Case Reports

Reflex Sympathetic Dystrophy After Overuse: The Possible Relationship to Focal Dystonia

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Abstract—We have seen four musician-patients with reflex sympathetic dystrophy (RSD) that followed the muscle-tendon overuse syndrome. Two of these patients responded to minimal treatment that included oral corticosteroids, vigorous physical therapy, and transcutaneous electrical nerve stimulation. A third required a brachial plexus block before symptoms began to resolve. The fourth had a spontaneous resolution, but developed a focal dystonia as the RSD improved. The clinical features, treatment, and pathophysiologic mechanisms that cause RSD are reviewed. RSD may be more common after overuse than previously suspected, and early diagnosis and vigorous treatment are important to prevent complications. Based on our own case and other published examples of dystonia following RSD, we suggest that the similarities in the physiology of the two disorders make it reasonable to hypothesize that RSD is a sensory analog of focal dystonia. *Med Probl Perform Art* 4:114–117, 1989.

Reflex sympathetic dystrophy (RSD) is a term used to describe a syndrome of severe burning dysesthetic pain, hyperesthesia, and disturbances of sympathetic nervous system function manifested as altered sweating, hair and/or nail growth, edema, and mottled skin. Unless aggressive treatment is instituted, dystrophic changes such as osteoporosis and ultimately atrophic changes including pericapsular fibrosis and contractures of joints, severe osteopenia, skin, and fingertip atrophy may develop.[1] Syndromes indistinguishable from RSD have been reported with a variety of other names.[1] Until recently, late complications involved connective tissue, skin, muscle, and bone, but recent reports suggest that movement disorders, including focal dystonia, may be encountered.[2–4]

The earliest descriptions of RSD were reported by Mitchell, who observed Civil War soldiers with gunshot wounds affecting peripheral nerves.[5] Reflex sympathetic dystrophy is now known to follow virtually any kind of injury, including some so minor that details of the incident may be difficult to remember. Children and adults of either sex may be affected.[1]

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We have treated four patients in whom the disorder followed overuse. Although the florid syndrome should be easily recognized, more subtle forms are less easily identified. All three of the patients who had been seen by prior physicians had not been diagnosed as having RSD, and appropriate treatment had been delayed. One of them developed a focal dystonia as the RSD resolved.

CASE REPORTS

Case 1

A 12-year-old female pianist developed pain in her left hand while preparing a Haydn piano concerto. The cadenza featured rapid octave scales, played with the left hand, that were problematic because her hands were still somewhat small. Octaves required a stretch but she could reach a ninth. She practiced the piano about 2.5 hours per day and also played the viola in her school orchestra. The initial symptoms were paresthesias and pain on the ulnar half of the dorsum of the left hand. About 2 weeks after the onset of symptoms she noted pain in the wrist near the pisiform bone. She continued to practice and the pain worsened to the point where all uses of the hand were precluded. She

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stopped playing the viola because of the pain and improved somewhat during the next 5 days without any therapy.

On examination, tenderness of extensor and flexor muscle bellies and tendons serving the left ring and small finger was noted. The flexor carpi ulnaris tendon was the most severely affected. Contraction of these muscles increased the pain, which was maximal with forced abduction of the small finger. The muscle and tendon of the extensor pollicis longus was also affected. There were no sensory abnormalities. A diagnosis of musculotendinous overuse was made, and treatment with complete rest, ibuprofen 400 mg tid, and heat was recommended. After 5 days, there was no further improvement, and she was fitted with a resting splint.

The pain grew worse with use of the splint, spread proximally up the arm to the elbow and shoulder, and became burning and dysesthetic. The pain was associated with swelling and the emergence of an erythematous reticulated mottled rash on her left forearm, wrist, and hand. X-ray films of the wrist, a complete blood count, erythrocyte sedimentation rate, antinuclear antibody, and rheumatoid factor were all normal. A rheumatological consultant agreed that the patient had reflex sympathetic dystrophy, and prednisone 20 mg po daily was prescribed. Vigorous physical therapy emphasizing hand mobility was supplemented by transcutaneous electrical nerve stimulation (TENS) and fluidotherapy with little effect.

