e- ISSN 0976-0342
Print ISSN 2229-7456

International Journal of Pharmacy & Therapeutics

Journal homepage: www.ijptjournal.com

Research article

DERMATOMYOSITIS ASSOCIATED WITH INTERSTITIAL LUNG DISEASE: A CASE REPORT

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ABSTRACT

Dermatomyositis is an inflammatory myopathic disease which is characterized by chronic muscle inflammation accompanied by muscular weakness. The prevalence rate of dermatomyositis is 1 in 10,00,000 population with high risk in females than males. Interstitial lung disease is the most common and severe complication of inflammatory muscle diseases like dermatomyositis that affect the tissue and spaces around alveoli. In these patients, muscle weakness is usually accompanied by shortness of breath and cough and can lead to severe respiratory distress. We report a case of a male patient, 45 years old whose clinical history was onset in March 2019 with classical muscular weakness, heliotrope rash, gottron's papule and respiratory symptoms like cough and shortness of breath. Patients was on steroids and cyclophosphamide therapy. In this brief case report, we want to emphasize on the pulmonary complication of dermatomyositis.

Key Words:- Dermatomyositis, Interstital Lung Disease, Cyclophosphamide.

Access this article online

Home page: http://ijptjournal.com/

DOI: http://dx.doi.org/10.21276/ijpt.2020.10.1.1

Quick Response code

Received:12.09.19 **Revised**:22.09.19 **Accepted**:16.10.19

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INTRODUCTION

The inflammatory myopathies are a group of diseases that involve chronic muscle inflammation and weakness. They are autoimmune disorders in which our body natural defense mechanisms which are meant to act on foreign particles, in contrast, attacks healthy cells which lead to inflammation and damage to cells.

Dermatomyositis is one type of inflammatory myopathy characterized by inflammatory and degenerative changes of the muscles and skin. The average age at diagnosis is 40 years, and almost twice as many women are affected as men, with a prevalence rate of 1 per 100,000 in the general population (Kalyan M *et al*., 2016). Muscle symptoms initially presented as pains and weakness of the muscles of the trunk, upper arms, hips, and thighs (proximal muscles). In dermatomyositis, muscles tend to be stiff, sore, tender and, eventually, show signs of degeneration (atrophy). Dermatomyositis patients may experience difficulty in performing some functions, like raising their arms and/or climbing stairs or develop speech and swallowing difficulties. Dermatomyositis is usually the easiest type of myositis to diagnose because of the skin rash, which often appears before any muscle weakness is felt. The rash looks patchy, dark, and reddish or purple. It is most commonly found on the eyelids, cheeks, nose, back, upper chest, elbows, knees, and knuckles. While the rash of dermatomyositis may be the first sign of the disease, those with darker skin may not notice the rash as readily. Other characteristic DM symptoms include scaling and redness of the knuckles, elbows, knees, and/or other

extensor regions (Gottron papules and sign); an abnormal accumulation of fluid (edema) in body tissues surrounding the eyes; and/or other features (Anonymous 1).

The association of Interstitial lung disease and dermatomyositis had been well established. In rheumatic diseases (RDs), Interstitial lung disease (ILD) is a major complication. Approximately 25% of all ILD deaths and 2% of deaths due to respiratory failure are estimated to be due to underlined autoimmune disorders (Salimbene I *et al*., 2013).

Interstitial lung disease is a group of diseases that affect the tissue and spaces (interstitial) around the alveoli in the lung. When these spaces were obstructed by inflammation, the exchange of gases is impaired. If it is untreated for too long then it can lead to fibrosis of lung tissue in which lungs are scarred causing severe breathing problems. Interstitial lung disease is the most often and serious complication of the inflammatory myopathies. Researchers estimate that 30-40% of myositis patients have some form of lung disease due to autoimmunity directed on alveolar tissue (Anonymous 2).

