

Reflex Sympathetic Dystrophy: A Case of Total Body Pain

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Reflex sympathetic dystrophy (RSD), also known as complex regional pain syndrome type I (CRPS I), is a chronic condition characterized by burning and aching pain; hyperesthesia and allesthesia; motor disturbance and soft

clear, studies have shown that RSD is associated with many different medical conditions including trauma, drugs, and unidentified precipitating factors.³ Previously reported RSD patients had localized pain or pain involving only limited areas of the body.⁴ Systemic cases are rare. The following report examines a patient with

- Harrington rod fusion, T8 through sacrum, for scoliosis and spinal instability in 1978
- Harrington rod removal in 1980
- restrictive lung disease and respiratory insufficiency since her late 20s, diagnosed in 1983
- galactorrhea-amenorrhea syndrome between early 1992 and 1993.

During the examination, the response to mild sensory stimuli may produce severe pain.

tissue change; vasomotor and autonomic changes; and psychosocial disturbance.¹ Early signs and symptoms of the disease usually include inflammatory signs in the affected limb and muscular paresis with easy fatigue.²

RSD can be difficult to diagnose and often requires excluding other conditions that produce similar symptoms. A thorough history and neurological examination is important for accurate identification. During the examination, the practitioner may notice that response to mild sensory stimuli produces severe pain. Physical examination involves observing the skin color and temperature; swelling and vascular reactivity; overgrown and grooved nails; swollen and stiff joints; and muscle weakness and atrophy. Other conditions are ruled out with appropriate testing that may include a magnetic resonance imaging scan, a full laboratory panel, and electrophysiological studies of the nerves and muscles.

While the etiology of RSD is not

global pain that affected all of her extremities and had multiple medical complications associated with RSD.

■ Patient Characteristics

The patient, a 52-year-old African-American woman, was diagnosed with RSD in 1992. She had several medical conditions before her diagnosis throughout a specific stretch of time, which may have predisposed her to RSD. These included:

- a long history of pain in her back and legs since 1973, managed with a variety of indicated regimens
- a fracture of the sacral spine in 1973 at age 19
- scoliosis, cervical, and lumbar spinal stenosis diagnosed in 1973
- arachnoiditis since 1974 from complications of several myelograms
- laminectomies from L4 to S1 for herniated discs in 1975
- facet denervations by ablation and epidural injections in the lumbar and cervical spine for pain treatment in 1976

In 1992, she began to experience burning pain, excessive hair growth compared to the opposite extremity, and swelling in the left arm, which were considered early signs and symptoms of RSD. She was diagnosed with brachial plexitis and treated with physical therapy that included acupuncture, desensitization procedures, warm compresses, and range of motion exercises. The treatments were ineffective, and the patients' symptoms advanced to her right upper extremity and bilateral lower extremities over a period of 1 month. Later, the pain spread to her torso, including her back, abdomen, and chest.

The pain was described as a deep aching and burning sensation. She experienced hypersensitivity with marked allesthesia of the skin in local areas. She also suffered tremors, myoclonic jerks, and mood changes that included anxiety and depression. Despite intensive treatment, her pain syndrome became chronic. She developed muscle atrophy and contractures of the extremities, especially at the fourth and fifth fingers of her hands. She has been unable to walk since 1993.

Continuous abdominal pain occurred, along with ileus, which re-

quired total colectomy with ileorectal anastomosis in 2002. Dumping syndrome emerged soon afterwards. Since the total colectomy, she developed partial ileus of the small bowel, and menopause began in 2000. Furthermore, the patient has a history of angina and suffered a myocardial infarction (MI) in 1996.

At present, the patient still has constant, generalized pain, chronic ileus, muscle spasms, and atrophy of all four extremities. She uses an electric wheelchair for mobility and has lived in a nursing home since 1993.

■ Disease Management

Currently, the patient's pain is reasonably controlled with a self report of pain between 4 and 6 out of 10. Pain is treated utilizing a multimodal approach, which includes morphine (Avinza), gabapentin (Neurontin), prednisone, oxcarbazepine (Trileptal), and lidocaine via patch. Various medicines are used for other conditions (see Table: "Medical Conditions and Associated Medications Used"). Treatments used in the past included other analgesics, tricyclic antidepressants, sympathetic blocks, desensitization techniques for pain, and therapies for preventing muscular atrophy or contracture.

