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CASE REPORT / OLGU SUNUMU

Polymyositis Lung Involvement Mimicking COVID-19 Infection – A Case Report

COVID-19 Enfeksiyonunu Taklit Eden Polimiyozit Akciğer Tutulumu - Olgu Sunumu

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ABSTRACT

Polymyositis is an autoimmune connective tissue disease that can cause interstitial lung disease. The clinical diagnosis is confirmed through muscle biopsy, electromyography findings, and laboratory parameters. Pulmonary involvement due to polymyositis can be radiologically confusing, as many clinical conditions, including other interstitial lung diseases and viral infections, have similar radiological findings. This case, which is clinically and radiologically similar to COVID-19 infection, is presented to highlight the early clinical presentation of polymyositis-associated interstitial lung disease and to emphasize the importance of differential diagnosis in interstitial lung diseases.

Keywords: COVID-19, lung, polymyositis.

ÖZ

Polimiyozit, interstisyel akciğer hastalığına sebep olabilen otoimmün bağ dokusu hastalıklarından biridir. Klinik tanı, kas biyopsisi, elektromiyogram bulguları ve laboratuvar parametreleri ile doğrulanır. Polimiyozite bağlı akciğer tutulumu, radyolojik olarak, diğer interstisyel akciğer hastalıkları ve viral enfeksiyonlar gibi birçok klinik durum ile karışabilmektedir. Klinik ve radyolojik olarak COVID-19 enfeksiyonu ile benzerlik gösteren bu olgu, polimiyozit ilişkili interstisyel akciğer hastalığının erken dönem klinik tablosuna dikkat çekmek ve interstisyel akciğer hastalıklarında ayırıcı tanının önemine vurgu yapmak amacıyla sunulmuştur.

Anahtar kelimeler: COVID-19, akciğer, polimiyozit.

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INTRODUCTION

Idiopathic inflammatory myopathies, including polymyositis and dermatomyositis, are autoimmune connective tissue diseases with varying degrees of muscle and systemic involvement. Interstitial lung disease is a common complication of idiopathic inflammatory myopathies and is directly associated with increased mortality.[1] In interstitial lung disease due to polymyositis, reticular density and honeycomb appearance are observed radiologically in the lower lobes and peripheral parenchymal tissue. Differential diagnosis should be made for viral pneumonias, opportunistic infections, chronic hypersensitivity pneumonia, pneumoconiosis, sarcoidosis, chronic eosinophilic pneumonia, cryptogenic organizing pneumonia, and drug-associated interstitial lung diseases.[2]

This case was diagnosed with polymyositis-related lung involvement at the peak of the COVID-19 pandemic, so it is presented to draw attention to the importance of non-infectious differential diagnoses and the early clinical appearance of idiopathic inflammatory myopathy-associated interstitial lung disease since its clinical and radiological features are similar to COVID-19 infection.

CASE REPORT

A 50-year-old male patient was admitted to the outpatient clinic with symptoms similar to the COVID-19 infection, such as a nonproductive cough, dyspnea, and muscle pain that lasted about 2 months, and difficulty performing daily tasks during the pandemic period. The patient didn't have any known comorbidities. No additional features were found when his family history was examined. Crackles were heard in both lung bases on lung auscultation. The neurological system examination and other system examination findings were normal. Laboratory parameters revealed creatine kinase 2432 U/L, erythrocyte sedimentation rate 76 mm/h, C-reactive protein 11.2 mg/dL, and lactate dehydrogenase 528 U/L. Electrolyte values were found in the reference range. In the evaluation of lung parenchyma areas in thorax computed tomography (CT), nodular lesions with reverse halo characteristics were detected in the lateral segment of the right lung middle lobe and the anterior segment of the left lung upper lobe (Fig. 1). In addition, peripheral ground glass areas and consolidation were observed in the lower lobe posterior segment of both lungs (Fig. 2). The patient was hospitalized by the pandemic service with an initial diagnosis of COVID-19. Although the COVID-19 real-time polymerase chain reaction test (RT-PCR) of the patient was negative, intravenous methylprednisolone treatment at a dose of 1mg/kg was started in the patient whose clinical symptoms and radiological findings supported COVID-19 infection. In the follow-up, the patient was discharged with oral 48mg methylprednisolone treatment due to a regression in myalgia and respiratory symptoms.