Symptoms vary among affected individuals, which include shortness of breath, cough (usually a dry cough with no sputum), or no symptoms at all. Generally Symptoms do not progress rapidly, but in contrast, respiratory distress occurs quickly. Interstitial lung disease can even present before muscle symptoms become apparent. In addition, the severity of muscle or skin disease is not necessarily an indication of severe lung disease. Mild muscle and skin symptoms may not clearly indicate the severity of underlined interstitial lung disease (Anonymous 2).

Radiologic abnormalities in DM are characterized by a high incidence of airspace consolidation and a low incidence of honeycombing. Chest computerized tomography (CT) scan, especially a high-resolution CT, is preferred as a way to detect interstitial lung disease, identify the severity of the disease, and distinguish between fibrotic disease and active inflammation in the lungs (Cottin V *et al*., 2003). Classic signs of ILD include ground glass opacities (a characteristic appearance that indicates inflammation in the air sacks), reduced lung volume, and bronchiectasis (enlargement of the airways) (Douglas W *et al*., 2001).

Recent research has revealed that myositis-specific autoantibodies (MSAs) are closely linked to clinical phenotypes in PM/DM. The measurement of MSAs is useful for predicting clinical course, clinical characteristics, response to treatment, and prognosis in PM/DM patients. The MSAs that are strongly associated with ILD include the anti-melanoma differentiation-associated gene 5 (MDA5) and anti–aminoacyl-tRNA synthetase (ARS) antibodies ILD activity and severity are dependent on the subtype of PM/DM. (2) Anti-ARS includes anti–Jo-1, anti-EJ, anti-OJ, anti–PL-7, anti–PL-12, anti-KS, anti-Zo, and anti-Ha. DM-specific rashes, such as heliotrope and Gottron's sign, have been frequently observed in patients with anti–Jo-1, anti-EJ, anti–PL-7, and anti–PL−12.9 (Hozumi H *et al*., 2105). Corticosteroids and immunosuppressive agents should be co-administered as soon as possible in ILD with PM/DM patients. However, other causes of pneumonia or pneumonitis, such as infections and drug use, should be excluded before treatment initiation. combination therapy of corticosteroids, IVCY, and CNIs, such as CSA and TAC, should be immediately administered in ILD patients with hyperferritinemia and/or anti-MDA5. CNIs or IVCY should be added to corticosteroids in patients with chronic progressive ILD if pulmonary function is gradually deteriorating and/or the lesions of ILD are spreading on CT findings. CNIs should be administered with PSL in ILD with anti-ARS patients to prevent the progression and recurrence of ILD (Kawasumi *et al*., 2015).

CASE PRESENTATION

We report a case of 45 years old male patient whose clinical symptoms were started in march 2019 with recurrent high-grade fever associated with chills and rigor. The patient had complaints of pain at large and small joints accompanied by generalized myalgias and muscular weakness. Four months after the presentation of these symptoms patient paid a visit to a tertiary care teaching hospital with some pulmonary symptoms like shortness of breath and severe cough. Upon inquiry about the history of present illness, patient disclosed all the above symptoms along with loss of appetite and weight. On examination patient neck we found sign of bilateral lymphadenopathy and periorbital puffiness. The patient was admitted to hospital for assessment of chronic febrile illness with polyarthralgia. After initial assessment patient was sent to rheumatology for assessment of patients symptoms. The patient was suffering from generalized muscle weakness from 4 months with gradual onset of proximal weakness leading to difficulty in performing daily activities involving proximal muscles. Upon careful examination patients body, we found the heliotropic sign (bluish rashes) on eyelids and face. Patients joints especially small joints like metacarpophalangeal joints were identified with erythematous papules.

The blood count was normal except for an increase in white blood cells (20,000 cells/cumm). The erythrocyte sedimentation rate was slightly increased (37 mm/1st hr), and the creatine kinase (CK) was found to be normal. Patients chest radiography revealed abnormalities which were confirmed by high-resolution chest computerized tomography as Interstitial lung thickening with honeycomb-like consolidation in the lower lobe of lungs. Patient blood was investigated for

autoimmune antibodies like Anti nuclear antibodies and Anti-Jo-1 antibodies found to be positive.

