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Successful Treatment of Post-COVID Severe ANCA Associated Vasculitis' Case; Could Faster Treatment Be Better Prognosis?

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Abstract

The SARS-CoV-2 pandemic has become the most serious health problem of today globally. Kidney involvement in patients with coronavirus disease 2019 (COVID-19) is common and associated with high mortality. Although acute tubular necrosis due to hemodynamic instability is the most common cause, other complex and destructive processes related to cytokine storm and activation of innate and adaptive immunity have also been reported.

Herein, we present successful treatment of proteinase-3 anti-neutrophil cytoplasmic antibodies (PR3-ANCA)-associated vasculitis case presenting with severe pulmonary-renal syndrome as a rare and fatal complication of COVID-19 infection.

Keywords: AKI; ANCA; COVID-19; cresentic glomerulonephritis; pulmonary-renal syndrome

List of Abbreviations

AAV: Anca-associated vasculitis; GFR: Glomerular filtration rate

Introduction

Coronavirus disease 2019 (COVID-19) caused by severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2) results with a high degree of morbidity and mortality since its appearance in late 2019. While most COVID-19 cases are asymptomatic or only display mild influenza-like symptoms, a significant number of patients may experience severe pneumonia, acute respiratory distress syndrome, multiple organ failure, and even death.1 COVID-19 causes an exaggerated immune response, especially in susceptible individuals.2 Epitope expansion and antigen mimicry are the first triggers for antibody production.3 Antineutrophil cytoplasmic antibody (ANCA) associated vasculitis (AAV) is a systemic disease that causes vasculitis in various organs such as kidney, lung, skin and intestine. Although the exact mechanism for development of the disease is not clearly understood, infection has been reported to be a triggering factor especially by cytokine storm.4,5 AAV requires rapid diagnosis and treatment. Otherwise, delay in diagnosis and subsequent treatment makes morbidity and mortality as inevitable.6,7 On the other hand, in cases of severe organ involvement, urgent diagnosis of the disease may be very difficult which can delay therapy, hence worsen prognosis. We present a previously healthy 51-year-old case of new onset severe ANCA-associated vasculitis (AAV) following a mild COVID-19 infection who presented with both acute kidney injury (AKI) and diffuse alveolar hemorrhage (DAH), successfully treated empirically before identifying ANCA positivity.

Case Description

A 51-year-old man with a no prior comorbidity referred to our hospital with AKI, anuria and hemoptysis. The patient, who had two doses of biontech mRNA vaccine (last one 8 months ago), was positive for COVID-PCR test about 2 months ago due to complaints of runny nose, fever and fatigue. No hospitalization or medical treatment was required during the COVID-19 infection and the patient became PCR-negative. However, fifteen days after the PCR negativity, the patient started to complain of weakness, loss of appetite, joint pain and nausea. Hemoptysis started with coughing almost every day. In the last ten days, the patient's urine output gradually decreased, He was admitted to the local hospital with these complaints. Chest computed tomography (CT) showed cavitary lesion with central opacification in the right pulmonary apex and ground-glass shadowing in the lungs bilaterally. (Figure 1) Bacterial and fungal infection of lungs were ruled out. Also, Tuberculosis culture of sputum, asido-resistance bacilli (ARB) staining and immunological tests including PR3-ANCA and MPO-ANCA, ANA, anti-GBM were all negative. The patient, whose creatinine level increased progressively is referred to our hospital because of his worsening clinic, need of hemodialysis and evaluation for pulmonary renal syndrome. At admission, labs revealed serum creatinine 12.02 mg/dl, estimated glomerular filtration rate (eGFR) 4 ml/min/1.73 m^2, blood urea nitrogen (BUN) 118.65 mg/dl, albumin 2,7 g/dl and C-reactive protein level of 292 mg/dl. In the urine analysis proteinuria and erythrocyturia were found. Severe anemia was observed with normal platelet count and elevated white blood cells count. On admission, the patient was tachypneic and hypoxic. Within two hours of admission, he was urgently intubated due to respiratory failure secondary to sudden developed massive pulmonary hemorrhage. The clinical appearance was so devastating that treatment with IV pulse steroid 1000 mg and plasmapheresis was started immediately without serologic and/or tissue diagnosis by kidney biopsy. However, re-examination of serologic tests revealed positive result for PR-3-ANCA. Then, 500 mg intravenous pulse cyclophosphamide applied, and plasmapheresis treatment was completed to 5 sessions. After induction with 1000 mg pulse steroid for three days, steroid dose decreased gradually to 80 mg intravenous prednisolone as maintenance dose. The patient was taken to intermittent hemodialysis due to uremia and anuria. Tracheostomy was performed in the intensive care unit for the patient since being intubated for a long time. The patient's hypoxia regressed day by day, urine output gradually increased, his tracheostomy was closed, kidney functions improved, and he was discharged with recovery after a 50-day intensive care hospitalization. (Figure 2, Figure 3) We planned maintenance therapy with 60 mg oral prednisolone daily and 500 mg pulse cyclophosphamide with 15 days intervals.