During the follow-up, the patient's symptoms and radiological findings regressed, and the methylprednisolone treatment was discontinued by reducing the weekly dose. Two months after methylprednisolone treatment was discontinued, the patient was admitted to the hospital again with increased dyspnea and muscle weakness. In laboratory parameters, elevations of creatine kinase (2565 U/L) and lactate dehydrogenase (598 U/L) continued. TSH (4.2 Uu/Ml) and T4 (0.52 ng/dL) values of the patient with myopathy findings were found to be normal. On the chest X-ray, an increased density in the bilateral lower zones and a fibroatelectatic band in the right middle zone were observed (Fig. 3). Repeated COVID-19 RT-PCR samples also resulted in negative results. Thoracic CT was performed for the differential diagnosis of the patient. On thorax CT, consolidation areas with air bronchogram in the medial segment of the right lung middle lobe and in the lingula of the left lung; ground glass areas and fibrotic changes in the posterior segments of the lower lobes of both lungs were detected (Fig. 4). The progression can be seen more clearly when the current thorax tomography is compared with the sections of the thorax tomography 2 months ago (Fig. 5).

Fiberoptic bronchoscopy was performed with the initial diagnoses of organizing pneumonia, hypersensitivity pneumonitis, chronic eosinophilic pneumonia, and rheumatologic disease lung involvement. Normal endobronchial findings were observed during bronchoscopy. Bronchial lavage and bronchoalveolar lavage pathology were benign. Bronchoalveolar lavage cell count analysis revealed 52% histiocytes, 32% lymphocytes, and 16% neutrophils.

The patient was thought to have lung involvement from a systemic disease with newly developed findings, so an evaluation of neurological and rheumatological diseases has been made. Raynaud's phenomenon, malar rash, signs of arthritis, and significant skin lesions were not detected. When the rheumatological markers were examined, the anti-nuclear antibody level was positive at 1/100 titer and anti-Jo-1 was positive. Electromyography of the lower extremities revealed findings consistent with moderate myopathy with proximal involvement. Histopathological data could not be obtained because the patient did not accept the muscle biopsy procedure. Due to the risk of transmission of the SARS-CoV-2 virus, pulmonary function tests and diffusion capacity measurements could not be made on the patient at the time of diagnosis. After the results, the patient was diagnosed with polymyositis and azathioprine 100mg and methylprednisolone 40mg treatments were started. The patient was followed up monthly by the rheumatologist and the methylprednisolone treatment dose was adjusted according to the patient's symptoms. After a 1-year follow-up, the patient without respiratory symptoms was evaluated in terms of lung involvement. In the 6-minute walk test, the effort capacity was compatible with age, the pulmonary function test parameters (FVC 3.49 L, FEV_1 2.93 L, FEV_1/FVC 84%) were normal, and the chest X-ray findings showed (Fig. 6). The figure shows the chest X-ray side by side in chronological order (Fig. 7).

DISCUSSION

Although the predominant symptoms of COVID-19 infection are fever and cough, patients may also experience headaches, sore throats, weakness, dyspnea, myalgia, and taste and smell disorders. During COVID-19 infection, a decrease in the lymphocyte count and fibrinogen level can be observed in laboratory parameters, while an increase in d-dimer, lactate dehydrogenase, prothrombin time, creatine kinase, C-reactive protein, ferritin, and interleukin-6 levels can be observed.[3] The most common thorax CT finding of COVID-19 pneumonia is ground glass opacities, which typically appear bilaterally, peripherally, multilobarly, and localized posteriorly. Ground glass opacity can be seen alone or may be accompanied by different findings such as consolidation, interlobular