Based on clinical findings like heliotrope rashes, gottrons papules and positive for Anti-Jo-1 antibodies patient were diagnosed as Dermatomyositis. Proximal muscular weakness which is a classical sign of dermatomyositis rules out Amyopatic dermatomyositis from the scenario. Lung consolidation confirms the presence of interstitial lung disease which is a common complication of dermatomyositis.

The patient was started on corticosteroid therapy for suppression of autoimmunity and progression of the disease. Oral corticosteroid prednisolone was given at a dose of 1mg/kg/day which translates to a dose of 70 mg /day. For the treatment of interstitial lung disease patients was on Intravenous cyclophosphamide therapy monthly which was found to be beneficial for the treatment of ILD with dermatomyositis. Patient came to the hospital for administration of the second cycle of intravenous cyclophosphamide (IVCY) and his case was reviewed.

Table 1. Clinical parameters of the patient

LAB PARAMETERS	OBSRERVED VALUES	NORMAL VALUES
Pulse Rate	98 bpm	60-100 bpm
Blood Pressure	110/70 mm of hg	130/80 mm of hg
Spo 2	98 %	95-100 %
ESR	25 mm/1st hr	0-22 mm/1st hr
WBC	39,000 IU/L	3400-9600 cells/mcl
Hemoglobin	13.7 g%	12-16 g%
SGOT	82 IU/L	5-40 IU/L
SGPT	32 IU/L	7-40 IU/L

DISCUSSION

In this brief case report, we want to highlight the pulmonary complications and their treatment in dermatomyositis. ILD in dermatomyositis is fatal if left untreated. Pulmonary complications in patients with dermatomyositis may be aspiration pneumonia secondary to dysphagia, ventilatory failure secondary to muscular weakness, primary or metastatic malignancy, pleural effusion, spontaneous pneumothorax.

The onset of pulmonary symptoms in these diseases is highly variable. In some cases, pulmonary symptoms precede dermatomyositis symptoms and vice versa. In this case, the patient experienced myalgias and muscular weakness before the onset of pulmonary symptoms.

The five rheumatic diseases most frequently associated with the pleuropulmonary disease are rheumatoid arthritis, systemic lupus erythematosus, progressive systemic sclerosis, polymyositis/dermatomyositis, and Sjögren syndrome (Kawasumi *et al*., 2015). The overall 5-year survival rate is 77%–95% in PM/DM, and cardiac dysfunction, malignancy, respiratory failure, and infection are the major causes of fatal outcome. Previous studies reported that cardiac involvement and respiratory muscle involvement are significant prognostic factors for death in patients with PM/DM, except in patients with cancer. Yamasaki *et al* retrospectively investigated 197 patients with Polymyositis, Dermatomyositis, and Amyopathic dermatomyositis. Survival in the entire group at 1, 5, and 10 years was 85%, 75%, and 67%, respectively, and the mortality in Amyopathic dermatomyositis and Dermatomyositis was 61% and 77% at 5 years, respectively. Dermatomyositis patients exhibited significantly lower survival compared to PM (91% at 5 years), and ILD was the major cause of death in Amyopathic dermatomyositis (71%) and Dermatomyositis (60%) patients. Most of these patients died within the first few months (Kawasumi *et al*., 2015).

The patient was diagnosed as Interstitial lung disease with dermatomyositis is based upon CT report which showed interstitial thickening in both lungs with honeycomb-like structures in the lower lobe of lungs and some classical dermatomyositis like Heliotropic rashes, gottrons papules, proximal muscle weakness and presence of myositis specific antibodies.

