International Journal of Research in Medical Sciences
Namala SP et al. Int J Res Med Sci. 2023 Jan;11(1):396-398
www.msjonline.org
pISSN 2320-6071 | eISSN 2320-6012

DOI: https://dx.doi.org/10.18203/2320-6012.ijrms20223673

Case Report

Evan's syndrome secondary to COVID-19 infection

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Received: 14 November 2022
Revised: 09 December 2022
Accepted: 15 December 2022

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ABSTRACT

Wide range of autoimmune diseases are known to occur following SARS-CoV-2 infection. There are very few case reports of Evan's syndrome secondary to COVID-19. We hereby report a case of Evan's syndrome secondary to COVID-19 infection and discuss its management.

Keywords: Evan's syndrome, COVID-19 infection, Thrombocytopenia, Autoimmune hemolytic anemia

INTRODUCTION

COVID-19 (Coronavirus disease 2019) is caused by the SARS-CoV-2 (Severe Acute Respiratory Syndrome Coronavirus-2).[1] Viruses for long are known to be a part of mosaic of autoimmunity with COVID-19 being no exception to it.[2] Recent studies suggest the occurrence of autoimmunity and production of auto antibodies in COVID-19 patients.[3] So far, there has been only one case report of Autoimmune hemolytic anemia with immune thrombocytopenia also known as Evan's Syndrome (ES) in a COVID-19 patient.[4]

CASE REPORT

A 30-year-old lady, from Nellore, Southern India, with no known comorbidities presented with complaints of shortness of breath on exertion for 20 days progressing to breathlessness at rest for 2 days with history of petechial rashes over extremities for 1 week duration. There was no history of fever, cough or sick contact. She was initially evaluated at outside hospital. On arrival at outside hospital, she was in shock and severe respiratory distress. She was intubated there in view of respiratory failure. Blood investigations done there showed severe anemia (Hb- 2.4 g%) and thrombocytopenia (Platelet count- less than 10,000 cells/mm^3). CT chest was suggestive of COVID-19 Pneumonia with bilateral diffuse subpleural GGOs but COVID-19 RT PCR was Negative. Tropical infection screening including MP QBC, Scrub serology, Dengue serology and Leptospirosis were negative. Direct Coomb's test was positive. She was managed with IV antibiotics, stress dose of steroids (Hydrocortisone), 7 units packed red blood cells, 8 units fresh frozen plasma, 2 units single donor platelet and 8 units random donor platelet transfusions. She was eventually extubated 4 days later and was referred to our hospital for further management.

In emergency room, patient complained of diminution of vision both eyes and shortness of breath. She was mildly tachycardic (Pulse rate-110 beats/minute) and hypoxic with oxygen saturation (SpO_2) of 88% in room air. She was initiated on oxygen supplementation via nasal canula. General examination was significant for pallor and icterus. Respiratory system examination revealed coarse crepitations in both lung fields. She had perception of light in both eyes.

Baseline investigations showed anemia (Hemoglobin-8.9 g%), neutrophilic leukocytosis (White blood cells-12490 cells/mm^3), severe thrombocytopenia (Platelet-less than

5000 cells/mm^3), elevated ESR (55 mm/hr), indirect hyperbilirubinemia (3.9). Peripheral blood smear showed predominantly microcytic hypochromic RBC's with nucleated RBC's, polychromasia, spherocytes, myeloid left shift with severe thrombocytopenia. Anemia workup was suggestive of iron deficiency anemia (Low transferrin saturations with normal Ferritin levels) along with elevated reticulocyte count (26%), LDH (460) with decreased haptoglobin levels (less than 1.8). The above investigations were keeping with hemolysis but direct Coombs's test (DCT) turned out to be negative. Our patient initially presented to another hospital with overwhelming hemolysis and had a hemoglobin of 2.4g% with DCT being positive there, following which she was heavily transfused. Therefore, the red cells that was subjected to DCT here in our hospital was a mixture of transfused and patient's own cells. This probably explains why the DCT turned out to be negative.

Bone marrow biopsy showed hypercellular marrow with erythroid and megakaryocytic hyperplasia keeping with the clinical and lab diagnosis of immune hemolytic anemia and thrombocytopenia. Viral Markers namely HIV, HBsAg and anti HCV were negative. C3, C4 levels were normal. ANA was positive (2+). Anti-ds DNA, ANCA, extractable nuclear antigens namely Sm, Sm/RNP, SSA (Ro), SSB (La), Scl-70, Jo-1 were negative.

CT chest was suggestive of COVID-19 with diffuse small patchy consolidations with surrounding GGO's in bilateral lower lobes (Figure 1). Though repeat COVID-19 RT PCR also turned out to be negative, she tested positive for anti SARS CoV-2. The diagnosis of COVID-19 Pneumonia was made clinico-radiologically.