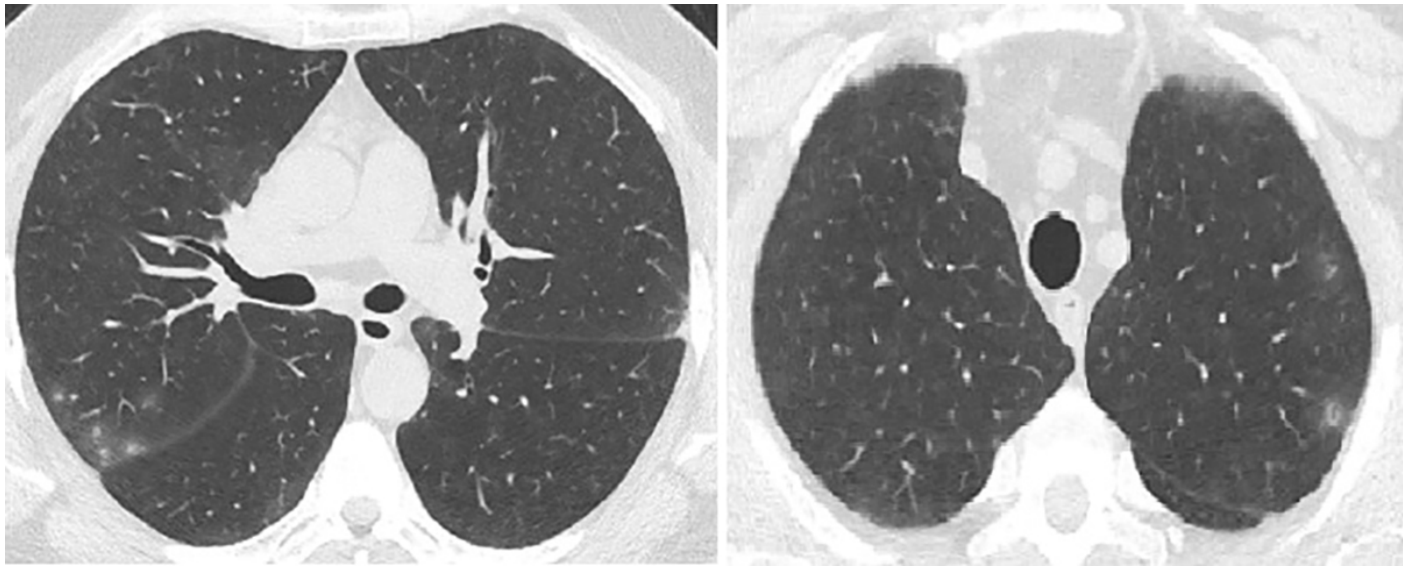

Figure 1: Nodular lesions with reverse halo characteristics were detected in the lateral segment of the right lung middle lobe and the anterior segment of the left lung upper lobe in the evaluation of lung parenchyma areas in thorax CT.

CT: Computed tomography.

septal thickening, and vascular dilatation. Consolidations are usually multifocal, segmental, patchy, mostly located in the lower lobe and peripheral, and may include air bronchograms.[4]

Our case presented symptoms similar to those of COVID-19 infection during the intense period of the pandemic. In the laboratory examination, elevated levels of C-reactive protein, lactate dehydrogenase, and creatine kinase were observed, and peripherally located ground glass areas were detected that were radiologically similar to COVID-19 lung involvement. Due to these features, the patient was followed up with an initial diagnosis of COVID-19. But after the COVID-19 RT-PCR tests were negative, examinations for a differential diagnosis were made and interstitial lung disease due to polymyositis was diagnosed. Although it is known that interstitial lung disease due to polymyositis can show itself as many other diseases such as viral pneumonia, drug-associated interstitial lung disease, sarcoidosis, and cryptogenic organizing pneumonia,[2] no cases appeared as COVID-19 pneumonia have been reported.

