

REFLEX SYMPATHETIC DYSTROPHY SYNDROME (SUDECK'S ATROPHY) AND MIGRATORY OSTEOLYSIS OF THE LOWER EXTREMITY

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Introduction

In practice the physician often encounters disease states and disease complexes which are perplexing yet treated satisfactorily. Often treatment for a set of presenting complaints is performed with satisfactory results without permitting the syndrome to become full blown. Since the condition under treatment might be unknown at the time, a diagnosis may not be applied to the condition. The author believes reflex sympathetic dystrophy syndrome is often such an entity. The entity will be discussed to remind the podiatric physician of its existence and to review appropriate diagnosis and treatment. Also a rare idiopathic form of osteoporosis termed migratory osteolysis of the lower extremity is discussed.

Reflex Sympathetic Dystrophy Syndrome

Reflex sympathetic dystrophy syndrome (RSDS) is a distinct symptom complex which includes pain, swelling, limitation of motion, vasomotor instability, trophic changes, and patchy demineralization. It is most prevalent in middle-aged and older adults although there is no age limitation. Bernstein and associates reported this entity in children (1). Disorders underlying RSDS include incidental trauma, fracture, peripheral neuropathy, osteomyelitis and infection, CNS abnormalities, vertebral degenerative joint disease, and myocardial infarction. The entity was first described by Mitchell and associates in 1864 (2). In 1882, Volkmann described post traumatic rarefaction of bone (2). In 1898, Destot noted osteoporosis following a long term painful ankle sprain (2). In 1900, Sudeck described acute atrophy of bone which he attributed to inflammation (2). In 1901 Kienbock reported acute bone atrophy following trauma (3).

Many terms have been used to describe this condition: reflex dystrophy, symptomatic dystrophy, post traumatic painful osteoporosis, Sudeck's atrophy, Sudeck's syndrome, causalgia, and shoulder-hand syndrome. The terms have added confusion and difficulty in recognizing it as a single entity. Steinbrocker and Argyros have described clinical factors in these conditions and are credited with the collective term of RSDS (4).

Pathophysiology

The pathophysiology to date remains obscure. Involvement of the sympathetic nervous system has been suggested by Leriche and others (2). It is related to the initiation and excitation of abnormal neural reflexes. An irritant focus such as a peripheral nerve injury is regarded as an afferent stimulus. The sympathetic system functions as the efferent branch of the reflex circuit.

The mechanism of the disorder apparently involves widespread disturbances in the internuncial pool of the spinal cord. This entity has a self-aggravating and self maintaining mechanism. Pain impulses entering the spinal cord from the peripheral site of irritation cause a constant re-excitation of the internuncial neurons. Stimuli extend to the anterior horn cells and efferent sympathetics resulting in muscular spasm, edema, and vascular changes which aggravate afferent pain impulses. A vicious cycle ensues and must be interrupted if symptoms are to improve. Livingston has postulated three factors needed to establish the vicious cycle (3):

1. Initial trauma and tissue damage resulting in chronic irritation of a peripheral nerve.
2. Abnormal state of activity in the internuncial neuron center produced by the increased afferent impulses.
3. Continuous and increased stimulation of efferent motor and sympathetic neurons resulting in a variety of peripheral impulses.

Initially after injury a vasomotor reflex spasm occurs. Loss of vascular tone ensues (persistent vasodilatation). This leads to vascular congestion and hyperemia resulting in loss of bone substance. Moberg believes that in the hand and arm there is decreased circulation by interference with the pumping mechanism of the voluntary muscles mediated by inhibition of motor nerves resulting in a frozen hand (2). Improper lymphatic and venous drainage results. The immobility results in edema and local fibrosis of muscles, tendons, and ligaments. This theory is consistent with findings seen in the foot and lower extremity.

Clinical Findings

Severity of clinical findings are unrelated to severity of injury. Vasomotor reactions can be varied. Initially mild pain and edema are noted. There is increased blood flow and tissues feel warm. Plethysmographic studies have shown an average of 30% increase in blood flow. Hyperhydrosis may be present. This may last weeks to months. Later more dystrophic changes appear as a result of vasomotor constriction. These include atrophy, severe cutaneous paresthesias, cold and shiny skin, and joint motion limitation. Patchy discoloration is usually present. Pain is moderate to severe. With time atrophic changes are severe enough that findings are consistent with Volkmann's ischemic contracture. Psychiatric factors play an important role in the condition as reported by Miller and DeTakats (2). Although no one particular personality type has been clearly identified, functional instability and predisposition to RSDS has been associated with obsessive-compulsive, anxious, hypochondriacal, and narcissistic types.

Radiological Findings

The most obvious roentgenographic finding is a mottled rarefaction of bone most evident in the first 4-6 weeks of the condition. Diffuse osteopenia (demineralization) is represented by thinning and dissolution of bony trabeculae. Osteopenia is noted by a generalized decrease in bone density. Peri-articular or juxta-articular involvement causes a relative radiolucency around joints as a result of accentuated demineralization in the cancellous bone of the metaphyseal regions. Cortices show varying degrees of thickness. This results from both periosteal and endosteal resorption giving a scalloped appearance to the bony surfaces. The latter are hard to see on standard radiographs and best seen on fine grain film.

