Case Report

Once in a blue moon: A rare case of Duchenne muscular dystrophy complicated by COVID-19

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ABSTRACT

Patients suffering from Duchenne Muscular Dystrophy (DMD) are at higher risk of suffering significant morbidity resulting from COVID-19, considering their pre-existing respiratory insufficiency and immunocompromised state. We present such a case who was admitted to our intensive care unit. A 21-year-old patient, who was a diagnosed case of DMD since the age of 8 and on treatment with steroids, angiotensin-converting enzyme inhibitors, and intermittent home-oxygen support, presented with fever and breathlessness and was diagnosed to have COVID-19 pneumonia. Oxygen support was provided by non-invasive ventilation (NIV), along with therapeutic and supportive treatment, namely, azithromycin, remdesivir, dexamethasone, and heparin. Dyselectrolytemia was corrected and convalescent plasma was transfused. The patient was weaned off NIV and discharged on significant improvement in his general condition. Although the treatment of COVID-19 using convalescent plasma has now fallen out of favor, we found some clinical improvement in our patient. DMD complicated by COVID-19 can seem like a daunting challenge, but providing fundamental, yet, simple treatment measures goes a long way in the patient care.

Key words: Convalescent plasma, COVID-19, Critical care medicine, Duchenne muscular dystrophy

The pandemic caused by the severe acute respiratory syndrome (SARS-CoV-2) virus has ravaged the world since December 2019. In addition to causing symptoms of the common flu, it can exacerbate into a severe infection causing pneumonia, acute respiratory distress syndrome, and even myocarditis. It is established that the disease often has a much more severe course in those who have concurrent pre-existing medical conditions, such as diabetes, hypertension, or renal failure.

Duchenne's muscular dystrophy (DMD) is an X-linked recessive disease that is caused by dystrophin gene mutation and leads to progressive skeletal and cardiac muscular atrophy [1]. DMD patients are at higher risk of suffering significant morbidity and mortality resulting from COVID-19 as they have respiratory insufficiency and are often on chronic treatment with corticosteroids leading to immunosuppression [2]. Passive immunization has previously been used with some degree of varying success in treating these patients for respiratory diseases.

We would like to present a case of DMD who was admitted to the Intensive Care Unit (ICU) in JSS Hospital, Mysuru due to COVID-19 infection, and the various challenges faced while treating this patient.

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CASE REPORT

A 21-year-old male patient presented to the emergency department with 5 days history of fever, easy fatigability, and breathlessness. He reported no other symptoms. The patient was a known case of DMD since 2002, diagnosed by multiplex polymerase chain reaction and showing deletion of a segment of the DMD gene involving exons 46–47. The patient was on occasional home oxygen support, angiotensin-converting enzyme (ACE)-I therapy, and steroids for his condition.

On examination, the patient was conscious and oriented, with a pulse rate of 130/min, blood pressure of 110/70 mmHg, oxygen saturation of 90% at room air, and respiratory rate of 37/min. Laboratory examination revealed a hemoglobin of 15.2 g/dL, total leukocyte count of 7130/μL (neutrophils-83.5%, lymphocytes 14.3%, eosinophils 0.0%, monocytes 2.1%, and basophils 0.1%), neutrophil-lymphocyte ratio of 6 indicative of mild stress, low procalcitonin value, and elevated inflammatory markers. Serum electrolytes were suggestive of hypokalemia and hyponatremia (Sodium-115 mEq/L and potassium-2.5 mEq/L).

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Oxygen was delivered to the patient through non-invasive ventilation (NIV) by the orofacial mask interface (pressure support mode-FiO_2-60%, PEEP-6 cmH_2O). Potassium correction measures were employed – 60 mEq KCl over 12 h was administered intravenously. Treatment with antibiotics (Inj. Ceftriaxone-Sulbactam 1.5 g 1-0-1, Tab. Azithromycin 500 mg 1-0-0, and Cap. Doxycycline 100 mg 1-0-1), antivirals (Inj. Remdesivir 200 mg IV followed by 100 mg 1-0-0 IV for a total of 5 days), steroids (Inj. Dexamethasone 6 mg 1-0-0), and anticoagulants (Low molecular weight heparin 40 mg subcutaneously 1-0-0) was started. The patient was monitored with serial arterial blood gas analysis twice a day and serum lactate dehydrogenase, ferritin, and electrolytes (Table 1).