Idiopathic inflammatory myositis, first described by Bohan and Peter in 1975, is a rare, heterogeneous group of diseases that can involve multiple organs such as muscles, skin, joints, and lungs.[5,6] Inflammatory myositis can be divided into subgroups, including dermatomyositis, immune-mediated necrotizing myopathy, overlap myositis (containing antisynthetase syndrome), inclusion body myositis, amyopathic dermatomyositis, polymyositis, cancer-associated myositis through new myositis-specific autoantibodies, histopathological developments, and classification criteria. Different subgroups have different clinical symptoms, histopathological findings, autoantibody profiles, prognosis, and treatment responses.[5] Polymyositis and dermatomyositis are the major subtypes of idiopathic inflammatory myopathies.[7]

Diagnostic criteria for polymyositis include symmetric and progressive muscle weakness in the proximal extremity muscles, characteristic myopathic electromyography findings such as short-duration, low-amplitude complex repetitive discharges, fibrillations, and positive sharp waves; elevated creatine kinase, aspartate aminotransferase, alanine aminotransferase, lactate dehydrogenase, aldolase enzymes; and muscle biopsy findings showing CD8+ T-cell infiltration and the absence of characteristic findings of other myositis such as Heliotrope rash or Gottron's sign/papules.[5,8]

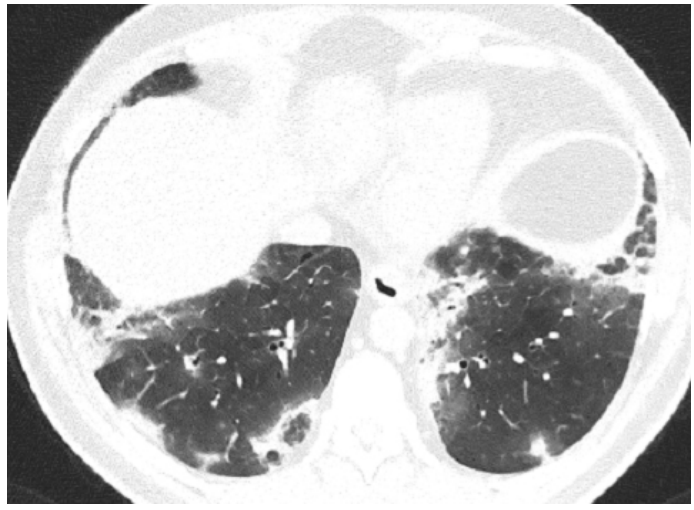

Figure 2: Peripheral ground glass areas and consolidation are observed in the lower lobe posterior segment of both lungs in the evaluation of lung parenchyma areas in thorax CT.

In our case, electromyography and laboratory findings were present, but the dermatological features were not observed. Idiopathic inflammatory myopathy is a rare disorder that occurs in approximately 14–17 per 100,000 people.[9] Pulmonary involvement occurs in 9–60% of idiopathic inflammatory myopathy; it causes about 40% of disease-related mortality.[6,9] Diagnosis may be delayed in patients whose initial sign is diffuse interstitial lung disease and who

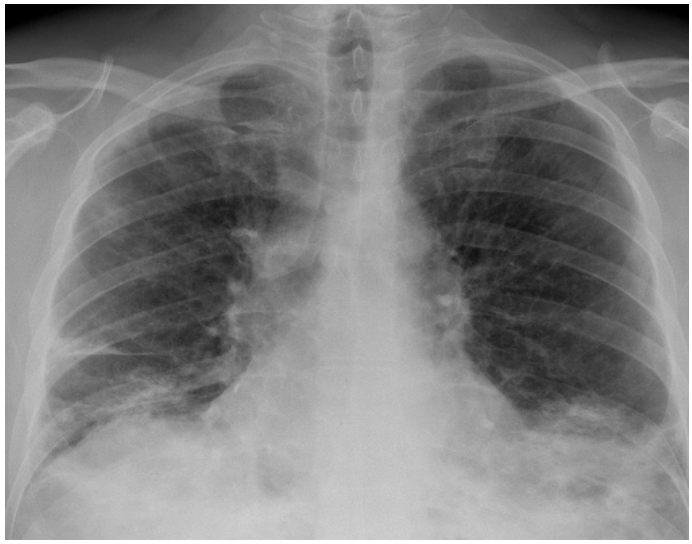

Figure 3: An increase in density in the bilateral lower zone and a fibroatelectatic band in the right middle zone are observed in the chest X-ray.

are receiving corticosteroid therapy due to the suppression of musculoskeletal findings.[10] In our case, the initiation of steroid therapy with the primary diagnosis of COVID-19 infection caused a temporary improvement in symptoms. For this reason, our case received the diagnosis of polymyositis in the follow-up process.

