Cryptogenic Organizing Pneumonia as the Initial Presentation of Polymyositis

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Introduction

Interstitial lung disease (ILD) is a diverse group of lung diseases that can be associated with various connective tissue diseases including polymyositis and dermatomyositis (PM-DM). The occurrence of ILD in patients with PM-DM varies widely among case studies, ranging from 20-80%.[1] It has been previously reported that interstitial lung disease (ILD) may be a presenting feature of polymyositis (PM),[2-3] however the diagnosis of PM may be missed if patients do not present with other symptoms typically associated with PM.

Case Report

A 53 year old hispanic female presented to the hospital with a two week history of dry cough and dyspnea on exertion. A CT scan of the chest excluded pulmonary emboli but showed bilateral lower lobe consolidations and she was started on treatment for community acquired pneumonia. She did not clinically improve with antibiotics and deteriorated to the point of developing ventilator-dependent respiratory failure. Microbial workup for both fungus and bacteria, including bronchial washings obtained via bronchoscopy were negative. Hepatitis, HIV, comprehensive ANA panel (ANA, SS-A, SS-B, SCL-70, dS-DNA, SM/RNP, RF, CCP), c-ANCA and p-ANCA were also negative. A lung biopsy was performed which revealed findings consistent with cryptogenic organizing pneumonia (COP). She was empirically started on prednisone with gradual clinical improvement and was eventually discharged home with a prolonged prednisone taper and on two liters of oxygen via nasal cannula. Four months after her initial presentation, she presented with new symptoms of arthralgias, lower extremity weakness, and joint swelling of bilateral wrists, hands and knees. Increased CPK, aldolase, and positive anti-Jo-1 antibodies were identified. A muscle biopsy showed findings consistent with polymyositis. She was diagnosed with PM and started on high dose corticosteroids. Her symptoms of lower extremity weakness significantly improved, however she still remained oxygen dependent on two liters nasal cannula secondary to unresolved ILD. She was discharged home on high dose prednisone with close out-patient follow up with rheumatology.

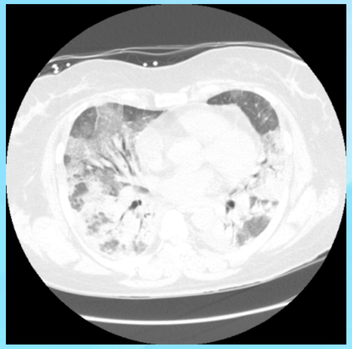

Figure 1: Patient's CT Scan showing bilateral lower lobe consolidation and diffuse groundglass opacities.

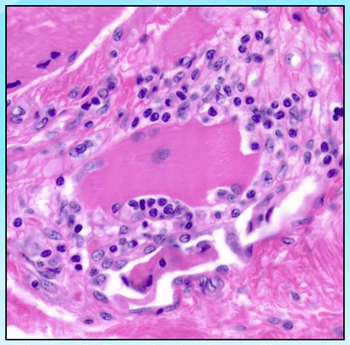

Figure 2: Example of polymyositis muscle biopsy. Cellular infiltrate is predominantly within the fascicle, with inflammatory cells invading individual muscle fibers.[1]

Discussion

Although proximal muscle weakness is the most frequent clinical manifestation and the presenting symptom in 80% of patients with PM, ILD maybe an early manifestation in patients with anti-Jo-1-positive PM-DM and may precede the onset of myositis.[2-4]

The presence of antisynthetase antibodies in PM-DM appears to be associated with a greater probability of developing ILD; one retrospective case series of anti-Jo-1 antibody-positive individuals reported the incidence of ILD as high as 86%.[5]

ILD has been reported to precede the onset of muscle involvement by months or even years, appear at the same time as symptoms of myositis, or months after the development of muscle involvement.[1, 4]

The patterns of lung pathology that have been found to be associated with DM and PM include: idiopathic interstitial pneumonias, including nonspecific interstitial pneumonia, usual interstitial pneumonia, organizing pneumonia, and acute interstitial pneumonitis.[4]

The initial treatment for ILD in patients with PM-DM include systemic glucocorticoids at a dose of 1mg/kg per day. ILD in PM is associated with a high rate of morbidity and mortality[6] and often require a second immunosuppressive agent to slow the progression of ILD and to enable tapering of glucocorticoids.

Conclusion

This case demonstrates that ILD maybe the initial presentation of PM-DM, thereby preceding the symptoms of myositis. Therefore, PM-DM must remain within the differential of ILD in order to prevent a delayed diagnosis which can be associated with significantly increased morbidity and mortality secondary to progression of the disease. This case also highlights the utility of anti-Jo-1 as both a marker for polymyositis and its use in the evaluation of patients with ILD of unclear etiology.

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