Over the next few days, the potassium levels were corrected. Serial X-rays were taken on the 1st, 3rd, 5th, and 7th days of ICU stay (Fig. 1). NIV was continued over a period of 5 days with subsequently decreasing FiO_2 after 3 days (Fig. 2). 2D echo on day 2 was within normal limits. On the 5th day of admission, convalescent plasma was taken from an aged male donor who recovered from COVID-19 infection as higher neutralizing antibodies are expected in this subset of patients (as neutralizing antibody levels were not available at the time) [3].

Dyselectrolytemia was completely resolved, following which a considerable decrease was noted in the respiratory rate and oxygen requirements, with substantial improvement in the work of breathing. The patient developed ICU delirium for which he was counseled daily over video call by a known clinical psychologist who was consulted 3 years ago for depression. He was discharged on day 7 of admission for home care by teleconsultation with home oxygen support.

Table 1: Serial arterial blood gas analysis values on each day of ICU stay

Date	pH	paO_2	$paCO_2$	HCO_3-	Lactate	FiO_2
September 15, 2020	7.420	37.9	32.8	20.9	1.3	60
September 16, 2020	7.418	114	26.9	17.1	1.8	60
September 17, 2020	7.459	41.1	28.8	20.1	1.6	40
September 18, 2020	7.527	164	19.4	16.0	0.7	40
September 19, 2020	7.440	44.0	33.0	22.0	1.6	40
September 20, 2020	7.443	44.8	36.7	24.7	1.5	40
September 21, 2020	7.447	180	29.8	21.8	0.7	21

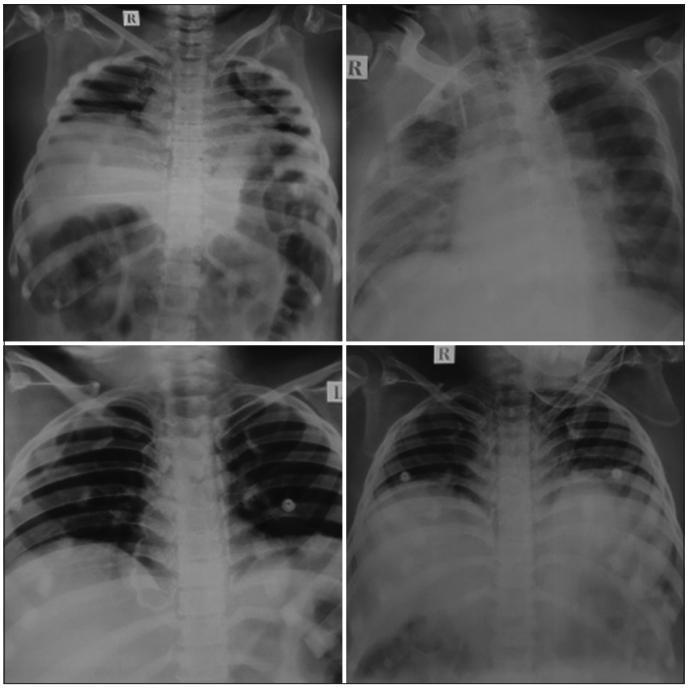

Figure 1: Serial X-rays taken on 1st, 3rd, 5th, and 7th days of intensive care unit stay

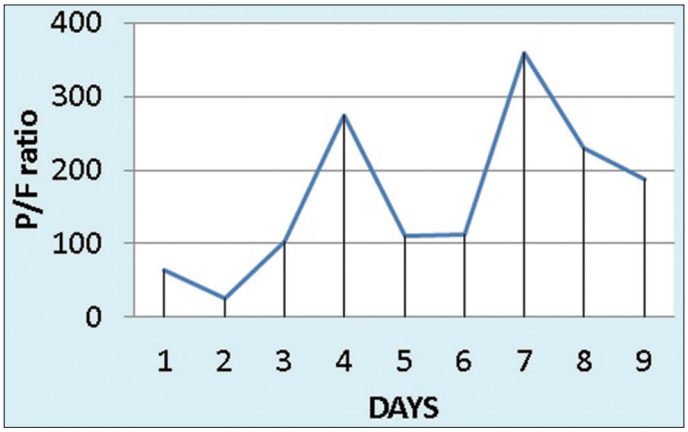

Figure 2: P/F ratio depicted on each D day of intensive care unit stay

DISCUSSION

DMD leads to progressive skeletal and cardiac muscular atrophy. It requires management strategies and a multidisciplinary approach to tackle this complicated condition; however, even the most aggressively treated patients have an average life expectancy of 28 years. This disease eventually results in loss of respiratory reserve and cardiac ailment. DMD patients are at higher risk of suffering significant morbidity and mortality resulting from COVID 19 as they have respiratory insufficiency and are often on chronic treatment with corticosteroids leading to immunosuppression.

