Polymyositis is an uncommon rheumatological disease of unknown etiology, with an estimated prevalence of between 0.5 and 8 cases per million. Proximal muscle weakness is the most frequent clinical manifestation and the initial symptom in 80% of patients. The diagnostic criteria are symmetric proximal muscle weakness, increased muscle enzyme levels (creatine kinase and aldolase), characteristic alterations in the electromyogram, and the demonstrated presence of inflammatory cell infiltrates and necrosis in a sample of muscle tissue.

Pulmonary complications appear in more than 46% of patients with polymyositis and are associated with reduced survival. We present the case of a 60-year-old woman with dyspnea and muscle weakness who was diagnosed with polymyositis and interstitial lung disease (radiography indicated possible nonspecific interstitial pneumonia). The patient responded well to prednisone and methotrexate.

Key words: Polymyositis. Lung diseases, interstitial. Nonspecific interstitial pneumonia.

We present the case of a patient whose condition manifested with simultaneous muscular and pulmonary symptoms. She was diagnosed with polymyositis and interstitial lung disease that improved with corticosteroids and methotrexate.

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She presented arthralgia in the knees and hands and reddening of the fingers in response to cold temperatures, although Raynaud disease was not evident. The most noteworthy aspect of the physical examination was Velcro-type end-inspiratory crackles on pulmonary auscultation; cardiac auscultation was normal. Of particular interest was considerable weakness at the level of the scapula and pelvis, which made it difficult for the patient to raise her arms above her head and rise from a chair without using her arms.

Standard chest radiograph revealed a predominant bibasilar interstitial pattern. Baseline arterial gas values were as follows: \( \text{PaO}_2 \), 59 mm Hg; \( \text{PaCO}_2 \), 42.8 mm Hg; oxygen saturation, 90%; and alveolar-arterial gradient in partial pressure of oxygen, 31.9 mm Hg. Based on a suspicion of interstitial lung disease coupled with a possible connective tissue disorder, the decision was taken to admit the patient to hospital for further tests.

The most relevant details of the laboratory analysis were as follows: aspartate aminotransferase, 111 U/L; alanine aminotransferase, 81 U/L; lactate dehydrogenase, 806 U/L; creatine kinase, 3976 U/L; and creatine kinase MB, 142 U/L. The remaining biochemical parameters, complete blood count, and coagulation study were normal. Serology provided positive results for antinuclear antibodies at a low-titer (1/80), and for smooth muscle and anti-Jo-1 antibodies. Tests for the remaining antibodies—anti-Ro/SS-A, anti-La/SS-B, anti-Sm, antiribonucleoprotein, and circulating antineutrophil cytoplasmic antibodies (with a perinuclear and cytoplasmic pattern)—and rheumatoid factor were negative. The protein profile was normal.

High-resolution computed tomography of the chest revealed a bilateral interstitial pattern, with septal enlargement, areas of ground glass opacity, reticulation, and traction bronchiectasis predominantly in the lower lobes and no clear areas of honeycombing (Figure 1). The results of lung function testing were as follows: forced vital capacity, 2400 mL (84.2%); forced expiratory volume in the first second, 1970 mL (81.7%); total lung capacity, 4080 mL (80%); diffusing capacity of the lung for carbon monoxide (DLCO), 4.65 mL/min/mm Hg (59.5%); and the DLCO/alveolar volume ratio, 1.33 (87%). Fiberoptic bronchoscopy did not reveal endobronchial lesions. A culture and smear of bronchial aspirate and bronchoalveolar lavage fluid were negative. A cell count in bronchoalveolar lavage fluid based on 400 cells revealed 244 macrophages (61%), 52 lymphocytes (13%), 44 neutrophils (11%), and 60 eosinophils (15%). Transbronchial biopsy yielded a fragment of lung parenchyma with fibrosis and an interstitial inflammatory infiltrate, as well as destruction of the alveolar parenchyma.