Three weeks after the diagnosis of RSD was made, the patient was referred to a pain center. A stellate ganglion block produced Horner's syndrome, but failed to relieve her symptoms. Several days later the upper portion of the cervical brachial plexus was injected with bupivacaine, which relieved the pain immediately, followed by steady improvement during continued physical therapy. The patient was lost to follow-up until about a year after onset of the problem.

The patient had experienced nearly complete resolution of symptoms, but had residual complaints of shooting pains in the affected arm associated with playing and other activities. These symptoms worsened during the subsequent 3 weeks and she again experienced dysesthetic sensations on the radial side of the index finger. On examination there was a suggestion of atrophy of dorsal interosseus muscles of the left hand and incomplete voluntary flexion of all fingers and extension of the wrist. A TENS unit was prescribed and physical therapy was reinstated. She was playing the piano about 45 minutes per day.

Case 2

A 33-year-old right-handed female pianist sought help because of pain in both index fingers. Two and a half years earlier she was diagnosed as having triggering of her right thumb and was injected with steroids, which produced pain that subsided over a month. As the pain resolved, the patient developed back pain that she attempted to relieve with forceful pressure on the lumbar paraspinal muscles applied with both index fingers for 30 seconds, a maneuver she repeated at least 50 times per day. This led to the development of pain in both index fingers. She was seen by an orthopedic surgeon who injected her right index finger with corticosteroids at the metacarpophalangeal joint and placed the finger in a splint. The pain worsened and became burning and dysesthetic. Her hand became swollen and the skin became shiny, thin, and blotchy in appearance. Blood tests for rheumatologic disease were negative. X-ray films of the hand were normal. Because of her inability to play the piano and budgetary considerations, she lost her university teaching position.

The patient's symptoms decreased in severity, but she continued to be unable to play. For the 6 months prior to her evaluation she reported increases in pain, particularly in the right index finger, whenever she attempted to play, accompanied by sensations of mild weakness.

On examination, there was some mild weakness of the superficial and deep finger flexors of both index fingers and weakness of the volar interosseous muscles inserted on the index fingers. The sensory examination of the hands was normal and there was no Tinel sign on percussion of median or ulnar nerves. Phalen's test was negative. With the exception of rather marked ulnar deviation of the right hand in most positions, examinations while playing were unrevealing. There was no evidence of focal dystonia.

A diagnosis of residual RSD associated with overuse was made and physical therapy was started, focusing on mobilization and strengthening the hands. A TENS unit was also used. X-ray films of the hands were again normal. During therapy, the exercises and stretching to relieve the RSD exacerbated the overuse, and there was little improvement. The right index finger (the site of maximal pain) was splinted and the patient was given prednisone 20 mg po daily and amitriptyline 35 mg po at bedtime. A week later there was substantial improvement. Medications were tapered and discontinued after 3 weeks.

Three months after her initial visit, the patient was able to play for 2 hours without significant problems and auditioned successfully for a new position. She continues to use the TENS unit intermittently, warm up carefully, rest during practice sessions, and apply ice after practicing.

Case 3

A 50-year-old right-handed female pianist experienced pain in her right forearm which had begun 4 years previously when she had returned to school to begin a major in piano performance. Soon after resuming a full schedule of practicing and playing, she experienced pain in the extensors of the right wrist and fingers. Initially the pain resolved when she stopped playing, but, as time passed, the pain became more severe, lasted longer, and was associated with other uses of the hand.

Three months prior to her evaluation she sought medical advice. A nonsteroidal anti-inflammatory agent and physical therapy consisting of ice and heat applications and ultrasound were prescribed but had little effect. After play-

ing for her fall-semester juries, she stopped playing completely. Two weeks later corticosteroids were injected into the region of the lateral epicondyle of the right elbow, and her right wrist and elbow were immobilized in a plaster cast for 3 weeks.

