REFLEX SYMPATHETIC DYSTROPHY SYNDROME

by Stephen J. Miller, DPM

Under normal circumstances, the human body will heal in an orderly fashion following trauma or disease. Occasionally, this does not occur and the patient will suffer from an unexpected degree of pain, swelling, stiffness and dysfunction, even though appropriate treatment has been provided.

This complex of symptoms is known as Reflex Sympathetic Dystrophy Syndrome or RSDS. It involves an abnormal response in the sympathetic nervous system which is thought to be the cause of the disorder. In RSDS, there is not only an increase in the sympathetic nervous system activity, but also an abnormal reflex that allows this increase in stimulation to perpetuate itself. The common denominator is pain in an extremity. The characteristic abnormality is vasomotor instability.

By definition, RSDS is a syndrome of pain, hyperesthesia, vasomotor disturbances and dystrophic changes that usually improves with sympathetic denervation. Causalgia is that presentation of the disorder that occurs after a major nerve injury, even though the majority of the presentation is the same. Causalgia is considered to be the severest form of RSDS.

Experimental and clinical evidence obtained over many years has provided significant support to the contingent that although the causative factors may be different, the pathophysiology, presumed mechanisms and symptoms are similar in that sympathetic interruption early in the course of this syndrome provides effective relief of pain and disappearance of the rest of the symptoms.

TRIGGERING EVENTS

RSDS has been reported in adults, adolescents and children. Most often, there is an identifiable "triggering event." However, one paper has stated that up to 35% of presenting cases have no identifiable initial cause. Almost half of all cases will present in the lower extremities.

The most common identifiable cause is some type of trauma such as accidental injury, surgical and other iatrogenic injury, or micro or macro trauma associated with certain occupations. RSDS has also been associated with diseases such as myocardial infarction, cerebral vascular accidents, and a variety of neurological disorders as well as malignancies.

IDENTIFYING THE SUSCEPTIBLE PATIENT BEFORE SURGERY

There are three factors which must be present at the same time before a person can develop RSDS. These include: a persistent painful lesion (traumatic or acquired); a diathesis and; an abnormal sympathetic reflex.

The diathesis, or pre-disposition, is a susceptibility to a certain disease state. With respect to diathesis, it is believed that there are two types of patients. The first type is the hypersympathetic reactor - the patient with evidence of increased sympathetic activity such as chronically cold or sweaty feet, summer or winter. Such a patient may exhibit pallor, slight cyanosis or coolness of the toes in spite of palpable pulses. The pulses themselves may be thready and weak. There may be a history of fainting, blushing easily or migraine headaches.

The second type of patient is described psychologically as having an inadequate personality. This patient is fearful, suspicious, and emotionally labile, and tends to be a chronic complainer. This type of patient can be difficult to treat because he often tries to control his own

treatment and habitually blames others for his problems.

In general, patients with such a hypersympathetic diathesis have been suggested to have some common characteristics. These include a background of vasomotor instability together with hyper-emotional temperament, both being prerequisites to developing RSDS. Since these patients have a physiologic makeup that allows their sympathetic nervous systems to become hyperactive, they have also been described as sympathetic reactors or hypersympathetic reactors.

Steinburg, the eminent podiatric diagnostician, recognized a sympathetic sensitive patient which he termed Raynaudian. This is a patient with a centrally induced hypertonicity of the sympathetic nervous system. Another disorder with the common trait of vasomotor instability is acrocyanosis frigida, also known as pernio or chilblains, the mildest form of cold injury. Patients with any of these conditions have a susceptibility to developing RSDS, especially following trauma or surgery.

CLINICAL PRESENTATION AND COURSE

Pain in the extremity is the one common denominator to all stages and for all patients affected with RSDS. The pain is characteristically of a burning or severely aching nature, usually superficial in perception and occurring in the periphery of the extremity. In the lower extremity it is usually more intense in the toes and sole of the foot.

The pain is often accompanied by *allodynia,* pain due to a noxious stimulation of normal skin or painful sensation to touch. It can also be associated with *hyperpathia,* the delayed over-reaction to a stimulus, especially a repetitive stimulus. The pain is usually out of proportion to the injury or precipitating event (known as hyperalgesia) and will progress outside the dermatome of the involved nerves. As a result, the patient suffers greatly and tends to protect the affected extremity. It becomes more intractable with time as long as it remains untreated.

