DIAGNOSTIC MODALITIES IN REFLEX SYMPATHETIC DYSTROPHY

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INTRODUCTION

Reflex sympathetic dystrophy (RSD) is a syndrome characterized by pain, swelling, hyperesthesia, trophic changes, and vasomotor instability. The syndrome is secondary to an abnormal hyperactive state of the sympathetic nervous system following injury to an extremity. Precipitating factors include soft tissue injuries, infections, fractures, sprains, dislocations, and operative procedures. The common mechanism underlying RSD may be injury to either central or peripheral neural tissue, including peripheral nerve branches. Reflex sympathetic dystrophy is the currently accepted term for this disorder, but many terms have been used, including causalgia, minor traumatic dystrophy, algoneurodystrophy, Sudeck's atrophy, and shoulder-hand syndrome.

The diagnosis of RSD is often difficult to establish. It is primarily a clinical diagnosis, but other diagnostic modalities are becoming available. Standard roentgenograms have been the main diagnostic aid in the recent past. Unfortunately, the characteristic osteoporosis of RSD is a relatively late finding. It is generally accepted that early recognition and prompt treatment provide the greatest opportunity for a successful outcome. A delay in diagnosis can result in severe physical and psychological problems. Early recognition with the aid of currently-used diagnostic studies, will facilitate diagnosis and direct treatment.

CLINICAL DESCRIPTION AND COURSE

Symptoms of RSD may begin gradually, days or weeks after the injury, or may manifest within a few hours. Signs and symptoms of RSD include pain out of proportion to the initial injury, which continues and increases in severity with time. The pain is characterized as a burning or aching pain which may be localized early, but becomes diffuse, nonanatomic, and outside dermatoma distribution. Other signs include edema, hyperhidrosis, and increased hair and nail growth of the affected extremity. Late signs include atrophy of skin, muscles, and bones, as well as joint stiffness from disuse.

Reflex sympathetic dystrophy has been classified into three stages. Stage 1 (Acute-first three months) consists of pain, edema, hyperthermia, and increased nail and hair growth. Stage 2 (Dystrophic-three to nine months) consists of pain, induration, edema, cool skin, hyperhidrosis, with a livedo reticularis appearance or cyanosis. Stage 3 (Atrophic-symptoms in excess of nine months) is characterized by pain spreading proximally, shiny skin, flexion contractures, bony demineralization, and ankylosis.

The frequency of RSD after peripheral nerve injury ranges from 1% to 15%. Many mild forms in the lower extremity probably go undiagnosed because the forced ambulation of daily living may be sufficient physical therapy to rehabilitate the extremity.

PATHOPHYSIOLOGY

In a normal response to trauma, vasoconstriction occurs in an attempt to control bleeding and prevent excessive swelling. Following the initial injury, vasodilation occurs to aid in healing. If vasoconstriction persists, painful, localized ischemia may occur, and thus, reflex sympathetic dystrophy begins.

There are numerous theories which attempt to explain the pathogenesis of this disease. Livingston postulated that increased afferent impulses arising from a peripheral nerve injury cause an abnormal state of heightened activity in the internuncial neuron pool within the spinal cord. This, in turn, propagates continuous stimulation of sympathetic nerve fibers producing vasoconstriction and persistent ischemia, further accelerating the vicious cycle.

More recently, Melzak and Wall have suggested that certain cells in the substantia gelatinosa of the spinal cord act as a computer, directing incoming afferent impulses. The large myelinated fibers act to inhibit the control system. The small afferent sympathetic fibers tend to stimulate the substantia gelatinosa, thus increasing the transmission of pain impulses to the brain.

DIAGNOSIS

The diagnosis of RSD is primarily clinical. It is important when evaluating any patient with long term pain to rule out other somatic causes of pain, such as fracture-dislocation, infection, neoplasm, other neurologic causes (such as herniated nucleus pulposus), and vascular compromise (such as arterial occlusion).

Roentgenographic studies were the first used to confirm this disorder. Findings include patchy demineralization in cancellous bone. These changes are not specific for RSD, nor always present. This demineralization has been reported as a positive finding in 30% to 70% of RSD cases.

Bone Scintigraphy

Bone scintigraphy is now the foremost diagnostic adjunct for RSD. It is both sensitive, and specific for the diagnosis of this syndrome. Three-phase radionuclide bone scans of technetium (Tc 99m) demonstrate a pattern of diffuse increased tracer with increased periarticular uptake in the involved extremity. The three-phase bone scan technique consists of Phase I, a radionuclide angiogram; Phase II, a blood pool phase; and Phase III consisting of delayed images obtained three to four hours after radionuclide injection.

Kozin et al., in their study of 64 patients, showed scintigraphy to have a sensitivity close to radiography (60% vs. 69%) but a significantly greater specificity (92% vs. 79%). In Mackinnon and Holder's study of 145 three-phase bone scans of the hand, results revealed a 96% sensitivity, 98% specificity, and a 99% negative predictive value by the use of the delayed images alone.

Holder et al., in a prospective study of 51 patients with a presumptive diagnosis of RSD, performed three-phase radionuclide bone scans to support the diagnosis. An additional 100 patients with a variety of foot problems were retrospectively reviewed. Overall, sensitivity in this study was 100%, specificity 80%, positive predictor value 54%, and negative predictive value 100%.