Intracortical areas show resorptive tunneling or striations best seen in the metatarsals. Genant and associates using quantitative analyses, found that there was usually 33% loss of cortical thickness mostly attributed to endosteal resorption (5). As the syndrome continues, arthropathy ensues. This is represented by juxta-articular and subchondral erosions, peri-articular soft tissue swelling, and capsular distention. The latter is identified by prominence of soft tissue around joints and obliteration of normal peri-articular fat planes.

Osteoporosis is a common radiologic finding. Burkhart and Jowsey evaluated the importance of parathyroid and thyroid hormones in mediating bone resorption (6). They concluded that circulating parathyroid and thyroid hormones are essential effectors in disuse osteoporosis and local factors (hyperemia) induce greater sensitivity to the

circulating hormones in the immobilized limb. It is therefore possible, based on this study, that acute osteoporosis of disuse is mediated by local factors such as hyperemia and diminished mechanical stress in conjunction with systemic factors such as circulating parathyroid and thyroid hormones.

Technetium-99 bone scans usually show hot spots in the peri-articular areas, probably as a result of rapid bone turnover from increased local blood supply. Kozin and associates revealed that 69% of the time bone scans in patients with RSDS bone scans were positive and that scintigrams (mTc99-MDP) showed a 92% specificity in patients with RSDS (7). The degree of specificity helps in identifying patients that would be responsive to systemic corticosteroid therapy.

Histopathology

Genant took synovial biopsy specimens and revealed that there was mild to moderate synovial edema, hyperplasia, and fibrosis. Capillary proliferation and mild perivascular inflammatory infiltrates accompanied the above suggesting the component of arthropathy in this entity. There was absence of diffuse inflammatory cell infiltrates differentiating this condition from rheumatoid arthritis and other inflammatory arthritides.

Treatment

Treatment is directed at getting the part to normal function as soon as possible after diagnosis. Since this syndrome can be so disabling and mentally taxing, the physiologic regimen of treatment should be support by physiological reassurance of the patient. Usual treatment regimens consist of antiinflammatory drugs, NSAIDS, anti-anxiety agents, muscle relaxors, vasodilators, and anti-depressant drugs. Cortico-steroids have been of particular value in the treatment of RSDS. Repeated peripheral sympathetic nerve blocks or regional intravenous nerve blocks in conjunction with the above have been useful. In severe intractable cases, surgical sympathectomy of the sensory nerves, dorsal roots, or spinothalamic tracts have been performed. Even amputation has been reported.

A typical podiatric patient seen in early stage of RSDS may be treated with NSAIDS, posterior tibial nerve blocks repeated at weekly intervals, followed by passive range of motion exercises and appropriate analgesics. Physical therapy is usually performed 2 to 3 times a week. Physical therapy aims at reducing muscle weakness, joint stiffness, edema, and indirectly diminishing osteopenia. Nerve blocks help break the pain cycle and allow for aggressive physical therapy while the extremity is somewhat numb. The patient is encouraged to keep the

limb warm and maintain weightbearing. In patients with vasomotor symptoms prednisone, 60-80 mg. every other day, has been used by Kozen and associates with good results (4). In addition vasodilators have been implemented. In patients with severe pain, posterior tibial or ankle blocks are supplemented with analgesics, sedatives, and muscle relaxors. With unremitting pain, narcotic analgesics in combination with antidepressants and psychiatric therapy are useful.

To date no studies have been published documenting clinically objective and symptomatic long term successful results post treatment. Generally in the early stages pain diminishes. Absence of pain allows joint mobility which reduces edema by reestablished lymphatic and venous drainage. As hyperemia subsides, osseous structural and architectural remodeling occurs. Bones become more capable of accepting and reacting to weight-bearing stresses. Gradually roentgenographic evaluation shows a homogenous osseous appearance with re-established thicker, smoother, cortices and absence of epiphyseal and metaphyseal radiolucencies. The above are dependent as to when the diagnosis of RSDS was made, treatment instituted, age and mental stability of the patient. Suffice it to say that the earlier the diagnosis is made, treatment instituted, and the younger the patient, the more rewarding the results.

Treatment in children has generally given excellent results since symptoms are not as severe as adults and since trophic changes and osteoporosis are usually absent.

Prognosis

Even though the above therapy has rendered many patients partially or completely asymptomatic, others show little or no response. Symptoms may persist for years and disability may become permanent. This syndrome has caused patients to have disastrous lives due to severe emotional and economic problems related to the disability. Some have been driven to suicide.

Summary

Reflex sympathetic dystrophy syndrome (RSDS) is seen in the podiatric medical practice. The symptom complex has a predilection for the lower extremity. Severe trophic and osseous changes are part of the syndrome. The diagnosis should be made as soon as possible for best long term results.