While in the cast, she noted the onset of burning dysesthesis over the radius which became progressively more severe. When the cast was removed a week prior to her evaluation, the dysesthetic pain continued to increase in severity to the point where she stopped using the hand for everyday activities. She noted that the light touch of fabric on the affected skin accentuated the burning pain.

On physical examination, she had slightly reduced mobility of the right shoulder and she failed to extend the right elbow fully. There was incomplete flexion of all fingers of the right hand, and extension of the right wrist was limited. The skin over the right radius was darker than on the left and hairs were slightly longer. Mild atrophy of flexors and extensors of the right wrist and fingers was noted. Sensation to light touch and pinprick was very slightly reduced on the distal radial side of the forearm: sensory stimuli evoked dysesthetic sensations.

Prednisone, 20 mg po daily, physical therapy to remobilize joints and strengthen the right hand, and a TENS unit to desensitize the skin were started. Three weeks later, there was a marked improvement and she began to play again.

Case 4

A 22-year-old right-handed male electric guitar player was seen on only one occasion for multiple complaints related to the left arm. Two years earlier he had just completed his first recording and had concentrated on one piece that involved extensive use of a first inversion E chord that required a very long stretch of the small finger of the left hand. On the morning after the recording session he awoke with pain in the radial side of the small finger. The problem worsened over several days as his finger became swollen, and a reddish mottled discoloration appeared in the skin. He sought medical attention and was told he had tendinitis and was treated with a nonsteroidal anti-inflammatory agent that produced a bronchospastic reaction. During the next several weeks he grew steadily worse as the symptoms spread to the rest of the hand and the pain became burning and dysesthetic, radiating to the elbow. Additional therapeutic modalities tried included the use of heat, ice, ultrasound, and ultimately a corticosteroid injection into the web space between the thumb and index finger. By that time he was unable to use his hand for any purpose because of the severity of symptoms.

The patient then began to improve slowly and was later seen by a hand surgeon, who found pain and tenderness at the left cubital tunnel. An ulnar neuropathy was suspected and a long arm splint was applied. The pain at the cubital tunnel associated with a positive Tinel sign persisted, but an EMG and nerve conduction studies were normal.

He began to play again, but noted rapid fatigue of the left hand. For the 6 months prior to his evaluation he noted an uncontrolled tendency of the small finger to flex while playing, which led to inaccurate, imperfectly played notes and marked decrements in his playing ability. At the time of his visit he indicated that he held the finger in a position of forced flexion to keep it out of the way. He developed complex re-fingerings and used the ring finger in place of the small finger whenever possible.

The patient is Caucasian and indicated that he had never taken any neuroleptics or used drugs for recreational purposes. He had a prior negative rheumatologic evaluation and had no current symptoms of rheumatologic disease or Wilson's disease.

On physical examination there was no Kayser-Fleischer ring and pertinent findings were restricted to the left arm and hand. The skin had a brown discoloration at the site of the steroid injection. A very mild generalized weakness of most muscles in the arm was slightly more marked in muscles innervated by the ulnar nerve. The left first dorsal interosseous muscle was smaller than the right. A positive Tinel sign was elicited in an ulnar nerve distribution at Guyon's canal and the cubital tunnel where slight tenderness persisted.

A focal dystonia, characterized by flexion of the small finger of the left hand, was noted when he played. Trihexyphenidyl therapy was refused.

DISCUSSION

The case histories presented illustrate typical features of the musculotendinous overuse syndrome complicated by the development of RSD. The diagnosis of RSD was based on a history of pain that increased in spite of rest and that became burning and dysesthetic with an intensity far in excess of that expected from the initial overuse problem. In addition, abnormal vasomotor responses, edema, alteration in skin color, accelerated hair growth, loss of joint mobility, and spread of the syndrome away from the site where the injury occurred were noted as described in the individual case reports. Only the first patient developed evidence of dystrophy.

