Treatment of People with Evans Syndrome in the Setting of COVID-19 Pandemic

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SUMMARY

A new type of pneumonia had developed from Wuhan Province in China in December 2019, caused by a novel member of the Coronaviridae family named Severe Acute Respiratory Coronavirus 2 Syndrome (SARS-CoV-2) [1]. The disease is characterized by fatigue, dry cough, fever, and dyspnea [2]. In a more severe case, the picture may become more complicated by the onset of interstitial pneumonia with alveolar damage, which clinically can lead to severe Acute Respiratory Distress Syndrome (ARDS) and even death [3]. Since the initial outbreak, the epidemic has had a rapid global spread worldwide, which led the World Health Organization (WHO) to declare the disease now called COVID-19, a pandemic on 11th March 2020 [4].

In addition to the fact that COVID-19 is a highly infectious disease by its nature, people with autoimmune diseases have a higher infectious risk compared to the general population; because of an overall impairment of their immune system combined with the iatrogenic effect generated by corticosteroids and immunosuppressive drugs [5]. So, the COVID-19 pandemic is certainly conditioning the treatment strategy of such complex disorders as Evans Syndrome.

Evans syndrome is a rare disorder in which the body’s immune system produces antibodies that mistakenly destroy red blood cells, platelets, and sometimes neutrophils. This leads to abnormally low levels of these blood cells in the body (cytopenia). It was first described by Evan and Duane in 1951. Primary Evans syndrome with no cause is very rare and is seen in children. Secondary Evans syndrome may occur in association with other autoimmune disorders or lymphoproliferative disorders as a secondary disorder [6]. It is one of the rare presenting features of autoimmune disorders, especially Systemic Lupus Erythematosus (SLE). Evans Syndrome occur in patients with severe multisystem SLE manifestations and sometimes may even precede the onset of disease [7].

Evans syndrome is not considered a ‘serious’ health condition in the situation of the current COVID-19 pandemic. However, some of the treatments used for Evans syndrome will affect the immune system and may reduce the ability to fight the infection. So, we will state the treatment strategies of people with Evans syndrome in the setting of the COVID-19 pandemic.

No modification is required if patients are stable on low doses of immunosuppressive drugs. Changing treatments requires increased monitoring.
and could potentially result in relapse, and thus may be riskier than making no changes. For patients on higher doses of corticosteroids or immunosuppressive drugs, the use of IVIG (intravenous immunoglobulin) could allow for tapering and possibly discontinuation. Currently, there is a belief that rituximab should be avoided, and lower doses of cyclosporin may be as effective as full-dose [8].

Corticosteroids (CS) inhibit the immune response and delay the clearance of the pathogen. On the other hand, they suppress the host inflammatory response, which in the case of viral infections of the respiratory tract is the major responsible for lung damage and the occurrence of ARDS [9]. The latter represented the rationale for the widespread use of CS for the management of the Middle East Respiratory Syndrome (MERS)-CoV and SARS-CoV outbreaks [10,11] both histologically characterized by lung inflammation and diffuse alveolar damage [12]. Methylprednisolone has already been used in the treatment of COVID-19 in combination with antiviral (oseltamivir), antibiotics, and oxygen therapy [13]. Overall, no clear reason exists to expect that patients with COVID-19 infection will benefit from CS, and they might be more likely to be harmed with such therapy [14].

There is no sufficient evidence to support the beneficial role of antimalarials in the prevention of COVID-19. Gao et al, reported that the administration of chloroquine phosphate in 100 Chinese patients with COVID-19 infection was superior, compared with the control group [15]. In contrast to the above report, a small pilot study from China showed no difference between HCQ-treated patients compared with a control group [16]. Anecdotal reports from registries of patients with COVID-19 infection and autoimmune rheumatic diseases demonstrated that approximately 25% of infected patients were already taking HCQ, indicating HCQ might not have any protective effect. So there is urgent need for large clinical trials to assess the efficacy and safety of antimalarial treatments in patients with COVID-19 infection [17].

There is no data to suggest, and experts do not believe that splenectomized patients are more vulnerable to COVID-19. However, viral infections can be complicated by bacterial superinfections. Therefore, in the setting of fever, the management of a splenectomized patient should be the same as without COVID-19 infection through intravenous antibiotics, and close monitoring because severe sepsis can develop quickly [8].

As long-term natural immunity to SARS-CoV-2 is uncertain; As the required percentage of immune individuals has been estimated to be 50–66% of the population that will take long to be achieved, data from SARS-CoV suggest that the duration of immunity may not be sufficient significant, while the immunity response against SARS-CoV-2 may not be efficiently effective in all patients, as relapses have already been reported. Also, the development of mutant strains, which has already been documented, can cause the reemergence of the epidemic [18].

So herd immunity may depend on effective vaccination even more. Therefore, the development of an effective vaccine is of paramount importance especially for those patients with autoimmune disorders, not only for the control of the current outbreak, but also for the prevention of future outbreaks.

References
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