Migratory Osteolysis of the Lower Extremity

Migratory osteolysis of the lower extremity was

described by Duncan in 1967 (8). Other headings used to describe this entity have been: transient osteoporosis of hip, foot, and knee, transient osteoporosis of hip, peculiar artropatia rarefacente dell'anca, sympathetic dystrophy of foot, regional migratory osteoporosis, and transient osteoporosis of the lower extremities.

In his paper Duncan described three patients with a syndrome consisting of acute migratory and painful soft tissue swelling in the lower extremities associated with a rapid development of osteoporosis. Each regional episode of pain lasted less than nine months and resolved spontaneously without specific treatment. Remineralization of the affected sites occurred within 2 years. Non painful weightbearing and full activities were resumed much earlier than x-ray findings of bony remineralization. The essential features of the syndrome are:

1. Regional swelling and pain involving the lower limbs,
2. Rapid, severe osteoporosis localized to the same area,
3. Duration of symptoms 6-9 months,
4. Spontaneous involvement of other regions.

The typical reaction begins in an area of the limb with no history of trauma, surgery, or other precipitating factor. The areas become swollen, slightly to moderately tender and extremely painful on weightbearing. There is little pain non-weightbearing. In the first two to three weeks vasomotor changes may cause erythema, vasodilation, and moist skin. With time weightbearing becomes disabling and patients function with crutches. Months later signs and symptoms diminish. First to subside are color changes, then swelling, and last pain. The sequence of events is not the same in all patients. Months to years may pass before a different extremity is affected.

Radiology

The most remarkable x-ray finding is a rapidly developing osteoporosis during the first four to eight weeks. Intramedullary cystic changes, endosteal and periosteal erosions have been described. Serial x-rays reveal decreased cortical thickness and slight periosteal elevation. Trabeculae appear very thin and some disappear completely. Normal thickness of the cortices returns one to two years later. Residual trabeculae thicken, but the original number does not return. The most commonly affected parts of the extremity in Duncan's studies were the ankle and foot. Osteoporosis can extend into the knee, hip, and foot.

Laboratory Studies

Exhaustive studies for metabolic bone disease, infection, rheumatic, or collagen diseases are usually negative. Sedimentation rate is normal. Renal, neurologic, and all other systemic disorders are absent. Duncan did report three patients with two hour postprandial blood sugar levels ranging 140-160 mg/100 ml., and a slightly delayed return to normal of three hour glucose tolerance curves. Bone biopsies are sterile. Stains reveal marked tissue edema with increased osteoblasts and osteoclasts surrounding individual bone trabeculae.

Discussion and Differential Diagnosis

At the outset the clinician would think he might be faced with any of the acute inflammatory or rheumatic disorders, since patients usually present when symptoms are most acute. Other conditions in a differential diagnosis include gout or pseudogout, sarcoidosis and infection.

Gout can be ruled out on the basis that synovium in this case is not hypertrophied nor are there uric acid crystals present. Absence of crystals would also rule out pseudogout. Gout usually runs in an acute course of one to two weeks. This entity lasts three to six weeks. Rheumatoid arthritis (RA) and palindromic rheumatism can be ruled out on the basis of morning stiffness which is not present in this entity. Untreated palindromic rheumatism lasts one to two weeks.

Rheumatoid arthritis usually presents bilaterally symmetrical. Radiographically, RA presents with multiple joint changes, juxta-articular osteoporosis, narrowing and subsequently subchondral osteolysis.

Infection would reveal positive cultures and laboratory studies which generally reveal leucocytosis. In addition malaise and fever would be present.

Reflex sympathetic dystrophy syndrome (RSDS) is generally induced by trauma which results in signs and symptoms secondary to vaso motor reactions.

Metabolic disorders such as hyperparathyroidism, renal osteodystrophy, and Mediterranean fever can be ruled out on the basis of history, physical examination, and laboratory studies. Other conditions in a differential diagnosis would include tuberculosis, disuse atrophy, demineralizing synovial chondromatosis, and other neoplasms.

Treatment

Treatment during the initial stages with mild analgesics,

NSAIDS, muscle relaxants, and anti-anxiety agents are useful. Excellent results have been reported with corticosteroid therapy (15 mg prednisone daily for 4 weeks) followed by gradual tapering. Physical therapy, range of motion exercises, and forced gradual weightbearing have been shown to prevent associated localized muscle atrophy. Weightbearing helps promote cortical thickening giving greater architectural strength to the bones. Local sympathectomy using posterior tibial nerve blocks have been helpful in conjunction with the above.

Summary

The syndrome of migratory osteolysis of the lower extremity, including a differential diagnosis has been presented. It behooves the podiatric physician to be aware of the syndrome since it is most commonly reported in the lower extremity and resembles RSDS, but has definite and distinct features, the most important of which is spontaneous remission.

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