Unless severe pain disappears spontaneously or is relieved with treatment, it usually spreads proximally to involve the entire extremity in time. This devastating ability to spread in space and increase in time is one of the most distressing characteristics of causalgia and the severe forms of RSDS.

THE STAGES OF RSDS

RSDS has been classified into three different stages based upon objective, radiographic and clinical signs. Stage I, or the acute stage, presents constant pain which may start immediately or several weeks after the precipitating event. It may be of a burning or aching nature and is not limited to a specific dermatome. It is exacerbated by cold and usually will be accompanied by diffuse swelling with use.

Since the vasomotor instability can present at either end of the vasoconstrictive or vasodilatory spectrum, one may see warm or cool skin, erythematous or waxy white discoloration of the skin, as well as hyperhidrosis or anhydrosis. Trophic changes are not yet apparent although deossification sufficient to be visualized on radiographs may present within four to six weeks.

Stage II, or the dystrophic stage, is characterized by the edema becoming more indurated although the pain persists. The skin is usually cool, pale, and may take on a laquer look. Diffuse osteoporosis on x-ray is visible and the nails may become cracked, brittle and ridged. In this stage, the patient is still capable of improvement.

Stage III is known as the atrophic stage. Now the intractable pain spreads proximally to involve the entire limb and trophic changes lead to irreversible tissue damage. There is decreased dermal blood flow, thin and shiny skin, and the fingertips become wasted and pointed as the fat pads atrophy. The fascia becomes thickened and joints will stiffen even to the point of ankylosis. Radiographs show a marked demineralization. Most of these changes are irreversible at this stage.

It is difficult if not impossible to predict the course of RSDS once it has been identified. The disorder may be self-limiting but not without much agony. There is general agreement that early recognition and treatment will realize a much better response to treatment, thus limiting the duration of the syndrome.

Patients afflicted with RSDS may seem emo-

tionally unstable, anxious, and socially withdrawn. This is thought to be more a result of the frustration of the persistent symptoms rather than a predisposition to RSDS.

Unrelenting pain, distress and discouragement can lead to chronic invalidism, drug addiction, psychiatric treatment and even suicide. Emotional sequelae and the disparity between the degree of pain and the physical examination may mislead clinicians to believe the pain is psychogenic. When added to misguided therapeutic efforts at relieving the intractable pain, this serves only to further aggravate the psychologic symptoms. The increased anxiety then increases sympathetic discharge which exacerbates the pain, and the vicious cycle continues.

TREATMENT

The importance of early diagnosis and treatment of RSDS have been widely recognized as critical to successful resolution of this syndrome. Best results in treating RSDS are obtained when the diagnosis is made early and treatment is begun in Stage I, or at the very latest, Stage II.

The most important initial factor in treatment is to prevent the patient from protecting the limb unnecessarily. This seems to perpetuate the reflex nature of the disorder. Thus, prompt initiation of physical therapy is essential and the orders should be specific for any combination of the following: hydrotherapy, underwater ultrasound, range of motion exercises, joint distraction, active and passive stretch, deep friction massage, desensitization, contrast baths, paraffin baths, gait training, jobst pump, as well as a home exercise program.

Pain control is essential, with the goal of getting the patient through his or her therapy program. Narcotics should be avoided, but tranquilizers can be helpful to subdue the emotional turbulence that can tend to perpetuate the pain cycle.

Oral steroid therapy for a specific period can be quite efficacious. Nonsteroidal anti-inflammatory drugs are found to be helpful in the control of pain thought to be due to trophic changes or inflammatory changes with the bone. Alpha-adrenergic blockers that provide sympatholytic activity, as well as calcium channel blockers that interfere with peripheral vasospastic response, are also effective.

Chemical sympathetic blockade is the foundation of remarkable and sustained relief in a majority of patients. It can be provided through paravertebral blockade of the sympathetic ganglia, regional intravenous blockade by way of the Bier block or peripheral nerve blockade by itself. None of the sympathetic blockades should be undertaken without already having initiated the patient on aggressive physical therapy and other measures.

Keep in mind that much remains to be learned about this potentially devastating disorder. It is always important to seek outside consultation when RSDS is suspected. For your part, remember that a detailed and compassionate explanation relative to the pathophysiology of the disorder can result in great relief to any patient.

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