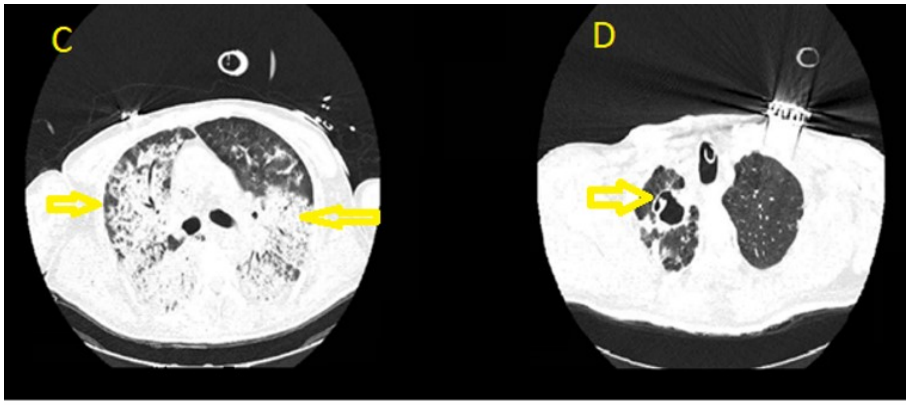

Figure 1: Thoracic computed tomography images showing a widespread consolidation with central and peripheral distribution bilaterally, These findings are suggested of diffuse alveolar hemorrhage (C), also a cavitary lession seen in right pulmonary apex (D).

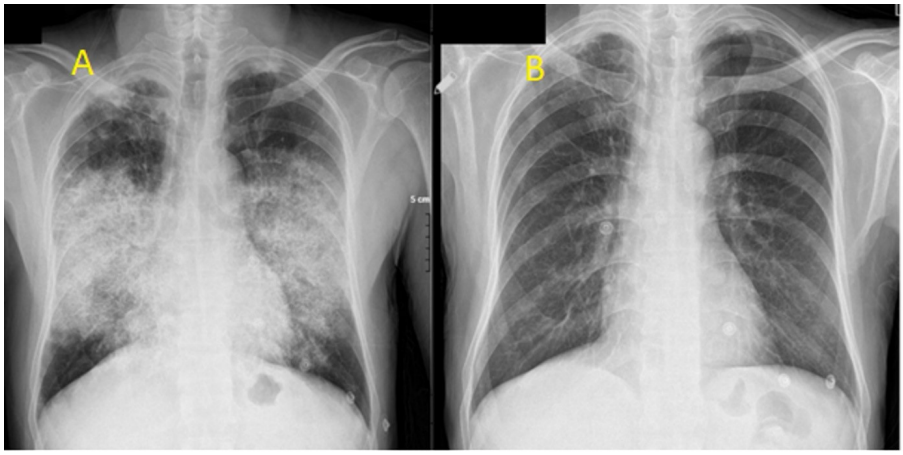

Figure 2: Chest X-ray (Posteroanterior view) findings at presentation (A), and 8-weeks after (B)

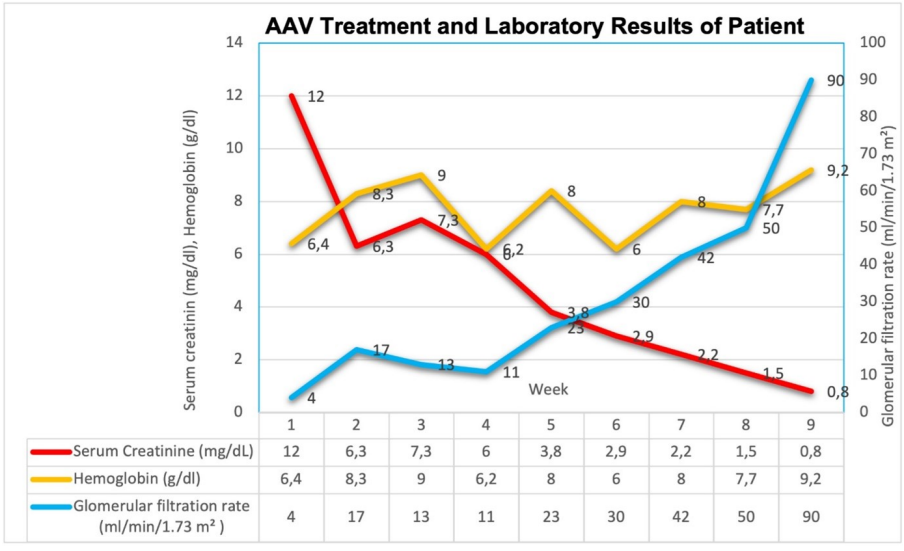

Figure 3: IV pulse steroid 1000 mg and plasmapheresis was started immediately. Then, 500 mg intravenous pulse cyclophosphamide applied, and plasmapheresis treatment was completed to 5 sessions. After induction with 1000 mg pulse steroid for three days, steroid dose decreased gradually to 80 mg intravenous prednisolone as maintenance dose. In his follow-ups, urine output gradually increased, and there was no need for hemodialysis. The decrease in hemoglobin stopped and serum creatinine value decreased from 12 mg/dl to 0.8 mg/dl. GFR increased to 90 ml/min/1.73m².

Discussion

Herein, we present a case of a severe antineutrophil cytoplasmic antibodies (ANCA) vasculitis who presented with AKI which requiring hemodialysis and DAH after SARS-CoV-2 infection. Granulomatosis with polyangiitis is a small vessel vasculitis with a wide spectrum of presentation which may present with clinical pictures ranging from mild disease to life-threatening conditions resulting from massive alveolar hemorrhage. Immunosuppression is the corner stone of the treatment and if left untreated, the mortality risk increases dramatically.7 Although the cause of the AAV onset is unknown, infection has been reported to be an important suspected factor.4,5 It has been reported that the immune response and cytokine storm during SARS-CoV-2 infection and following period, a strong activation of the immune system with the release of pro-inflammatory cytokines and immune system

Conclusion

AAV is a serious disease and prompt treatment is one of the most important factors in patient survival.

Acknowledgement

None

Conflicts 0f Interest

The authors have no conflicts of interest to declare.

Consent

Written informed consent was obtained from the patient fort the publication of this case report.

Data Availability Statement

The datasets used and/or analyzed during the current study are available from the corresponding author on request.

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