Case Reports

Complex Regional Pain Syndrome Type II Associated with Lumbosacral Plexopathy: A Case Report

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Abstract

Introduction. Complex regional pain syndrome (CRPS) is a well-known clinical entity, first described in the 1800s, consisting of pain, hyperalgesia, edema, and sudomotor changes either without (Type I) or with (Type II) a definable nerve injury. CRPS Type II is most commonly caused by high velocity missile injuries, mononeuropathies, and partial nerve transections.

Case Report. In this case, a 25-year-old soldier who sustained a blast injury causing multiple spinal compression fractures, extremity fractures, and pelvic and sacral fractures was transferred to a U.S. Army medical center for surgical management and rehabilitation. He complained of weakness, sensory changes, and pain in his left lower extremity. The patient also demonstrated swelling and hyperesthesia of the left foot and ankle. Undiagnosed soft tissue injury, fracture, and deep venous thrombosis were ruled out by imaging studies. The patient had an electromyogram/nerve conduction study (EMG/NCS) that showed widespread left sided lumbosacral plexopathy as well as possible cauda equina injury. Triple phase bone scan demonstrated findings consistent with CRPS of the left foot and ankle. He was started on a tricyclic antidepressant and an anticonvulsant. Physical and occupational therapy were quickly engaged to incorporate range of motion exercises, mirror therapy, and physical modalities. The patient continued conservative management and rehabilitation and eventually was discharged with significantly improved function and decreased pain.

Conclusion. Although many causes of CRPS Type II have been described, this is only the second reported case of CRPS Type II secondary to lumbosacral plexopathy in the literature.

Key Words. Causalgia; Lumbosacral Plexopathy; Complex Regional Pain; Syndrome

Introduction

Complex regional pain syndrome (CRPS) was first described in 1813, and the term "causalgia" was later coined in 1864 by Civil War physician Silas Weir Mitchell to describe a syndrome characterized by distal burning pain, swelling, and skin changes in the setting of partial nerve transection, which could be affected by movement, loud noises, and strong emotions [1]. In the early 20th century, physicians began to recognize a clinical syndrome with similar symptoms but without a definable nerve lesion, which often responded to sympatholytic interventions. This syndrome became known as "reflex sympathetic dystrophy," according to the description by Evans in 1946 [2]. In 1994, the International Association for the Study of Pain (IASP) changed the name reflex sympathetic dystrophy to CRPS Type I and causalgia to CRPS Type II [3]. A recent meta-analysis of all reported cases of CRPS Type II from the 19th and 20th centuries (over 1,500 reported cases) concluded that CRPS Type II is very rare outside wartime and is most often associated with partial rather than complete transection of a nerve [4]. According to the meta-analysis, the most common cause of CRPS Type II is high velocity missile injuries, but many other causes have been reported, including blunt trauma, nerve stretch, various surgeries, venipuncture, and electrical injury [4]. Most cases of CRPS Type II describe a mononeuropathy, with the most commonly involved nerves being the median, ulnar, and tibial. The upper extremities are more often involved than the lower extremities. Cases of plexopathy most often describe involvement of the brachial plexus [4]. The following describes the unusual case of CRPS Type II associated with lumbosacral plexopathy, which has only been reported once in the literature [5].

Case Description

A 25-year-old male active duty Army officer sustained a blast injury resulting in multiple spinal compression fractures, several upper extremity fractures, left pelvic and sacral fractures involving the sacral foraminae, and a left pubic ramus fracture. In the combat zone, he underwent embolization of the left gluteal artery, exploratory laparotomy with note of a zone II non-expanding retroperitoneal hematoma, and surgical manipulation and external fixation of his pelvic and sacral fractures. He was transferred to a regional Army medical center for more definitive...
treatment of his orthopedic injuries and eventually to a U.S. Army medical center for inpatient rehabilitation. He screened positive for mild traumatic brain injury based on loss of consciousness of less than one minute but did not display any signs or symptoms of traumatic brain injury or post-traumatic stress disorder throughout his hospital stay. Approximately 1 month after his initial injury, the physical examination was significant for decreased sensation to light touch and pinprick over the left scrotum and penis, lateral thigh, posterior thigh, lateral calf, lateral foot, and hyperesthesia of the dorsum and sole of his left foot. The patient had weakness of left ankle dorsiflexors, toe extensors, and plantarflexors. The patient also had swelling of the left foot and ankle but no point tenderness. Skin temperature and texture were equal bilaterally. The patient complained of a constant burning and throbbing sensation in the left foot and calf, worse at night and with dependent positioning.

In addition to the opioids he was already receiving, he was started on gabapentin with partial relief of his symptoms. In order to rule out previously undetected fracture, plain X-rays were obtained of the left hip, femur, tibia and fibula, ankle, and foot. No fractures or abnormalities were visualized. Venous ultrasonography of the left lower extremity was ordered, and it showed adequate venous flow and no evidence of deep venous thrombosis (DVT).

An EMG/NCS of the lower extremities was performed in order to fully document the extent and location of his neurologic injury. The results indicated widespread involvement of the left lumbosacral plexus, including the femoral nerve, sciatic nerve, pudendal nerve, and gluteal nerves. Additionally, there were abnormalities in the lumbar paraspinals indicating possible involvement at the level of the nerve root or cauda equina.