In 1993, the patient underwent multiple sympathetic nerve blocks for both upper and lower extremities and surgical sympathectomies for upper extremities without significant change. Her pain was reported as constant, with a self-reported severity averaging 8 to 10 out of 10 prior to her current treatment regimen.

■ Multiple Complications

RSD is a rare syndrome that is difficult to diagnose and is often initially missed by practitioners not familiar with it. Its incidence is unknown both in the United States and internationally. The reported prevalence of RSD

Medical Conditions and Associated Medications Used	
Medical Conditions	Medications Used
Clonic spasms	Clonazepam and tizanidine
Hypertension	Nifedipine
Osteoporosis	Risedronate, vitamin D and calcium
Diffuse esophageal spasms	Nitroglycerin patch
Asthmatic bronchitis	Salmeterol discus, flunisolide inhaler, and theophylline
Gastrointestinal upset/pain	Esomeprazole, simethicone
Atypical bipolar disorder	Olanzapine
Hypothyroidism	Levothyroxine

is 1% to 2% after various fractures, 2% to 5% after peripheral nerve injury, and 7% to 35% in prospective studies of Colles fracture. The likelihood of developing RSD is higher if the lesion is distal or the nerves are affected.

RSD affects all races, with no racial predilection, and both sexes (the female to male ratio is 2:1). The mean age of RSD patients is approximately 40 to 42 years. The highest incidence of the disease is in adults aged 40 to 49 years. RSD has been found occasionally in children, but its incidence is much lower than in adults. This patient is consistent with those statistics.⁵ She has all the common manifestations of RSD, including severe pain, altered sensation, motor disturbance (spasm, restriction), vasomotor or autonomic changes (skin color, temperature, swelling and edema, and sweating or trophic changes), and altered psychosocial functions.

As with other reported cases, the course of this patient's disease included three stages: 1) acute or hyperemic; 2) ischemic; and 3) atrophic. Although pain in the patient's back and neck began in 1973 following the fracture of her sacral spine, definite diagnosis was not made until 1992, when she began to experience burning

pain, hair growth, and swelling in the left arm. The key differential diagnosis of this case is brachial plexitis, which was made prior to RSD diagnosis.

Her total body pain, however, is unique, as are the multiple severe complications resulting from her condition. While diagnostic criteria for RSD are mainly focused on many different aspects of localized sensory and autonomic features,³ this patient manifests pain and motor dysfunction involving the entire body except the head and face. A Medline search from 1994 showed only three cases with involvement of more than two limbs, and only one other case involved the whole body.⁴ Additionally, investigators searched PubMed, which covers the remaining years after this literature review, and found no reported cases involving the whole body. This case expands the diagnostic criteria for CRPS I and challenges the validity of the term "complex regional pain syndrome."⁶ While indeed complex, RSD is not necessarily localized. Moreover, in this particular patient, RSD involved the entire body and had consequences for many systems. While a causal relationship between RSD and her varying complaints cannot be proven, they occurred after the confirmative diagnosis in a gradual manner with no additional explana-


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tion. These nontypical RSD complications are believed to include: chronic intestinal pseudo-obstruction (resulting in removal of large intestine) and ileus of the small bowel, restrictive lung disease with respiratory insufficiency; angina, MI, transient ischemic attack, hypertension, hypothyroidism, multiple sebaceous cysts on the back, and occasional bouts of anxiety and depression, which greatly improved with better pain control.

The pain associated with RSD is difficult to manage, but can generally be controlled at a tolerable level with multiple drugs and other therapies. At present, the patient is able to attend social activities, including concerts and church with the use of an electric wheelchair. She can sleep fairly well. Because there is no uni-

form treatment protocol for RSD patients with total body pain, medications and their combinations were researched. Minimal effective dosages were used, which, although nominal, is beyond regular uses.

■ The Verdict

Ultimately, the patient was diagnosed with total body RSD. This implies that pain can involve more than just assorted extremities or regions. The disease became chronic and caused pain, physical disability, and complications involving multiple systems. To improve her treatment result, multiple drugs and therapies are needed. For more information, clinical practice guidelines are available from the International Research Foundation for RSD/CRPS at <http://www.rsdfoundation.org>. 

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