphalyngeal joints involving left 3rd, 4th, and 5th digits [Figure 1]. There was associated soft tissue swelling and skin changes in the form of blackish discoloration and dryness and scaling but with no synovitis. Clubbing of digits with increased nail angle along with whitening and excessive shining of nails of 3rd, 4th, and 5th digits was also noticed [Figure 2]. Atrophy of thenar and hypothenar muscles of left hand was evident. Other proximal joints, namely elbow and shoulder, were normal on clinical examination. Pain was assessed with Visual Analogue Scale (VAS), the score of which was 9 at presentation.

Her previous lab reports were reviewed. Blood counts were within normal limits. Liver function tests, lipid profile, thyroid profile, and renal function tests were all within normal limits. Her chest X-ray showed no abnormality. Serology for viral hepatitis, gonococcal, and HIV was negative. Rheumatoid factor, antinuclear antibody, and HLA-B27 were also negative. Radiograph of hand and wrist showed marked regional osteopenia with bone resorption involving mainly around the metacarpo-phalyngeal and interphalyngeal joints without any erosions or sclerosis [Figure 3]. A Technitium labeled MDP triple phase bone scan was advised that revealed increase in radiotracer accumulation in the region of left forearm and left wrist at inflow and pool phase images. Delayed whole body static images showed increased peri-articular radiotracer uptake in left shoulder joint, left elbow joint, and left wrist joint. Focal increased radiotracer uptake was also seen in metacarpophalyngeal joint and proximal interphalangeal joint of 3rd and 4th fingers of left hand.

Hence a diagnosis of CRPS type I was made according to Budapest criteria. As the patient did not respond to NSAIDs, amytryptiline, and pregabalin. Left stellate ganglion block was given [Figure 4]. After 5th sitting of stellate ganglion block, patient rated a score of 3 on VAS scale. With time, patient was able to do her routine day to day activities and is presently being followed up regularly.

Discussion

Pathogenesis of CRPS is unknown and may include alterations in cutaneous innervation, central and peripheral sensitization, altered function of the sympathetic nervous system, increased levels of local and systemic pro-inflammatory cytokines, lower systemic levels of anti-inflammatory cytokines, genetic factors, and psychologic factors.[2] In this case, we used the Budapest criteria to make the clinical diagnosis of CRPS [Table 1].[3] The diagnosis may have been missed if not for using the diagnostic criteria.

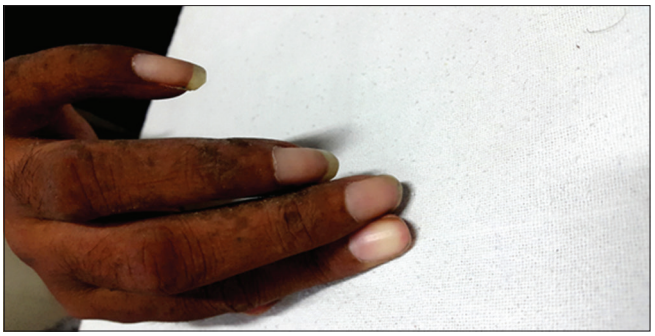

Figure 1: Flexion deformity at proximal and distal interphalyngeal joints involving left 3rd, 4th, and 5th digits

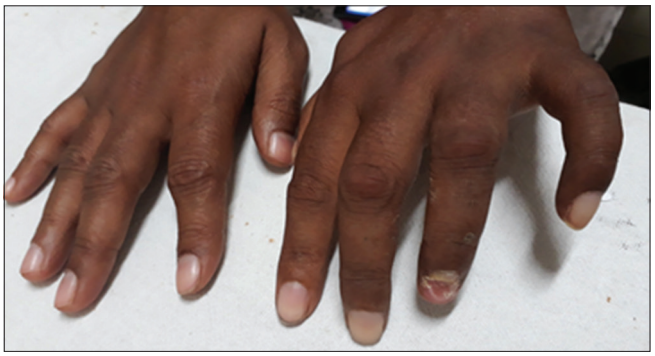

Figure 2: Clubbing of digits with increased nail angle along with whitening and excessive shining of nails of 3rd, 4th, and 5th digits

