A 52-year-old woman presented with a 1-year history of weakness, fatigue, and joint pain. During this time, the patient reported periods of weakness in her legs and arms that was not associated with exercise. She also complained of difficulty getting out of the bathtub and arising from a chair. The patient also experienced easy fatigability and joint pains in her wrists and hands. She denied redness or swelling in her joints; however, she had increasing episodes of stiffness that worsened as the day progressed. The review of systems was negative for skin rashes, alopecia, Raynaud’s phenomenon, dyspnea, cough, or wheezing. Her weight had been stable over the past year. The patient previously had been in good health and was currently employed full time as a hotel inspector. She had never smoked or consumed alcohol.

**Physical Examination**

Vital signs: temperature, 37°C; respiration, 20/min; pulse, 100/min and regular; BP, 156/84 mm Hg. General: no respiratory distress. Cardiac: normal heart tones without murmurs. Chest: bibasilar crackles. Extremities: mild tenderness of the joints of the wrists and hands without erythema or swelling. Results of neurologic examination were normal with exception of decreased strength (3/5) in the deltoid, triceps, and hip flexor muscles.

**Figure 1.** Bilateral interstitial infiltrates and hilar adenopathy.

**Figure 2.** Photomicrograph of muscle containing a nonnecrotic epithelioid granuloma with inflammation extending between adjacent muscle fibers. One adjacent degenerating fiber is basophilic and contains macrophages within the sarcoplasmic envelope (hematoxylin-eosin, original magnification ×250).
Laboratory Findings

Hematocrit, 34.8%; WBC, electrolytes, and renal indices: normal. Creatine phosphokinase (CPK), 1,561 U/L; aspartate aminotransferase, 68 U/L; erythrocyte sedimentation rate, 33 mm/h; angiotensin-converting enzyme, 60 U/L. Thyroid function tests: normal. Antinuclear antibody, negative. Chest radiograph: bilateral interstitial infiltrates and hilar adenopathy (Fig 1).

What diagnosis can explain the patient’s clinical presentation?

Diagnosis: Sarcoid Myopathy

Sarcoidosis is a systemic disease of unknown etiology. Characteristic noncaseating granulomas can be found in a variety of organ systems. The most common sites are the lungs, lymph nodes, liver, eyes, and the skin. Other tissues that may be infrequently involved include muscle, joints, spleen, bones, nervous system, blood vessels, and the endocrine glands. There is a higher incidence in the Afro-American female population. It typically occurs in the third or fourth decades; however, it can present at any age. Patients with sarcoidosis may be asymptomatic or they may present with fatigue, malaise, cough, dyspnea, and chest pain.

Muscle involvement is rarely the presenting manifestation of sarcoidosis. Asymptomatic myopathy can be in detected 25 to 50% of patients by muscle biopsy specimen. However, symptomatic muscle involvement is rare and found in less than 1% of patients with sarcoidosis. Sarcoid myopathy without other organ system involvement has never been documented; therefore, it is necessary to search for other organ involvement in these cases.

Three clinical types of sarcoid myopathy have been described—acute, chronic, and a palpable nodular myopathy. The chronic type is the most common with diffuse, symmetric, progressive muscle weakness of the extremities with a remitting and relapsing course. Frequently, muscle pain can precede the weakness and atrophy. The CPK levels are often normal. The acute form is infrequently observed with fewer than ten cases reported. Muscle weakness is the initial presentation and can be confused with polymyositis since the CPK levels are usually elevated. The nodular form of myopathy is also rare and presents with pain and tenderness of the skin.

Sequelaes from the myopathy is thought to result from pressure atrophy and muscle degeneration with fibrosis and not from granuloma invasion of the muscle fibers. A neuropathy can also occur and affect the course and response to therapy. Muscle biopsies are warranted for a definitive diagnosis in patients with symptomatic muscle involvement.

Corticosteroid therapy is the treatment most often used. Patients with the acute form of the disease appear to respond rapidly and with better long-term results; however, this impression is based on limited case studies. It appears prudent to begin patients on a regimen of high-dose oral corticosteroids and monitor serum CPK level as well as clinical response.

The present patient underwent a quadriiceps femoris muscle biopsy; the biopsy specimen revealed an inflammatory myopathy with noncaseating granulomatous material and giant cells (Fig 2). Results of pulmonary function tests were as follows: FVC, 1.70 L; FEV1, 1.30 L/s; and FEV1/FVC, 76% consistent with moderate restrictive ventilatory impairment. Transbronchial biopsy specimen obtained by fiberoptic bronchoscopy revealed scattered noncaseating epithelioid granulomas and multinucleated giant cells. Stains and cultures for fungi and mycobacteria were negative.

The patient received 40 mg/d of prednisone. Within the next 6 months, her serum CPK level decreased to 682 U/L, muscle weakness improved, and joint pains resolved. Corticosteroid doses were tapered slowly; however, dyspnea developed within a year of diagnosis. She is presently receiving 20 mg of prednisone for pulmonary dysfunction without muscle weakness.

Clinical Pearls

1. Sarcoid myopathy may be the initial manifestation of this multisystem disease. The normal clinical presentation is a diffuse, slowly progressive, symmetric proximal muscle weakness with muscle atrophy that resembles muscular dystrophy.

2. Although muscle weakness may be the primary symptom, other organ systems usually are involved at the time of diagnosis.

3. The CPK level may be elevated or normal. Muscle biopsy specimens are necessary to exclude a neuromuscular disorder that may occur concomitantly with sarcoidosis.

4. Corticosteroid therapy is recommended at high doses (40 to 60 mg/d). Tapering the dose based on the CPK level and the patient’s symptoms is suggested.

Suggested Readings