The diagnosis of RSD is based on clinical features. X-ray films may reveal evidence of the antecedent injury or evidence of rapid bone turnover.[6] Although scintigraphy with technetium 99m has been advocated as a useful adjunct, there is usually little doubt about the diagnosis.[1,6,7] Nerve blockade may be useful both diagnostically and therapeutically.

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The mechanisms that produce RSD are unknown; however, a thoughtful hypothesis has been proposed by Devor.[8] He suggests that tissue injury damages or transects the axons of sensory nerves. During the recovery process the exposed regenerating surface of the axon gains a higher than normal number of alpha-adrenergic receptors that contribute to the development of abnormal electrical properties of the nerve, leading to spontaneous nerve depolarizations and the development of ectopic pacemaker capabilities. This abnormal barrage of neural impulses has secondary effects on sympathetic reflexes in the spinal cord and also alters other aspects of the central processing of sensory information.[9] As a result of these changes, the central nervous system receives neural impulses that originate because of altered chemosensitivity and mechanosensitivity of neurons and not as a consequence of physiologic stimuli. Aberrant sensory processing produces the sensation of pain, and altered sympathetic reflexes produce the sympathetic nervous system abnormalities characteristic of RSD. This theory not only accounts for the signs and symptoms of RSD but also for the success of the therapies directed at altering the sympathetic nervous system.

Successful treatment of RSD may be complex and arduous for both the patient (including the family) and the physician. Uncomplicated mild cases may resolve with simple analgesics and intense physical therapy. Most patients require more aggressive therapy, and many successful approaches have been reported. Physical therapy is a component of all regimens. The goals of therapy include desensitization, often employing TENS, and the maintenance of mobility and strength to avoid contractures, bone demineralization, and other dystrophic and atrophic complications.

Pharmacologic therapies are diverse and include anti-inflammatory agents, corticosteroids, and analgesics as well as a variety of agents that act on the sympathetic nervous system. These include propranolol, reserpine, or guanethadine, which may be given orally or in the form of a Bier block—a regional anesthesia technique employing an intravenous injection of an agent distal to an occlusive tourniquet. Other forms of regional anesthesia directed at the sympathetic nervous system or its ganglia such as stellate ganglion blocks or paravertebral sympathetic ganglion blocks may be necessary. When these modalities provide only transient relief, surgical ablation may be indicated to provide longer lasting effects. Regional anesthesia techniques should be performed by experienced individuals, preferably in a tertiary pain care center.

Peripheral trauma may lead to the development of movement disorders, including focal dystonia.[2–4] Almost half of all posttraumatic dystonias have been preceded by RSD that antedated the movement disorder by as much as 8 years.

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In many cases this interval is much shorter, and the two disorders may develop simultaneously.

Although the pathogenesis of RSD and focal dystonia are unknown, there are interesting parallels that may be of importance. Physiologic studies of patients with focal dystonias have shown abnormalities of reciprocal inhibition of antagonist muscles that suggest lesions in Ia interneurons in the spinal cord.[10] In addition, there may be additional abnormalities in the control of the affected Ia neurons attributable to disturbances in the basal ganglia and supplementary motor cortex.[10] These altered neural control systems may be analogous to the alterations in sensory and autonomic impulse-processing in RSD hypothesized by Devor.[8] Thus focal dystonia may be the motor system analog of RSD, which is a syndrome of disturbed sensation.

All four of our patients had symptoms and signs of overuse that preceded the development of RSD. Three of our four patients had been seen by other physicians who had not recognized the symptoms of RSD; the fourth had not sought prior health care for her problem. This suggests that RSD, like other problems that affect musicians, may be underdiagnosed by those unfamiliar with the special needs and demands of performers.

The development of RSD in our patients also emphasizes the need for a better understanding of the pathophysiology of the overuse syndrome. Although recent pathologic studies are an important step in this direction,[11] substantial improvements are needed before better treatment and prevention of this pervasive problem and its complications are possible.

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