Autoantibodies are thought to play an important role in the pathogenesis of myositis in patients with idiopathic inflammatory myopathy and have been identified in more than 50% of patients. Among the autoantibodies; anti-Jo-1 and anti-melanoma differentiation factor-5 antibodies have high rates of interstitial lung involvement. Anti-Jo-1 positivity usually results in a more severe clinical course of myositis compared to other autoantibodies.[1] Overlapping myositis is defined as the presence of a connective tissue disease such as scleroderma, systemic lupus erythematosus with inflammatory myopathy, and the most frequently represented form of overlapping myositis is antisynthetase syndrome. Anti-synthetase syndrome is characterized by the presence of one or more of the clinical features and the positivity of one of the autoantibodies targeting amino acid tRNA synthetases. Anti-Jo1, anti-PL7, and anti-PL12 antibody positivity can be seen in this

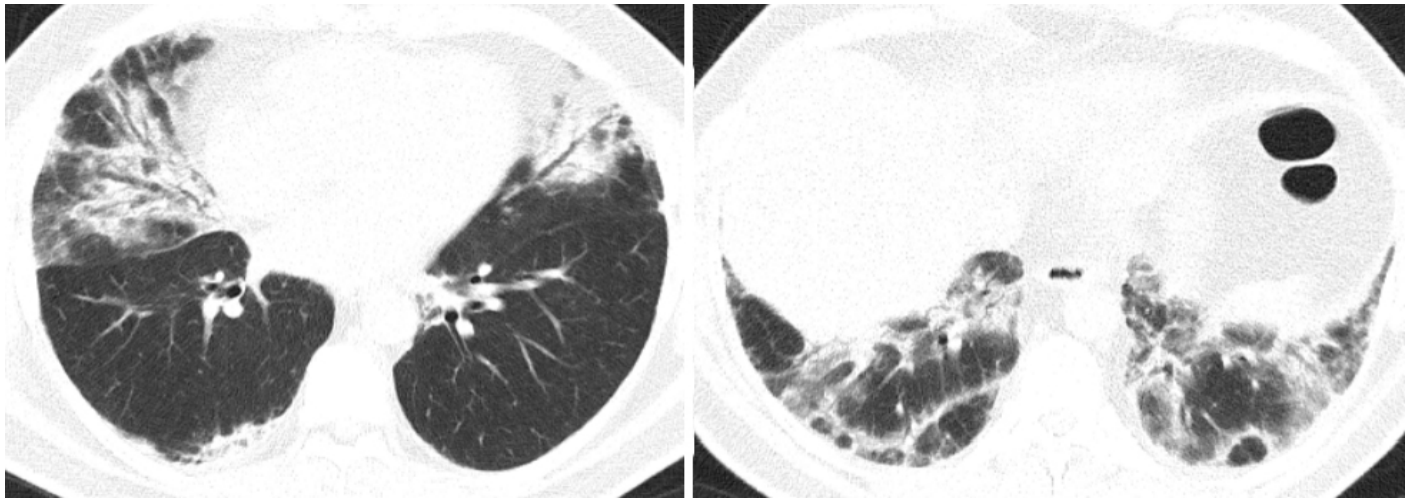

Figure 4: There are consolidation areas in the medial segment of the middle lobe of the right lung and in the lingula of the left lung with an air bronchogram. Ground glass areas are observed in the posterior segments of both lung lower lobes.