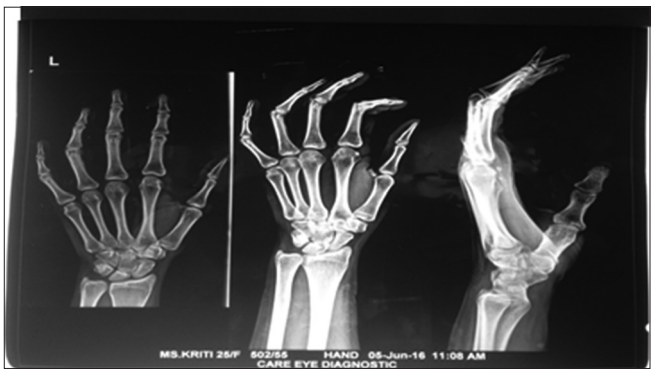

Figure 3: Hand and wrist radiograph demonstrating periarticular osteopenia at metacarpophalyngeal joint and patchy osteopenia of wrist bones

The psychiatric medical treatment is not less important,[4] since most of these patients seem to have lower baseline levels of stress and depression. Bisphosphonates seem to play a role when osteopenia is massive[5] whereas corticosteroids, non-steroidal anti-inflammatory drugs as well as gabapentin-related painkillers have also proven useful for pain alleviation.[6,7]

Blockade of the somatic nerve or sympathetic ganglion is performed in more complex and refractory cases of the syndrome.[8] Heir *et al.*[9] observed treatments for this syndrome

Table 1: Budapest criteria for CRPS

All of the following statements must be met:
The patient has continuing pain that is disproportionate to any inciting event
The patient has atleast one sign in two or more of the categories below
The patient reports atleast one symptom in three or more of the categories below
No other diagnosis can better explain the signs and symptoms

No.	Category	Signs/symptoms
1.	Sensory	Allodynia and/or hyperalgesia
2.	Vasomotor	Temperature asymmetry and/or skin colour asymmetry
3.	Sudomotor/edema	Edema and/or sweating changes and/or sweating asymmetry
4.	Motor/trophic	Decreased range of motion and/or motor dysfunction and/or trophic changes

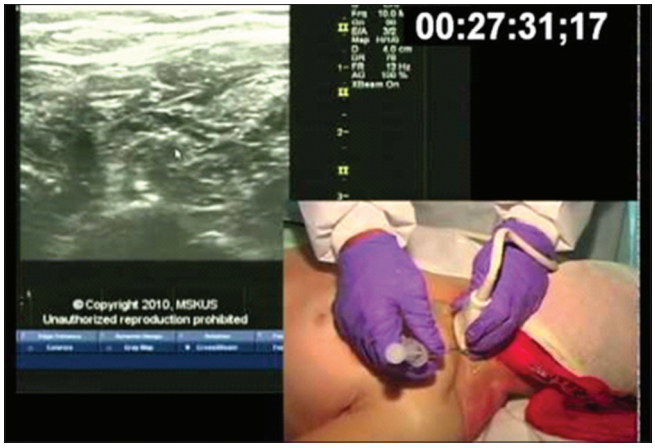

Figure 4: Showing technique of ultrasound guided stellate ganglion block

in two reported cases. In the first, two stellate ganglion blockades were performed, obtaining an improvement of 80%; and in the second, after numerous failures in treatment, it was only possible to abolish the pain with this blockade.

Conclusion

This case has been presented to sensitize the common physician towards this common disease which is very frequently misdiagnosed as cervical spondylosis, radiculopathy, or even brachial plexus syndrome. The physician should look upon severe pain without significant trauma, atrophy, and color changes which different it from others.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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