The patient continued to describe burning and throbbing pain in the left foot and ankle, variable swelling, and hypernessitvity to touch. He was started on nortriptyline, and a triple phase bone scan was ordered. The images showed asymmetric increased uptake in the left ankle and foot during blood flow and pool images, with particular note of uptake within periartricular structures. These scintigraphic findings were consistent with CRPS of the left ankle and foot. The patient was taken off gabapentin and started on pregabalin, which was rapidly titrated to effect and resulted in further improvement of his symptoms. Physical therapy was quickly engaged to incorporate range of motion exercises, contrast baths, mirror therapy, and desensitization techniques into his treatment plan. The patient was subsequently transferred to a VA polytrauma rehabilitation center where he continued aggressive physical therapy and medical management. He was transferred on the following medications with approximately 50% improvement of his symptoms: enoxaparin 30 mg twice daily, docusate/senokot, polyethylene glycol, lidocaine 5% patch to left foot 12 hours per day, clonazepam 0.5 mg three times daily as needed, pregabalin 100 mg three times daily, morphine sulfate immediate release 45 mg every four hours as needed, morphine sulfate sustained release 45 mg three times daily, and nortriptyline 25 mg at bedtime.

His pelvic external fixation device was removed, and the swelling of the left foot and ankle improved as he was able to bear weight and ambulate. MRI of the left foot and ankle was obtained, showing diffuse periarticular edema consistent with CRPS. There was no evidence of soft tissue injury. He was offered a lumbar sympathetic block but preferred to maximize conservative management.

He is ambulating independently and continues to functionally improve as an outpatient.

Discussion

According to the IASP, the diagnosis of CRPS can be made if the following criteria are fulfilled: preceding noxious event without (CRPS Type I) or with (CRPS Type II) associated nerve injury; spontaneous pain or hyperalgesia/ hyperesthesia not limited to a single nerve territory and disproportionate to the inciting event; evidence at some time of edema, skin blood flow (temperature) or sudomotor abnormalities in the region of pain; and other diagnoses are excluded [3]. Because the IASP permits diagnosis based on patient report and considers pain/hyperalgesia, vasomotor abnormalities, and sudomotor abnormalities as equivalent diagnostic criteria, it has been suggested that their criteria may be adequately sensitive (rarely missing a diagnosis of CRPS) but not sufficiently specific, leading to potential false positives. A consensus group met in Budapest in 2003 to propose new diagnostic criteria for CRPS. According to the proposed criteria, the patient must report one symptom in 3 out of 4 of the following: sensory (hyperesthesia/allodynia), vasomotor (temperature asymmetry, skin color changes), sudomotor (edema, sweating), and motor/trophic (decreased range of motion, weakness, skin changes) as well as the presence of two or more signs at the time of evaluation: sensory (hyperesthesia/allodynia), vasomotor (temperature asymmetry, skin changes), sudomotor (edema, sweating), motor/trophic (decreased range of motion, weakness, skin changes) [6].

As unilateral leg swelling usually implies a local mechanical or inflammatory process, our differential diagnosis in this case included soft tissue or ligamentous injury of the foot/ankle, fracture, DVT, and venous or lymphatic insufficiency. Undiagnosed fracture, venous insufficiency, and DVT were excluded by plain X-rays and venous ultrasonography. MRI of the left foot and ankle ruled out soft tissue injury. With severe soft tissue injury, we would have expected a more immediate onset of symptoms, gradual improvement over time, and different pain descriptors than “burning” or “throbbing.” In fact, as burning is the most common pain descriptor reported in CRPS Type II, some authors have concluded that the diagnosis of CRPS Type II is unlikely if burning pain is not present [4,7]. Others have noted the primarily distal location of symptoms that was also demonstrated in our patient [1,8].
Like most medical conditions, early diagnosis and treatment is important in maximizing outcome. It is worth noting, however, that in complex polytrauma patients, distracting injuries can effectively delay the onset of symptoms [9]. Our patient had a multitude of orthopedic injuries and had been through a number of major surgeries including an urgent cholecystectomy. His left foot and ankle symptoms appeared later and could easily have been missed early in the hospital course because of his more critical surgical issues.

There is currently a paucity of evidence supporting the use of medications to treat CRPS. Medications such as tricyclic antidepressants and anticonvulsants are commonly used because of their documented efficacy in other neuropathic pain syndromes. Gabapentin has shown promise in some randomized trials [10,11]. Although opioids were once thought to have no role in the treatment of neuropathic pain, more recent research has indicated otherwise. In our patient, the pain caused by his multiple injuries would have been difficult to manage without opioids. A recent study showed that, at least for diabetic neuropathy and post-herpetic neuralgia, the combination of gabapentin and morphine was more effective than either agent alone [12]. Another study indicated that opioids were successful at reducing neuropathic pain levels secondary to peripheral neuropathy, focal nerve injury, post-herpetic neuralgia, spinal cord injury, central post-stroke pain, and multiple sclerosis although the authors concluded that a high-dose therapy was more efficacious than low dose therapy [13]. Similar combinations have been used to treat neuropathic pain in military polytrauma patients with success. Medications are an important part of treatment; however, many experts advocate an interdisciplinary approach including PT, OT, and psychology services to achieve the best functional outcome [14].

This case describes a cause of CRPS Type II that has very rarely been reported in the literature. Our patient was very complex with multiple orthopedic and neurologic injuries and had an unusual cause of a well known pain syndrome. Fortunately, we were able to make an early diagnosis and initiate treatment that resulted in significant improvement. We believe this case demonstrates three important principles. First, while CRPS Type II commonly occurs in the setting of mononeuropathy, it should probably be considered in neurologic injuries as varied as plexopathy, radiculopathy, and other lower motor neuron injuries. Second, early diagnosis and treatment of CRPS is crucial, before central pain sensitization occurs and function is lost. Finally, when managing polytrauma patients with complex injuries, secondary causes of pain can be easily overlooked or attributed to routine musculoskeletal pain. Nonetheless, it is important to fully evaluate and consider a broad differential diagnosis in each pain complaint.

Conflict of Interest

The authors have no financial disclosures or other relationships that may lead to a conflict of interest.

References