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A case of COVID-19 in a patient with pemphigus on azathioprine: Successful management with methylprednisolone

Abdullah Demirbaş[1], Ömer Faruk Elmas[2], Mustafa Atasoy[3], Ümit Türsen[4], Asuman Kilitci*[5], Torello Lotti[6]

[1]*Department of Dermatology, Kocaeli University, Faculty of Medicine, Kocaeli, Türkiye*
[2]*Department of Dermatology, Medicana International Hospital, İstanbul, Türkiye*
[3]*Department of Dermatology, Memorial Ataşehir Hospital, İstanbul, Türkiye*
[4]*Department of Dermatology, Mersin University, Faculty of Medicine, Mersin, Türkiye*
[5]*Department of Pathology, Düzce University, Faculty of Medicine, Düzce, Türkiye*
[6]*Department of Dermatology, Guglielmo Marconi University, Rome, Italy*

ABSTRACT

COVID-19 is caused by severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2) virus that first appeared in Wuhan, China. It is not just a respiratory disease but a multisystem disease. Risk factors for mortality and morbidity include advanced age, male gender, hypertension and immunosuppressive conditions. We present the case of COVID-19 that developed in a 31-year-old female patient who received azathioprine therapy with a diagnosis of pemphigus.

Key words: Pemphigus, COVID-19, azathioprine, methylprednisolone.

* *Dr. Asuman Kilitci,*
Department of pathology, Düzce University, Faculty of Medicine, Düzce, Türkiye
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Introduction

Coronavirus disease 2019 (COVID-19) is caused by severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2) virus that first appeared in Wuhan, China [1]. COVID-19 is not just a respiratory disease but a multisystem disease. This is considered one of the most important reasons for high rates of mortality and morbidity. Other risk factors for mortality and morbidity associated with COVID-19 include advanced age, male gender, hypertension, and immunosuppressive conditions. Pemphigus is an autoimmune blistering mucocutaneous disease characterized histologically by intraepidermal blisters owing to the loss of cell-to-cell adhesion of keratinocytes and immunopathologically by immunoglobulin G (IgG) autoantibodies directed against the cell surface of keratinocytes. Patients with pemphigus are generally considered to be vulnerable to infections due to disruption of the epithelial barrier and immunosuppressive treatments [2-4,5].

Case reports

A 31-year-old female patient presented with fatigue, cough, and back pain for two days. She had a diagnosis of pemphigus vulgaris confirmed by histopathology and direct immunofluorescence (DIF) three years before.

No accompanying systemic disease was known. She was in remission with 100 mg/day azathioprine monotherapy and had close contact with a person with a known COVID-19 patient, three days before symptoms began. The nasopharyngeal swab sample was positive for SARS-CoV-2. Blood investigations including complete blood count, ferritin, C-reactive protein, erythrocyte sedimentation rate, d-dimer, fibrinogen, serum electrolytes, liver function tests, and renal function tests were within normal limits. Chest computed tomography scan showed no infiltration. No abnormality was found in her dermatological examination. Azathioprine was discontinued immediately and an oral 10 mg daily dose of methylprednisolone treatment was started, and gradually tapered by 2 mg every 10 days. No specific antiviral treatment was used. During the infection, the patient experienced only mild symptoms and was symptom-free after 10 days following the swab test. A 100 mg daily dose of azathioprine re-started and while she was on 2 mg/day methylprednisolone, three vesicles appeared in the buccal mucosa which were regressed spontaneously without dose adjustment. The patient remained in remission with a combination of 2 mg/day methylprednisolone and 100 mg/day azathioprine.

Discussion

Pemphigus vulgaris is the most common type of autoimmune bullous diseases, which usually affects adults. High mortality rates of the disease have significantly decreased thanks to common use of systemic corticosteroids and immunosuppressants. However, immunosuppressive drugs disrupt the mucosal and cutaneous integrity and make the patients vulnerable to infections [6]. High-dose systemic corticosteroids, azathioprine, mycophenolate mofetil, cyclosporine, cyclophosphamide, rituximab, and IVIG therapies are well-known treatment options pemphigus vulgaris. Apart from IVIG, all these options are considered to pose a risk for COVID-19 infection. Some authors recommended that, in patients with pemphigus vulgaris immunosuppressive therapy should be suspended until COVID-19 is taken under control, while others suggested that these treatments should only be stopped in confirmed SARS-CoV-2 infections [4].

Systemic corticosteroids are usually considered the first step in pemphigus. Although corticosteroids have revolutionized the therapeutic approach toward pemphigus, treatment complications such as the risk of serious infections, which has become more pronounced with the COVID-19 outbreak, are still of concern. It has been recently suggested that the cytokine storm in patients COVID-19, involves the overproduction of pro-inflammatory mediators causing increased vascular permeability. Notwithstanding the concerns that corticosteroids may disrupt viral clearance, the low dose systemic corticosteroids seem to have an exceptional role in the management of severe SARS-CoV-2 infection [7,8]. Kasperkiewicz et al suggested that immunomodulatory treatments including systemic corticosteroids should be continued when needed since unjustified withdrawal may trigger disease aggravation. They also suggested that 10 mg or lower daily doses of prednisolone can be maintained in patients COVID-19 while doses higher than 10 mg/day can be decreased considering the activity of the disease, accompanying systemic diseases, and severity of COVID-19 [9]. The guideline prepared by the European Academy of Dermatology and Venereology Task Force Autoimmune Blistering Diseases suggested to discuss reducing the dose of systemic

corticosteroids for COVID-19 positive pemphigus patients [10]. Shakshouk et al. also suggested that systemic corticosteroids should be reduced gradually to the lowest efficient dose during the pandemic [2].

To sum up, low-dose methylprednisolone may provide a safe and successful management of pemphigus vulgaris in patients with COVID-19. It is obvious that more reports are needed to empower conclusions deduced in the present report.

Main Points

1. Despite concerns that corticosteroids can interfere with viral clearance, low-dose systemic corticosteroids tend to play an exceptional role in the management of serious SARS-CoV-2 infections.
2. Low doses of prednisolone may be retained in patients with COVID-19 but higher doses will need to be reduced as the disease becomes more active.
3. We think that low-dose methylprednisolone can provide safe and successful management of pemphigus vulgaris in COVID-19 patients.

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