Of the few studies that have been conducted and the opinion of several expert panels, DMD patients should continue their corticosteroid therapy during this pandemic. Periodic assessment of respiratory function, physiotherapy, rehabilitation therapy, etc., should be continued as far as possible. The patients who are on therapy with ACE inhibitors or angiotensin receptor blockers should continue their drugs, even though the pathophysiology of the virus is due to its fusion with the ACE2 receptor. NIV is preferred as much as possible, to facilitate the removal of secretions. Electrolyte disturbances, especially hypokalemia, must be corrected. Clinical severity is assessed by O_2 requirements

of more than 6 L/min or respiratory distress with the presence of respiratory failure and cardiac involvement [4,5]. The presence of severe thoracic deformities and multi-systemic comorbidities such as obesity and hypertension often worsen the clinical scenario.

Passive immunization with antibodies has been used to treat infectious diseases for hundreds of years. It has improved humoral immunity and its use has been extended to the discovery and licensing of monoclonal antibodies for disease treatment and even post-exposure prophylaxis. In the early 20th century, plasma therapy was also used with some degree of success during the Influenza pandemic. More recently, serum therapy and the use of immunoglobulins have been used for the respiratory syncytial virus, cytomegalovirus, SARS, and the Ebola virus [6].

Several multicentric, randomized, and control trials on the efficacy of convalescent plasma in COVID-19 were conducted worldwide during 2020–2021 when supportive therapy seemed to be the only treatment modality. Before the authorization of usage of corticosteroids and Remdesivir in 2020 by the Food and Drug Administration (FDA), high titer plasma therapy was indicated [7]. Oritgoza *et al*. studied plasma efficacy by measuring the participant scores on the 11-point World Health Organization Ordinal Scale for Clinical Improvement (WHO scale) on day 14, day 28, and 30 day mortality rate. They concluded that further studies were needed to understand interactions between plasma, corticosteroids, and remdesivir. However, a possible benefit of convalescent was observed early in the pandemic when high titer convalescent plasma was used and corticosteroids and remdesivir were not in use [8]. Plasma therapy may be a feasible treatment option when other therapies are not in use or unavailable. In a retrospective study conducted by Joyner *et al*., convalescent plasma was identified as a potentially beneficial therapy in hospitalized patients with COVID-19. They found that among patients with COVID-19 who were not receiving mechanical ventilation, the transfusion of plasma with high antibody levels was associated with a lower risk of death than the transfusion of plasma with low antibody levels. In addition, patients who received plasma within 3 days after the diagnosis of COVID-19 had a lower risk of death than those who received a transfusion later in the disease course. They concluded that the benefit of convalescent plasma was more in patients who received plasma transfusions containing higher levels of anti-SARS-CoV-2 IgG antibodies early in the disease course [9]. On the contrary, the multicentric, randomized, and control PLACID trial conducted in India in 2020 found that convalescent plasma did not reduce 28-day mortality or progression to severe disease in patients admitted to the hospital with moderate COVID-19 [10].

At present, the FDA's most recent revision of the Emergency Use Authorization for COVID-19 convalescent plasma, published December 28, 2021, limits treatment with high-titer COVID-19 convalescent plasma to patients who have immunosuppressive disease or are receiving an immunosuppressive treatment, into which treatment profile our patient fits in [11]. Despite having many of the complications mentioned above including thoracic deformities, obesity, hypokalemia, and restrictive lung disease, our patient showed significant improvement with the above simple therapeutic measures, including convalescent plasma administration.

CONCLUSION

A concurrent diagnosis of COVID-19 and DMD can initially be intimidating to physicians and many arrive at the foregone conclusion that such patients ultimately have a poor prognosis. However, targets and measures taken to solve basic critical care issues can significantly improve such patient's outcomes.

CONSENT

Informed and written consent was obtained from the patient's party.

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