Case Report

Complex Regional Pain Syndrome Secondary to Leprosy

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Abstract

Introduction. Leprosy is a chronic infectious disease caused by Mycobacterium leprae affecting the skin and the nerves. A clinical spectrum of disease is seen ranging from purely neuritic, tuberculoid to lepromatous spectrum according to the immune status of the patient. During the course of the disease, immunologically mediated hypersensitivity lepra reactions can occur that are associated with painful neuritis and neuropathy [1].

Complex regional pain syndrome (CRPS) was formerly known as Sudeck’s dystrophy or causalgia, consisting of a triad of autonomic, sensory, and motor symptoms disproportionate to the inciting event (inflammatory, infective, or traumatic nerve damage).

Case. A 20-year-old male presented with continuous pain, aggravated by cold and emotions, loss of fine touch and temperature sensation, redness, swelling, along lateral aspect of left hand and forearm with weakness in the grip of 6 months’ duration. There was a 5-year history of sensory loss only over left index finger that he ignored. Examination revealed abnormal sensory and autonomic functions along left radial and median nerve distribution that were confirmed by nerve conduction studies suggestive of mononeuritis multiplex. Radial cutaneous nerve biopsy was suggestive of leprosy. Magnetic resonance imaging and ultrasonography showed no compressive etiology; however, MRI showed involvement of brachial plexus. Antileprosy, anti-inflammatory drugs, and steroids were given in view of neuritis because of lepra reaction with supportive measures of physiotherapy, transcutaneous electrical nerve stimulation, to no avail. A surgical median nerve decompression also failed to relieve the pain. Temporary stellate ganglion block improved the pain scale. Thus, excluding all other causes, the final diagnosis was CRPS secondary to leprosy. There is only one reported case of CRPS with leprosy.

Conclusion: Leprous neuropathy caused the nerve damage that lead to CRPS type 2. Very rarely leprosy can lead to CRPS. CRPS is a diagnosis of exclusion.

Key Words. CRPS; Neuropathy; Leprosy; Brachial plexopathy; Sympathetic Block

Introduction

Leprosy is a chronic infectious disease caused by Mycobacterium leprae affecting the skin and the nerves. A clinical spectrum of disease is seen ranging from purely neuritic, tuberculoid to lepromatous spectrum according to the immune status of the patient. During the course of the disease, immunologically mediated hypersensitivity lepra reactions can occur that are associated with painful neuritis and neuropathy [1].

Complex regional pain syndrome (CRPS) was formerly known as Sudeck’s dystrophy or causalgia, consisting of a triad of autonomic, sensory, and motor findings. International Association for Study of Pain has divided CRPS into two types [2].

CRPS Type 1: Previously known as Sudeck’s reflex sympathetic dystrophy. It develops after remote trauma or relatively minor injury and no identifiable nerve pathology is seen.

CRPS Type 2: Formerly known as causalgia. It develops after substantial injury to a major nerve with obvious underlying nerve pathology.

Clinical features consist of autonomic, sensory and motor symptoms, and signs.

Stage I (early acute stage) shows hyperalgesia, allodynia, signs of vasomotor dysfunction, and prominent edema and sudomotor disturbance.

Stage II (dystrophic stage) occurs 3–6 months after onset characterized by more marked pain/sensory dysfunction, continued evidence of vasomotor dysfunction, with development of significant motor/trophic changes.

Stage III (atrophic stage) is characterized by decreased pain/sensory disturbance, continued vasomotor disturbance, and markedly increased motor/trophic changes [3].
We are reporting a case of CRPS type II where leprosy is the cause of underlying neuropathy.

Case Report

A 20-year-old immunocompetent male presented with loss of sensation and numbness of the tip of left index finger of 5 years; duration. He gave history of excruciating pain, numbness, weakness, redness, and swelling along the lateral aspect of left hand and forearm 6 months prior to presentation. The pain was sharp, continuous, aggravated by cold and emotions causing disturbed sleep. There was no history of significant medical or surgical complaints or any preceding trauma. Patient had taken multiple treatments in the form of nonsteroidal anti-inflammatory drugs, gabapentin, carbamazepine, amitriptyline, lorazepam, and systemic steroids.

Examination revealed a well defined, erythematous, edematous plaque on the lateral aspect of left hand (Figures 1 and 2) with loss of fine touch and temperature sensations but dysesthesia to crude touch along the innervation of median nerve with an initial visual analogue pain scale (VAPS) of 9. Motor examination revealed wasting of the left thenar muscles and a muscle power 4/5 with absence of supinator reflex. Autonomic function testing showed increased temperature with absence of sweating in the affected area (Figure 3). The left radial cutaneous and left median nerves were uniformly thickened. Systemic examination did not reveal any significant abnormalities.

Differential diagnoses of borderline tuberculoid leprosy in type 1 lepra reaction, thoracic outlet syndrome, and carpal tunnel syndrome were considered.

The hematological and serum biochemistry were normal. Serology for human immunodeficiency virus, syphilis, and autoimmune profile were negative. Skin biopsy showed atrophic epidermis overlying a normal-looking dermis with no evidence of chronic granulomatous response suggestive of leprosy. Nerve conduction studies showed complete absence of sensory and motor responses in left median nerve and left radial cutaneous nerve. Hematoxylin and eosin-stained sections of radial cutaneous nerve biopsy showed foamy macrophages admixed with lymphocytes along with fibrosis of epineurium and perineurium suggestive of pure neuritic leprosy (Figures 4 and 5).