According to myositis association to classify as dermatomyositis, the patient must have at least 1 skin symptoms like heliotropic rashes, gottrons papule, Periungual telangiectasias, and shawl sign. Along with skin symptoms, a patient must have proximal muscle weakness, elevated muscle enzymes, myalgias, positive report for myositis related antibodies and with signs of inflammation. (Anonymous 3)

Diagnosis of idiopathic inflammatory diseases is a key part of prognosis. based on the type of autoimmune antibodies idiopathic myopathies are classified as Polymyositis, Dermatomyositis, and Amyopathic dermatomyositis. The MSAs that are strongly associated with ILD include the anti-melanoma differentiation-associated gene 5 (MDA5) and anti–aminoacyl-tRNA synthetase (ARS) antibodies The presence of anti-MDA5 is associated with clinically amyopathic DM (CADM), especially cutaneous ulcers. CADM involves the typical skin lesions that are revealed in DM with amyopathy or hypomyopathy. Therefore, patients with CADM do not typically present with muscle symptoms, such as myalgia and muscle weakness. (Kawasumi *et al*., 2015)

Anti-ARS is associated with clinical manifestations, including arthritis, mechanic's hand, Raynaud's phenomenon, myositis, and ILD. Anti-ARS includes anti–Jo-1, anti-EJ, anti-OJ, anti–PL-7, anti–PL-12, anti-KS, anti-Zo, and anti-Ha. DM-specific rashes, such as heliotrope and Gottron's sign, have been

frequently observed in patients with anti–Jo-1, anti-EJ, anti–PL-7, and anti–PL−12. ILD alone has been frequently found in patients with anti-OJ and anti-KS, and myositis is not complicated with ILD in some patients with anti-ARS. (Kawasumi *et al*., 2015).

Corticosteroids and immunosuppressive agents should be co-administered as soon as possible in ILD with PM/DM patients. However, other causes of pneumonia or pneumonitis, such as infections and drug use, should be excluded before treatment initiation. Therefore, combination therapy of corticosteroids, IVCY, and CNIs, such as CSA and TAC, should be immediately administered in ILD patients with hyperferritinemia and/or anti-MDA5. CNIs or IVCY should be added to corticosteroids in patients with chronic progressive ILD if pulmonary function is gradually deteriorating and/or the lesions of ILD are spreading on CT findings. CNIs should be administered with PSL in ILD with anti-ARS patients to prevent the progression and recurrence of ILD. The administration of steroid-sparing agents (eg, MTX, AZA, CNIs, and MMF) should be also considered in patients with chronic mild ILD. RTX or MMF administration may be considered when Rapid progressing ILD or chronic ILD (Kawasumi *et al*., 2015).

CONCLUSION

This case report emphasizes the pulmonary complication of dermatomyositis. Interstitial lung disease is main reason for mortality in dermatomyositis cases. Dermatomyositis should be considered in the differential diagnosis of patients presenting with pulmonary symptoms, rashes, papules, and facial edema. This diagnostic approach may enable early diagnosis and treatment of this potentially fatal disease and prevent progression to malignancies. Detection of myositis specific antibodies confirm the diagnosis in the absence of skin biopsy. The best approach for treating ILD with dermatomyositis is the use of corticosteroids along with monthly intravenous cyclophospamide is proven to be beneficial if the disease is detected in early stage.

ACKNOWLEDGMENT

We would like to acknowledge the guidance of my teacher for his invaluable guidance and suggestions in making this case report. We would like to thank the management of Sri Venkateswara Institute of medical sciences and Sri Padmavati medical college for women for their incredible support.

CONFLICT OF INTEREST

Nil

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Cite this article:
Suneel Babu TM, Sai Nelatha V, Vasantha K, Robin George. Dermatomyositis Associated With Interstitial Lung Disease: A Case Report. *International Journal of Pharmacy & Therapeutics*, 11(1), 2020,1-4.
DOI: **http://dx.doi.org/10.21276/ijpt.2020.10.1.1**

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