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LETTER TO THE EDITOR

Atypical antinuclear matrix protein 2-positive dermatomyositis presenting with anasarca and bulbar weakness after coronavirus disease 2019 infection requiring mechanical ventilation

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A 28-year-old female presented with a six-week history of rapid onset proximal muscle weakness, dysphagia, and inflammatory rash (heliotropic rash, Gottron papules, and livedo reticularis). Of note, three weeks prior to developing symptoms the patient had tested positive for severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2). The manual muscle testing score 8 (MMT8) was 72/150. The patient had marked anasarca. Laboratory assessment revealed high creatine kinase (CK) 9270 IU/L. Extended myositis panel revealed a positive antinuclear matrix protein 2 (NXP2) antibody. Full body MRI showed extensive muscle edema in the adductor and flexor muscle groups of the shoulder and hip girdle and erector spiny muscles. Muscle biopsy and skin biopsies were both consistent with dermatomyositis (DM). Computed tomography-positron emission tomography showed evidence of intense muscle uptake, particularly in the proximal muscles of the forearms, without any findings suggestive of an underlying malignancy. The initial treatment consisted of high dose intravenous methylprednisolone (1.5 g in total), followed by oral taper and a course of intravenous immunoglobulin (2 g/kg). The patient was initiated on mycophenolate 500 mg twice daily.

The patient's condition worsened, and two weeks following the initial presentation, the patient was bed-bound with worsening dysphagia, ascites, anasarca, and bilateral pleural effusions in the setting of low albumin (26 g/L). Due to deteriorating peak flow measurements at ward level, the patient was moved to the intensive treatment unit where the patient was intubated for airway protection. Magnetic resonance imaging of the brain and acetylcholine receptor antibodies were normal. Given the severity of symptoms and lack of response to treatment, the combination of intravenous 6-hourly 100 mg hydrocortisone, 500 mg of cyclophosphamide every two weeks (3 g in total), and 1 g of rituximab at days one and 15 was prescribed. Steroids were tapered slowly. The patient received one further course of intravenous immunoglobulin due to evidence of residual disease activity. Symptoms slowly improved with recovery of muscle function (MMT8=147/150) and the resolution of skin rash and dysphagia two months following rituximab treatment. CK peaked at 21,525 IU/L before slowly returning to normal range. The patient remains well six months after discharge, has been switched to mycophenolate 500 mg twice daily maintenance therapy, and prednisone has been discontinued.

During the coronavirus disease 2019 (COVID-19) pandemic, evidence suggested that 10% of COVID-19 patients developed myopathic symptoms along with hyperCKemia.[1] Recently,

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there have been case reports of DM following SARS-CoV-2 infection, specifically cases relating to NXP2 antibodies.[2] NXP2 binds to viral RNA (ribonucleic acid) and is an important mediator of viral transcription.[3] It has been hypothesized that NXP2 antibodies can be induced by both the SARS-CoV-2 virus and messenger RNA vaccines. Presence of anti-NXP2 has also been associated with a more severe SARS-CoV-2 infection course.[4,5]

In this report, we describe an atypical case of COVID-19-related DM, marked by several factors associated with an unfavorable prognosis and limited treatment response. While existing case reports featuring these attributes are scarce, they hint at a potential connection between anasarca and bulbar weakness, suggesting the possibility of a rare DM subtype.[6-8] To the best of our knowledge, this case report represents the first documented instance of a severe and refractory COVID-19-related DM patient presenting with such features to be treated successfully.

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Data Sharing Statement: The data that support the findings of this study are available from the corresponding author upon reasonable request.

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