The bone scans of all 26 patients with a confirmed diagnosis of RSD demonstrated a diffuse increased tracer throughout the hind-, mid-, and forefoot with juxta-articular accentuation of tracer uptake on delayed images. Sixteen of the 26 patients had an abnormal angiogram study with increased tracer appearing at least one frame earlier than on the unaffected extremity. Twenty-three of the 26 patients had an abnormal blood pool phase. None of the patients with a confirmed diagnosis of RSD had a normal delayed image after an abnormal angiogram or blood pool phase. In addition, none of the patients in this study (n=151) with a normal scan had RSD.

Thermography

Other diagnostic modalities include electronic thermography, lumbar sympathetic block, intravenous regional chemical blockade, magnetic resonance imaging, and the use of an algometer. Electronic infrared thermography was utilized by Perelman et al. to aid in diagnosis of 4 cases of suspected RSD. The involved, painful extremity demonstrated a decreased temperature in all cases.

Karstetter and Sherman believe that RSD is virtually always accompanied by decreased temperature of the affected extremity. Data indicates that the pain intensity increases in proportion to

the decrease in surface temperature. Limbs normally differ in surface temperature by up to 0.5° C. Thus temperature difference is considered to be significant when there is a discrepancy of at least 1.0° C. The affected limb in RSD is usually about 2.0° C. cooler than the unaffected limb. Cooling associated with early RSD begins distally and gradually progresses proximally.

Sherman et al. found thermography to be an excellent tool for monitoring changes in pain related to variations in surface temperature, such as those occurring during a sympathetic block.

Sympathetic Nerve Blocks

A sympathetic spinal blockade can be helpful as both a diagnostic and therapeutic modality. This technique involves the use of two solutions: saline, as a placebo, and 5% procaine, used as a pharmacologic spinal blockage. If the introduction of saline substantially relieves pain, then the pain is possibly of psychogenic origin. If the procaine block does not relieve the pain, the pain is considered to be psychogenic or malingering. The pain is assumed to be somatic if pain relief occurs with procaine, but the pain returns with pin-prick sensation, because somatic fibers recover from anesthesia faster than sympathetic fibers. If the pain relief continues after motor function and pin-prick sensation have returned and sympathetic functions remain blocked, the pain is presumably sympathetic in origin.

Intravenous Therapies

Various intravenous chemicals have been utilized as diagnostic aids in RSD. Guanethidine and reserpine are sympathetic blocking agents that can be used in an intravenous regional Bier block technique to produce a regional sympathetic blockade. Guanethidine works by displacing norepinephrine in presynaptic vesicles and preventing its re-uptake. Reserpine's mechanism of action interferes with the storage of norepinephrine, gradually causing depletion in nerve endings. Studies vary greatly in the diagnostic and therapeutic effects of these two pharmacologic agents. Pain relief with these modified Bier blocks are good indicators of RSD, but patients may have no response to these tests. These tests may offer an alternative to conventional spinal sympathetic blockade, yet neither guanethidine or reserpine are available in the United States for intravenous infusion.

Magnetic Resonance Imaging

The use of magnetic resonance imaging (MRI) appears to offer no benefit in the diagnosis of RSD. Kock et al. reviewed images of 25 patients selected on the basis on initial clinical findings, and positive finding on scintigraphy for RSD. Seventeen of the 25 patients had a final diagnosis of RSD based on later clinical course. Of these 17, ten patients showed completely normal MR images, six showed only nonspecific soft-tissue changes or bone marrow sclerosis, and one patient showed changes in bone marrow signal. MRI appears to be of little value in the diagnosis of RSD.

Algometer

Bryan et al. in Liverpool, England have begun utilizing an algometer to measure pressure-pain thresholds. In their study of 33 patients with RSD (18 upper extremity and 15 lower extremity), they used a clamp with rubber-tipped jaws attached to a pistol grip handle which was connected to the main body of the instrument by a flexible cable. This device, the Somedic algometer is attached to either the thumb or the hallux, and pressure is increased in a controlled fashion while pressure readings are recorded electronically. The patient is asked to press a button when the perceived sensation changes from pressure to pain. The pressure is recorded at this point. The unaffected side is examined first, followed by the affected side, with the mean of three measurements being recorded.

The study confirmed that there is a significant lowering of the pressure-pain threshold on the affected side in patients with RSD. Bryan et al. found a 100% accuracy in identifying patients with RSD of the upper extremity, but only a 73% accuracy in lower extremity cases.

CONCLUSION

Reflex sympathetic dystrophy is a challenging syndrome to diagnose and treat, especially in patients presenting with unremitting pain in the absence of identifiable pathology. In this setting, it is vital that the podiatrist recognizes the symptoms early in the disease process. While many

clinicians consider a positive response to lumbar sympathetic block to be the standard of reference for the diagnosis of RSD, in many instances the pain or autonomic signs are less evident. In more subtle cases, other diagnostic testing may be performed.

Many diagnostic modalities have been presented. Three-phase radionuclide bone scans might be performed on suspected RSD cases, with careful evaluation of the angiogram, blood pool, and delayed phases. More recently described noninvasive modalities, such as surface thermography and algometry, may offer a simpler means of confirming the diagnosis. Early recognition of the signs and symptoms of RSD will enable prompt treatment to provide the best long term results for the patient.

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