



First report of an adult patient with clinical amyopathic dermatomyositis associated rapid progressive interstitial lung disease triggered by COVID-19

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Received 10 Apr. 2022

Accepted 24 May 2022

Published online 31 May 2022

Keywords: Dermatomyositis, Interstitial lung disease, COVID-19

Abstract

Interstitial lung disease (ILD) is a common manifestation of dermatomyositis (DM) with indolent course but a unique variant of DM called clinically amyopathic DM (CADM) presents with rapid progressive ILD (RP-ILD) and is called CADM associated RP-ILD. Here, we report a case of a 52-year-old woman with CADM associated RP-ILD occurred 2 weeks after recovery from COVID-19 infection but unfortunately died from RP-ILD, almost 5 months later. This is the first report of CADM associated RP-ILD triggered by COVID-19 in an adult patient.

Citation:

Varghaei A, Mojdeganlou H, Rabieepour M, Mikaeilvand A, Abbasi A. First report of an adult patient with clinical amyopathic dermatomyositis associated rapid progressive interstitial lung disease triggered by COVID-19. *Immunopathol Persa.* 2022;x(x):e32406. DOI:10.34172/ipp.2022.32406.

Introduction

It is more than two years that the world is dealing with COVID-19 infection which its main manifestations are fever and respiratory distress; however, within this pandemic, other diseases which can present with interstitial lung disease (ILD) and cause respiratory distress should not be missed. ILD is a common manifestation of dermatomyositis (DM) with indolent course (1) but a unique variant of DM called clinically amyopathic DM (CADM) presents with rapid progressive ILD and is called CADM associated RP-ILD (2). Here, we report a case of a 52 year old woman experienced aggressive lung damage and skin problems after recovery from COVID-19. She had typical dermatologic signs of DM and was diagnosed with CADM associated RP-ILD. She died from RP-ILD, almost four months later due to COVID-19 infection. There is one report of CADM associated RP-ILD after COVID-19 in pediatric patients (3) but our case is the first of its kind in adults.

Case Presentation

A 52-year-old female with a history of hypothyroidism, was afflicted by COVID-19 in October 2020 with positive polymerase chain reaction (PCR) test. She had fever,

Key point

Clinically amyopathic DM associated RP-ILD triggered by COVID-19 is a rare but a fatal complication of COVID-19 infection. Prognosis of this clinical condition is poor and despite administration of high dose immunosuppressive therapy patients may not survive.

chills, cough, muscle ache, and weakness. She was admitted in hospital for five days and then discharged with good condition. About two weeks later, some erythematous itchy patches presented on metacarpophalangeal joints (MIPs) and proximal interphalangeal joints (PIPs), which followed by fever, cough, and dyspnea. As dyspnea worsened, she admitted for the second time, which was about 1 month after the first admission. She received intravenous immunoglobulin (IVIG), plasmapheresis, remdesivir, and interferon beta-1a for treatment. Computed tomography (CT) scan revealed ground-glass opacities in 40-50% of both lungs. She was discharged after one week with partially improvement in her symptoms however, her skin lesions gradually dominated in the following days. Erythematous patches were added on elbows, as well as telangiectasias

on nail-folds. Additionally, erythematous papules on MIPs and PIPs became sharper, which raise the suspicion of DM and simultaneously her respiratory symptoms flared up again (Figure 1). The deterioration of her condition and the development of dyspnea, fever, and cough led to third admission. The PCR test for COVID-19 was conducted again but results came negative. The test was repeated to rule out probable false negative results since, the repeated test was also negative. Echocardiography findings were normal too. According to clinical findings and negative COVID-19 PCR tests the diagnosis of RP-ILD was considered. Another CT scan was performed during the third admission and showed bilateral ground-glass features in almost 70% of lungs with evidence of pneumothorax (Figure 2). The worsening of dyspnea, elevated respiratory rate (>40/min), and an O₂ saturation <85% with non-invasive ventilation, prompted the medical team to use mechanical ventilation and intensive care unit (ICU), admission on the 6th day of the third admission. She received 3 g methylprednisolone as pulse therapy, 1 g cyclophosphamide and IVIG accordingly.

The patient's skin lesions were consistent with DM according to dermatologist consultation. In order to confirm DM skin biopsy was performed which the pathologic evaluation confirmed the diagnosis of DM. Further evaluation for autoimmune antibodies including ANA, anti Jo-1, anti-Ro, anti-La, anti-dsDNA, carcinoembryonic antigen, CA19-9, CA125 and also aldolase was performed and the results were in the normal range. In the absence of weakness or decreased muscle force, and considering laboratory results, the diagnosis of CADM was confirmed by dermatologist. Unfortunately, the therapeutic efforts were not successful and the patient died on the 28th day of the third admission.