X-ray showed no evidence of any bony changes. Ultrasound examination showed segmental thickening of median and radial nerve. MRI brachial plexus and cervical spine revealed no compressive etiology along with abnormal enhancement signals along the roots, trunk, and division of left brachial plexus, suggesting involvement of central nervous system in leprosy (Figure 6) [4].

Based on the clinical, histopathological, and radiological findings, a provisional diagnosis of pure neuritic leprosy with type 1 lepra reaction was made. Anti-leprosy drugs with systemic steroids for neuritis were started, and pain medications were continued along with supportive physiotherapy and transcutaneous electrical nerve stimulation.

After 2 months of therapy, repeat nerve conduction studies and electromyography showed mild improvement in motor response and no progression of disease. There was no significant improvement in the pain. Surgical decompression of the median nerve in the carpal tunnel did not improve the pain. A temporary stellate ganglion block with ropivacaine and bupivacaine showed an improvement in the VAPS from 9 to 6. Thus, we arrived at a final diagnosis of CRPS type 2 secondary to purely neuritic leprosy.

Discussion

M. leprae infects the Schwann cells of the nerves that stimulate an immunological response from the body that...
cause nerve infiltration with inflammatory cells leading to nerve dysfunction and nerve damage. Neuritis is also seen as a part of the immunologically mediated lepra reactions. Treatment of leprosy consists of multidrug therapy consisting of rifampicin, clofazimine, and dapsone. The reactions of leprosy are managed by steroids and other immunosuppressants [1].

The etiology of CRPS includes trauma, neuropathies secondary to alcohol, diabetes mellitus, vascular causes like stroke and myocardial infarction, postherpetic neuralgias, operative procedures, and cast immobilization leading to entrapment of nerves.

Pathogenesis is not exactly known; however, it has been postulated that a vicious cycle of axonal injury, inflammation, and vascular dysfunction sets in, which leads to malfunctioning of small-fiber axons causing remaining fibers to inappropriately fire action potentials and release neuropeptides. This causes activation of pain receptors, transmission, and modulation of pain [5,6].

Early diagnosis and treatment of underlying condition improves success of therapy. Therapy includes nonsteroidal anti-inflammatory drugs, glucocorticoids, calcitonin and bisphosphonates, tricyclic antidepressants, mem-

Figure 3 Autonomic function testing with starch iodine testing showing absence of sweating in affected area.

Figure 4 Radial cutaneous nerve biopsy was showing fibrosis of epineurium and perineurium.

Figure 5 Mononuclear lymphocytic infiltrate admixed with foamy macrophages suggestive of leprosy.
brane stabilizers—carbamazepine, gabapentin, lidocaine, mexiletene, and N-methyl-D-aspartate receptor antagonists—and ketamine [7]. Topical capsaicin cream, dimethyl sulfoxide, and lignocaine cream can be used [8]. Physical and supportive measures consist of exercise therapy, occupational therapy, and transcutaneous electrical nerve stimulation. Regional anesthetic techniques like sympathetic ganglion block with local anesthetics, serial conduction blocks of brachial or lumbar plexus, and neuromodulation techniques like spinal cord stimulation have been tried with some success [9]. The ultimate goal of treatment is to restore function with a multidisciplinary approach.

Our patient had long-standing pure neuritic leprosy that he neglected to treat initially, which then caused fibrosis, thickening, and structural changes in the affected nerves as seen in the ultrasound and MRI examination. On the basis of the chronic pain refractory to all modalities of treatment, with a triad of autonomic, motor, and sensory findings and no other diagnosis that better explained these findings, a diagnosis of CRPS type 2 was made.

In ruling out other causes for the severe pain, the MRI performed incidentally showed involvement of the brachial plexus roots in cervical spine. This could not be attributed to any other pathology other than leprosy. This is extremely rare in leprosy [4].

Our patient is being managed with a multidisciplinary approach by a team including a dermatologist, a neurologist, a pain management specialist, and a physiotherapist. Serial temporary stellate ganglion blocks along with selected sensory median nerve blocks and continuous infusion of local anesthetics with an indwelling catheter put under ultrasonographic guidance along with intralesional steroid and ketamine have been carried out. There was no evidence of a neuroma on ultrasound of the scar tissue. Intramuscular botulinum toxin injections in the hand have not helped. Even with multidisciplinary approach, complete pain relief has not been achieved. The pain escalates once the effect of the local anesthetic wears off. This highlights the importance of early diagnosis and treatment of leprosy as once the fibrosis of nerves sets in pain cannot be managed effectively.

Lately, the patient has been put on thalidomide 100 mg once daily orally to which he has reported a change in visual analog pain scale from 6 to 3 in 1 month. The dose of thalidomide has been increased to 200 mg per day, and we will wait and watch for further developments.

To date, only one case report has been published of leprosy being the cause of CRPS by Garg and Dehran in 2010 [10].

Conclusion

Leprosy can cause neuropathy that can lead to CRPS. Leprous neuritis is amenable to treatment with steroids and immunosuppressants, while CRPS is not. CRPS is a diagnosis of exclusion. Its association with leprosy is very rare and difficult to manage as in our case.

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He has given the sympathetic block for the patient to diagnose CRPS and has helped in the management of the pain.

Dr. A. B. Shah, Consultant Neurologist, Former Professor and Head of Department of Neurology, T. N. Medical College and B. Y. L. Nair Hospital, Mumbai 400008, India.

He has confirmed the diagnosis of CRPS and referred the patient for further management.

Dr. A. S. Baliarsing, Head of Department of Plastic Surgery, T. N. Medical College and B. Y. L. Nair Hospital, Mumbai-400008, India.

He has performed the median nerve decompression.

References


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