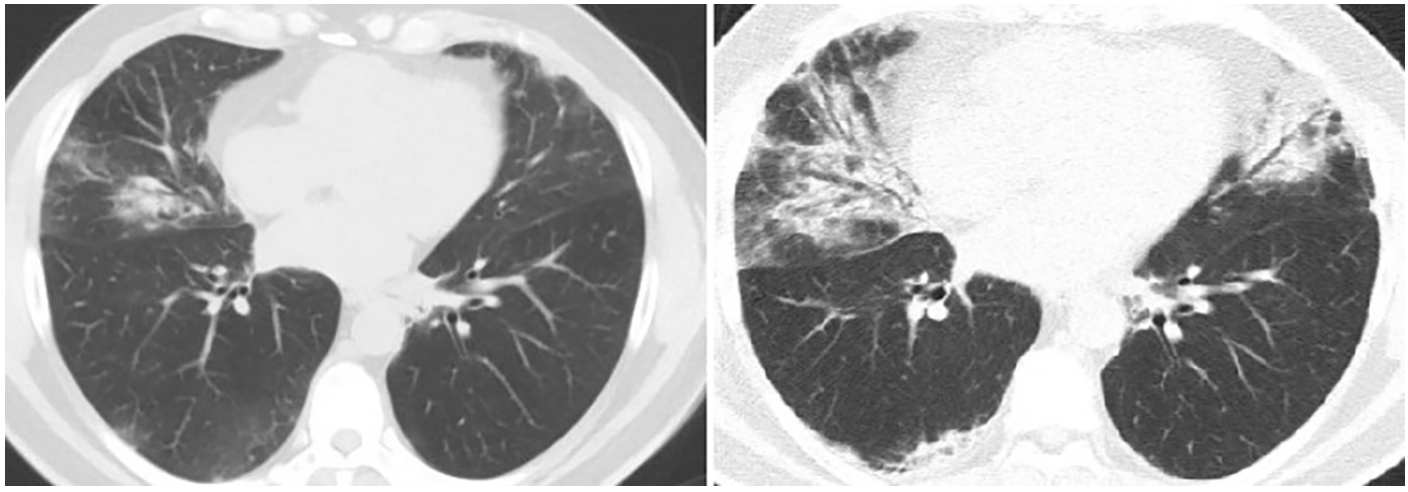

Figure 5: Comparison of two months ago and current thoracic CT; thorax CT sections show progression.

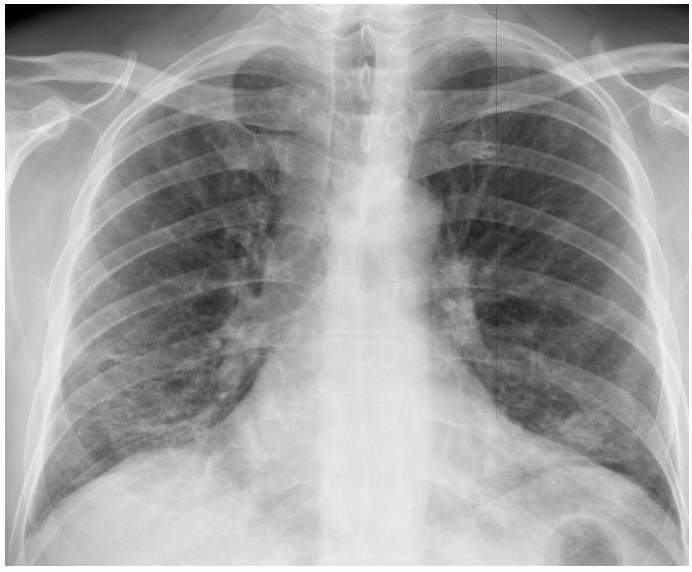

Figure 6: There is an increase in density in the lower zone of both lungs showing regression compared to the previous chest radiograph.

syndrome. Clinical features can be summarized as a mechanic's hand characterized by inflammatory myopathy, interstitial lung disease, arthritis, Raynaud's phenomenon, fever, and hyperkeratotic radial finger lesions.[5] The clinical features of interstitial lung disease due to polymyositis are variable. Patients may be asymptomatic or may describe symptoms such as exertional dyspnea and a non-productive cough.[9] Symptoms may resemble those of COVID-19. Our case had muscle weakness, dyspnea, and a non-productive cough, similar to the COVID-19 infection. In idiopathic inflammatory myopathy, lymphocyte or neutrophil dominance can be detected in the bronchoalveolar lavage, and interstitial pneumonia findings can be found in lung biopsy samples.[8] In the bronchoalveolar lavage examination of our case, lymphocytic alveolitis findings were similar to the literature data.