The electromyogram of the quadriceps and deltoid muscles revealed a myopathic pattern (Figure 2). Finally, a biopsy was taken of the deltoid muscle, and the sample tissue presented a marked inflammatory and interstitial infiltrate with a predominance of lymphocytes and necrosis. These abnormalities are indicative of polymyositis (Figure 3).

The diagnosis was of diffuse interstitial lung disease (DILD) associated with polymyositis, with possible nonspecific interstitial pneumonia (NSIP) according to the radiograph. Therefore, treatment was begun with methotrexate (25 mg weekly) and prednisone (60 mg/d). The patient progressed favorably, with an improvement in clinical symptoms, laboratory parameters, and respiratory function. Two months after treatment started,
According to the study, the prevalence of polymyositis ranging from 5% to 30% has been clearly established since then with an estimated Arch Bronconeumol. 2007;43(11):636-9.

The combination of ground-glass pattern, reticular opacities, and traction fibroblastic foci. In the case of DILD associated with polymyositis, the findings include septal enlargement (90%), reticular micronodules (27% to 73%), patchy consolidation (22% to 100%), and honeycombing (0% to 19%). The combination of a ground-glass pattern, reticular opacities, and traction bronchiectasis, without honeycombing, is indicative of NSIP, as was the case with our patient.

There is no relationship between creatine kinase concentrations or the extension of the muscle disease and the development of DILD. Most of the antibodies detected in patients with polymyositis recognize enzymes that intervene in the synthesis of transfer RNA (tRNA). The most common antibodies are those that target the histidyl-tRNA synthetase enzyme or anti-Jo-1 antibodies, which are present in only 30% of patients with polymyositis/dermatomyositis. Nevertheless, 75% to 100% of patients with polymyositis/dermatomyositis and positive anti-Jo1 antibodies present associated interstitial lung disease; therefore, this antibody is considered a marker of DILD, although it does not seem to have an added prognostic value. The so-called syndrome includes other types of myositis that present other antisynthetase antibodies such as threonyl-tRNA synthetase (anti-PL7), alanyl-tRNA synthetase (anti-PL12), or isoleucyl-tRNA synthetase (anti-OJ). These are also associated with interstitial lung disease.

Although there have been no randomized placebo-controlled clinical trials in patients with DILD associated with polymyositis, the decision to treat or not depends on the severity of lung dysfunction, the type of DILD, and whether or not glucocorticoids or immunosuppressants are contraindicated. Most authors recommend glucocorticoids at a dose of 0.75 mg/d to 1.0 mg/d (prednisone or an equivalent) and tapering the dose over time. A lack of response to corticosteroids or the onset of adverse events makes it necessary to introduce a cytotoxic or immunosuppressive agent. Although data are limited to isolated cases or small case series, favorable responses have been reported with cyclophosphamide (both orally and intravenously), azathioprine, methotrexate and cyclosporine A, and tacrolimus. In the study by Douglas et al involving 70 patients with DILD associated with polymyositis/dermatomyositis, the most common immunosuppressants were azathioprine (25 cases) and methotrexate (14 cases). We used methotrexate because of previous favorable experience with this drug in a similar case. Slow-release commercial formulations for oral, muscular, or subcutaneous administration make methotrexate a good treatment option, since they guarantee adherence and avoid the consequences of potential overdose by the patient.

The prognosis of DILD associated with polymyositis/dermatomyositis is better than that of idiopathic pulmonary fibrosis, as is the case with other collagen diseases. In the study by Douglas et al, mean survival at 3 and 5 years was 74% and 60%, respectively. By contrast, in the study by Marie et al, mean survival was greater: 90% at 3 years and 86.5% at 5 years.

In summary, most patients with polymyositis/dermatomyositis associated with DILD present clinical, radiographic, and histological evidence indicative of NSIP, which has a better prognosis and response to corticosteroids. Although no controlled randomized clinical trials have been carried out, most patients improve with corticosteroids and immunosuppressants.
REFERENCES