Pathologic evaluation

Skin biopsy showed epidermal atrophy with presence of some apoptotic cells and basal layer vacuolization. Basal layer is thickened. Dermis showed neutrophilic infiltration and nuclear debris especially in upper dermis



Figure 1. Erythematous papules on MIPs and PIPs, known as Gottron papules, hallmark of dermatomyositis.



Figure 2. Computed tomography (CT) scan showing extensive bilateral ground glass appearance with evidence of pneumothorax.

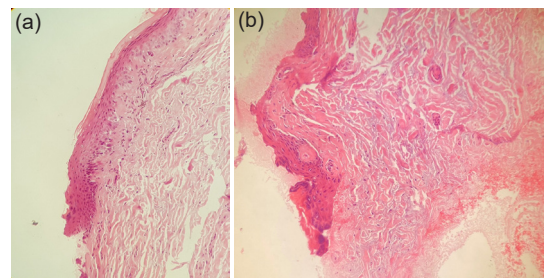


Figure 3. (a) Skin tissue showing basal layer vacuolization (H&E, 40×); (b) Dermal area of the previous figure (a) showing dermal mucinosis (H&E, 40×).

with evidence of mucinosis (Figure 3a and 3b).

Discussion

To best of our knowledge this is the first report of CADM associated RP-ILD in adults triggered by COVID-19. DM is an autoimmune disorder with different manifestations. ILD during DM is common. In CADM, patients experience a rapidly progressive ILD which has a very dismal prognosis (4). The patients usually present by skin manifestations of DM including rash, Gottron papules and elevated serum creatine kinase levels but the symptoms suddenly aggravate to fever and progressive respiratory distress. Radiologic studies usually show diffuse ground glass opacities in both lungs (5) and most patients die in a short time after the disease (2). This poor outcome is supposed to be related to high titers of autoantibodies specially anti-melanoma differentiation associated gene 5 (anti-MDA5) (6,7) and anti-Ro52 although some patients had no autoantibodies in their blood (8,9). Unfortunately, anti-MDA5 was not available in our center and we could not check it in our patient but her anti-Ro52 antibody was negative.

Considering the current pandemic situation of COVID-19, patients presenting by fever and respiratory distress are primarily considered as COVID-19 infection. Besides, it should be kept in mind that various diseases especially connective tissue disorders may have similar clinical presentations and thorough evaluation of patients and considering differential diagnoses is mandatory. In our case, the patient had a history of COVID-19 infection

with confirmed nucleic acid test 2 weeks before her first skin manifestations. Since COVID-19 may also have dermatologic manifestations it is recommended that every patient with presentations suspicious of COVID-19 infection and skin manifestations should be evaluated by a dermatologist for definite diagnosis. As in our case, there would be challenges in clarifying whether the patient's clinical deterioration is due to COVID-19 or CADM associated RP-ILD. Since the clinical presentation and also radiologic findings are very similar repeated nucleic acid test to rule out COVID-19 would be mandatory.

Conclusion

In summary we reported a case of CADM associated RP-ILD triggered by COVID-19 in a 52-year-old woman. To the best of our knowledge it is the first report of its kind in the literature. Prognosis of this clinical condition is poor and despite administration of high dose immunosuppressive therapy our patient did not survive. There are recent reports claiming that COVID-19 can trigger autoimmune diseases such as autoimmune hemolytic anemia and thrombocytopenia (10). Moreover, there are few reports of autoimmune thyroiditis, emerged after COVID-19 infection (10). Since our patient had no prior history or any signs connective tissue disorders and the manifestations of DM were observed shortly after COVID-19 infection, we concluded that the disease was triggered by COVID-19 infection.

Authors' contribution

AA, AV and MR were the principal investigators of the study. HM, AA, AV and AM were included in preparing the concept and design. AV and AA revisited the manuscript and critically evaluated the intellectual contents. All authors participated in preparing the final draft of the manuscript, revised the manuscript and critically evaluated the intellectual contents. All authors have read and approved the content of the manuscript and confirmed the accuracy or integrity of any part of the work.

Conflicts of interest

The authors declare that they have no competing interests.

Ethical issues

This study was approved by the ethics committee of the Urmia University of Medical Sciences. The patient's legal guardian gave the consent to publish as a case report. Besides, ethical issues (including plagiarism, data fabrication, double publication) have

been completely observed by the authors.

Funding/Support

None.

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