The typical setting for SRC is early dcSSc with a recent increase in skin thickening, palpable tendon friction rubs, and anti–RNA polymerase III antibody. During active, early dcSSc, patients should check their blood pressure once weekly and report a rise in systolic blood pressure of more than 20 mm Hg from baseline. Prednisone given at a dose of 15 mg daily or higher has been associated with the development of SRC and should be avoided in at-risk patients.

Cardiac Manifestations

Patients with SSc have three primary types of cardiac involvement: pericarditis, myocarditis, and myocardial fibrosis. The latter can lead to congestive heart failure and arrhythmias due to fibrosis of the conduction system. These complications can be asymptomatic and underrecognized in SSc patients, but pathologic changes have been found in most patients in older autopsy series. Later studies using cardiac magnetic resonance imaging (MRI) have confirmed the autopsy findings.

Diastolic dysfunction is becoming increasingly recognized as a complication of fibrosis and can be evaluated by echocardiogram during pulmonary hypertension screening. Many SSc deaths occur suddenly, possibly due to ventricular arrhythmias. It is prudent to obtain a resting electrocardiogram early in the disease course. Palpitations noticed by the patient should be addressed with a formal cardiac arrhythmia evaluation.

Gastrointestinal Tract Manifestations

At least one gastrointestinal manifestation will affect 80% or more of SSc patients, and all areas of the gastrointestinal tract may be affected. Gastrointestinal involvement is a significant cause of morbidity.

When the esophagus is affected, patients experience heartburn due to relaxation of the esophageal sphincter and distal dysphagia for solid foods due to esophageal dysmotility. Neuropathic changes and fibrosis of the muscularis of the small intestine can lead to motor dysfunction and symptoms of postprandial abdominal distention. Small intestinal hypomotility may lead to bacterial overgrowth, causing bloating and diarrhea.

When severe atony of the small intestine develops, patients occasionally develop a functional ileus or intestinal pseudo-obstruction. Parenteral nutrition may be necessary for severe malabsorption with accompanying weight loss and steatorrhea. Similar to the small bowel, the colon may develop impaired motor function leading to constipation and occasionally overflow diarrhea. Wide-mouthed diverticula on the antimesenteric border of the colon can be seen. The internal anal sphincter may become fibrotic, resulting in fecal incontinence.

Musculoskeletal Manifestations

Musculoskeletal manifestations are common. Tendons can become inflamed and fibrotic, particularly in early, diffuse disease. Palpable tendon or bursal friction rubs are virtually pathognomonic of SSc and often are a harbinger of progression to dcSSc before widespread skin thickening has occurred. Finger joint flexion contractures develop frequently within the first 2 years of diffuse SSc. True arthritis with palpable synovitis should raise the question of overlap with rheumatoid arthritis.

Some patients develop a bland myopathy with nonprogressive, mild proximal muscle weakness and wasting. A few, particularly with features that overlap with other connective tissue diseases, can develop true myositis, which can result in morbidity and disability.

DIAGNOSIS AND DIFFERENTIAL DIAGNOSIS

Raynaud's disease (i.e., primary Raynaud's phenomenon) is prominent in the differential diagnosis for SSc. Features that identify Raynaud's patients who have or may later develop SSc or another connective tissue disease are abnormal nail fold capillaries (i.e., capillary dilation, megacapillaries, and avascular areas), tissue loss at the tips of the fingers, and a positive antinuclear antibody (ANA) test result. None of these features is found in Raynaud's disease.

Mixed connective tissue disease (MCTD) is also on the differential for SSc. MCTD patients have features of two or more autoimmune diseases. This most frequently includes SSc, polymyositis, and SLE. Patients are positive for anti–U1-RNP antibody, a serologic marker for MCTD. Patients with MCTD can develop any or all of the following SSc manifestations: Raynaud's phenomenon, puffy fingers, limited or diffuse skin thickening, myositis, ILD, PAH, and esophageal dysmotility.

Scleroderma mimics are sometimes difficult to distinguish from SSc (Table 80-2). They include eosinophilic fasciitis, the localized forms of scleroderma such as linear scleroderma (more frequently seen in children), and plaque or generalized morphea.

Nephrogenic systemic fibrosis is a complication of gadolinium administration for radiographic studies that occurs in the setting of renal failure. Nephrogenic systemic fibrosis manifests as symmetrical, bilateral, fibrotic, indurated papules, plaques, or subcutaneous nodules, which can be erythematosus and occur on the lower legs or hands. The lesions are often preceded by edema and may initially be misdiagnosed as cellulitis. This diagnosis should be considered in patients being evaluated for a fibrotic disorder who have renal failure, regardless of the cause of renal disease.

Scleromyxedema and scleredema are cutaneous fibrotic disorders in which excessive mucin accumulation is found on skin biopsy. Scleromyxedema can mimic dcSSc on the physical examination or can manifest with multiple, firm, nodular skin lesions (i.e., papular mucinosis). A frequent association is a monoclonal gammopathy (i.e., immunoglobulin [IgG] paraprotein). Scleredema typically involves the nape of the neck and shoulders, sparing the distal extremities. All SSc mimics lack Raynaud's phenomenon, characteristic SSc internal organ involvement, and SSc-associated serum antibodies.

TREATMENT

Because no single therapy exists for SSc, patients must be appropriately monitored for visceral involvement to allow early identification and therapy targeted at specific organ complications. Consultation with a rheumatologist is helpful in this respect, and referral of severely affected patients to a dedicated scleroderma center should be the rule.

All patients should undergo screening evaluation for ILD and pulmonary hypertension throughout the course of their disease. Current expert recommendations suggest that patients with early, diffuse disease should be monitored at least yearly for these