Fundus examination revealed bilateral retinal hemorrhage with sub hyaloid bleeding involving macula of both eyes. She was transfused 1 unit of single donor platelets. In view of leukocytosis, blood and urine samples were sent for culture and sensitivity and was empirically started on IV antibiotics. Blood cultures flagged *Acinetobacter baumanii* and *Staphylococcus hominis*. Source reduction was done in the form of removal of central line (inserted at outside hospital) and IV antibiotics (Polymyxin B and Teicoplanin) were continued. Treatment with IV Ig (Immunoglobulins) for the ITP with hemolytic anemia (in view of Bacteremia) was planned. But due to financial constraints pulse IV methyl prednisolone 500 mg for 4 days under cover of antibacterial agents followed by oral steroids (1 mg/kg body weight) was given. She was also started on thrombopoietin analogue-Inj. Romiplostim 500 mcg weekly once.

Her oxygen supplementation was weaned off and her Vision improved gradually. Her counts showed steady improvement and on the day of discharge (after 7 days of steroids initiation), her hemoglobin was 8.9g%, white blood cell count was normalised to 7090 cells/mm^3 with a platelet count of 98,000 cells/mm^3. She was discharged on oral steroids along with pneumocystis pneumonia (PCP) prophylaxis and Iron supplements.

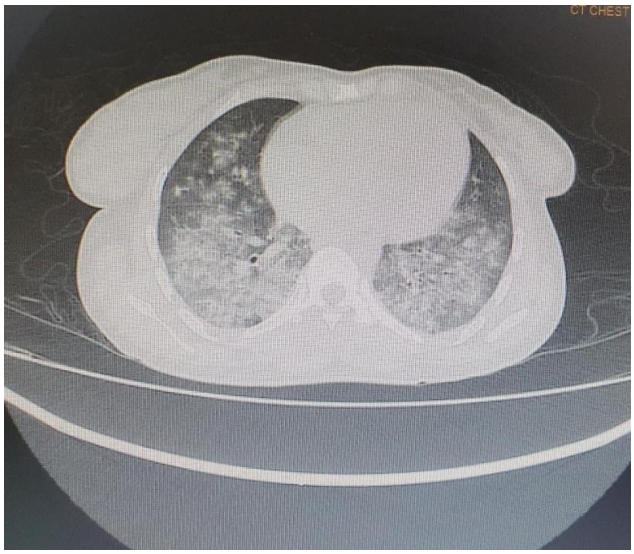

Figure 1: CT chest-suggestive of COVID-19 with diffuse small patchy consolidations with surrounding GGO's in bilateral lower lobes.

DISCUSSION

Wide range of autoimmune diseases are known to occur, after SARS-CoV-2 infection, ranging from systemic lupus erythematosus, autoimmune hemolytic anemia, immune thrombocytopenia to Guillain-Barré syndrome, vasculitis, multiple sclerosis and some autoinflammatory conditions in children.[5] Pathogenesis of these auto immune mechanism are thought to be due to molecular mimicry, immune dysfunction triggered by redistribution of immune cells and Immune reconstitution resulting in unregulated immune response.[5] There have been few cases reports of autoimmune hemolytic anemia and immune thrombocytopenia related to COVID-19.[6,7]

Our patient tested positive for anti-nuclear antibody (ANA). Though there hasn't been any prior documented ANA status of our patient, this could also be related to COVID-19 infection. Study by Gazzaruso et alhas reported prevalence of anti-nuclear autoantibodies (ANA) in 35.6% patient's admitted to the hospital for SARS-CoV-2 pneumonia.[3]

Occurrence of Evan's syndrome in association COVID-19 is a rare phenomenon. First-line therapy for Evan's syndrome in general is usually corticosteroids and/or intravenous immunoglobulin.[8] As our patient had bacteremia, it precluded the use of high dose steroids and IV Ig was the next ideal option.[9] Other therapies like Mycophenolate mofetil, cyclosporine, vincristine, azathioprine, sirolimus, and thrombopoietin receptor agonists can be considered second-line treatment options in ES independent of COVID-19 infection.[10] In view of affordability issues, she had to be treated with Steroids

and Thrombopoietin analogues, under the cover of appropriate antimicrobial agents.

Our patient showed excellent response with increment of both hemoglobin and platelet count to pulse IV steroids followed by oral steroids and thrombopoietin analogue. Hence steroids can be considered when it isn't feasible to use IV Ig. To the best of our knowledge, this also turns out to be the first case of Evan's syndrome secondary to COVID-19 which has been successfully managed with steroids and thrombopoietin analogues.

CONCLUSION

Evan's syndrome secondary to COVID-19 infection is a rare disease, which can be overlooked due to its ambiguous presentation. Hence high index of clinical suspicion and treatment at the time of initial presentation may help us in achieving favourable outcomes.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Namala SP, Marimuthu AK, Pandurangan P. Evan's syndrome secondary to COVID-19 infection. Int J Res Med Sci 2023;11:396-8.