Surgical lung biopsy in MA-ILD (myositis-associated interstitial lung disease) patients is not required to confirm the diagnosis and is not routinely performed because of its limited usefulness in treatment decisions.[11] Our case was diagnosed with polymyositis based on clinical and radiological features and laboratory parameters; a surgical biopsy was not considered necessary.

A pulmonary function test (PFT) should be performed both for the diagnosis of ILD and for disease follow-up in patients with polymyositis. PFT findings typically include decreased FVC, decreased total lung capacity with moderate restriction, and a moderate decrease in diffusion capacity.[1] Although polymyositis-related lung involvement was symptomatic in our case, no loss in lung capacity was observed in PFT.

High-resolution thoracic CT is the gold standard for early diagnosis and identification of idiopathic inflammatory myopathy patterns. It plays an important role in the management of these diseases to initiate medical treatment and monitor disease progression and complications. Although consolidation is more common in patients with anti-Jo-1 antibody-positive idiopathic inflammatory myopathy than lung involvement in other connective tissue diseases, the most common radiological findings are still ground glass opacities and reticulations. As the disease progresses, consolidations typically regress, and honeycombing and fibrosis become more common.[9] Ground-glass areas observed in thorax CT scans in the first presentation of our case were similar to viral pneumonia findings. However, on the thorax CT at the next admission of the patient, there was predominantly lower lobe fibrosis, areas of consolidation, and a reticular pattern, which may be compatible with connective tissue disease-related interstitial lung disease.

All diseases causing muscle weakness should be included in the differential diagnosis of polymyositis; in cases where there is a family history, hereditary muscle diseases such as myotonic dystrophy, mitochondrial myopathies, glycogen storage diseases, and myofibrillary myopathy should be considered.[11,12] A differential di-

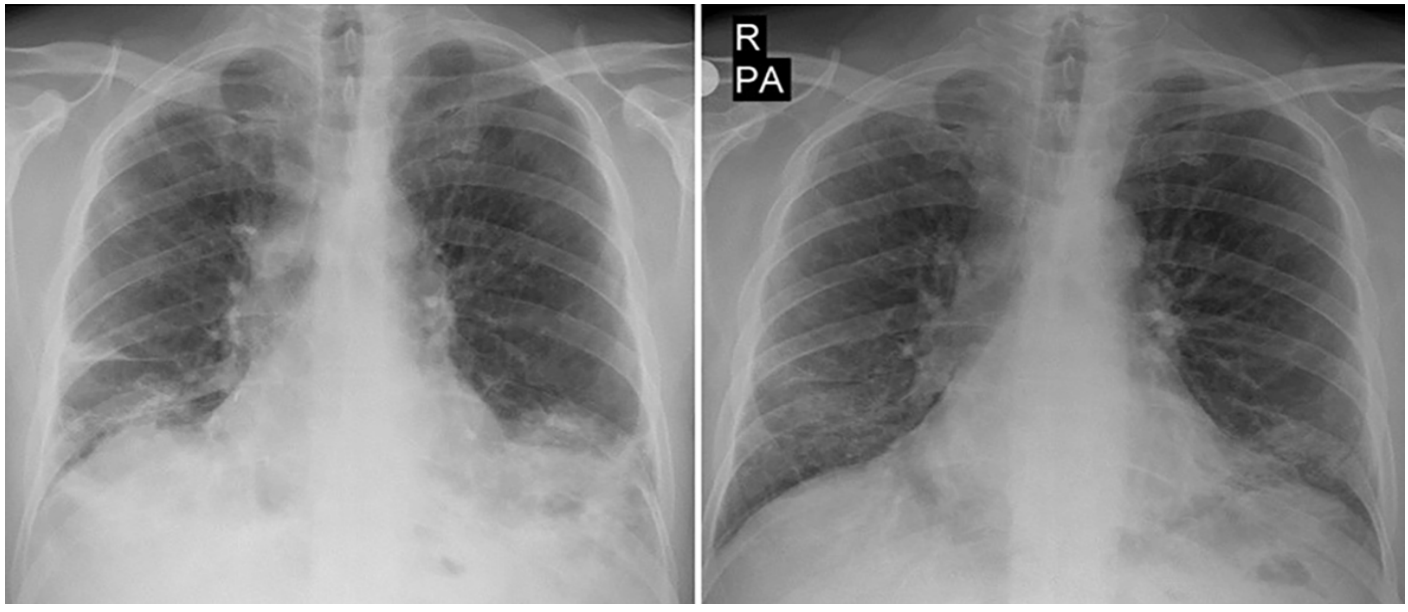

Figure 7: The figure shows the chest X-ray side by side in chronological order.

agnosis of inflammatory myopathies from other acquired muscle diseases and muscle-nerve junction diseases should be made.[11] History, physical examination, serological tests, neurological tests, and muscle biopsy are found helpful in the differential diagnosis of polymyositis.[5] Our case was diagnosed with polymyositis clinically, radiologically, and serologically.

There is no standard treatment regimen for the management of myositis-associated interstitial lung disease.[1] The goal of treatment is to stop inflammation as well as preserve muscle strength and function. In this group, high-dose glucocorticoid therapy should be initiated as early as possible.[12] Unfortunately, many patients do not respond to initial corticosteroid therapy or develop progressive disease when they are only treated with corticosteroids.[1] There are studies showing that the early addition of a second immunosuppressive agent to the treatment has a benefit for survival. Azathioprine, mycophenolate mofetil, cyclophosphamide, cyclosporine, tacrolimus, and rituximab are immunosuppressive agents that can be preferred.[12] Patients with anti-Jo-1 positivity respond better to glucocorticoid therapy, but with frequent relapses; therefore, immunosuppressive therapy in addition to glucocorticoids is necessary to achieve disease control.[7] Since our case with anti-Jo-1 positivity is symptomatic and has a progressive disease, dual immunosuppressive therapy in the form of glucocorticoids and azathioprine was started.

Polymyositis has a high mortality rate despite the widespread use of glucocorticoids with immunosuppressives; the 5-year survival rate is approximately 70%.[10] The main causes of death were cardiac (22%), pulmonary complications (22%), infections (15%), and cancer (11%).[11] ILD is one of the most important prognostic factors, resulting in poor survival. These days, early examination and effective management in patients with ILD are vital in clinical decision-making to complement the prognosis of polymyositis.[10]

CONCLUSION

In the period when the COVID-19 pandemic was followed intensively, our case was followed up with the diagnosis of COVID-19 infection, as it resembled COVID-19 in clinical, radiological, and laboratory findings. The patient was diagnosed with polymyositis when the findings of interstitial lung disease on Thorax CT, laboratory findings associated with muscle disease, and electromyelography were evaluated.

Lung infections, especially viral infections, can be confused with lung involvement in systemic diseases. Systemic symptoms, laboratory parameters, and radiological imaging of patients should be evaluated for a differential diagnosis.

Disclosures

Informed Consent: Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

Author Contributions: Concept – I.S., D.K., Ö.S.G.; Design – I.S., Ö.G.; Supervision – I.S., D.K.; Fundings – I.S., D.K., Ö.S.G.; Materials – I.S., D.K.; Data Collection and/or Processing – D.K., Ö.S.G.; Analysis and/or Interpretation – I.S., D.K., Ö.S.G.; Literature Search – I.S., Ö.G.; Writing – D.K., I.S.; Critical Reviews – D.K., I.S., Ö.S.G.

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