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RESIDENT'S FORUM

RF - Stevens-Johnson Syndrome and Toxic Epidermal Necrolysis: A Treatment Update[☆]



FR - Síndrome de Stevens-Johnson y necrólisis epidérmica tóxica. Actualización en el manejo terapéutico

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PALABRAS CLAVE

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Ciclosporina;
Etanercept

Stevens-Johnson syndrome (SJS) and toxic epidermal necrolysis (TEN) are potentially serious diseases, usually caused by a drug reaction, with a mortality rate of approximately 25%. Optimum treatment is not well established. The only randomized clinical trial, which took several years to complete, showed that thalidomide was not only ineffective but was associated with higher mortality.¹

Since then, cases have been published of treatment with cyclophosphamide, systemic corticosteroids, plasmapheresis, cyclosporin, intravenous immunoglobulins (IVIGs), and tumor necrosis factor inhibitors (anti-TNF agents). In this summary, we wish to comment on the findings of 2 recently published studies on the management of this disease.

In 1998, IVIG was reported as an effective therapy in patients with SJS/TEN for the first time.² Since then, the studies published show conflicting results, and it has not been possible to demonstrate a significant improvement in survival among patients treated with IVIG compared to those who receive only supportive care.³ However, a study has shown decreased mortality when IVIG is used at high doses (≥ 2 g/kg).⁴

Kirchhof et al.⁵ conducted a retrospective study and analyzed the outcomes of treatment with IVIG 1 g/kg/d for 3 days or cyclosporin 3-5 mg/kg/d for up to a maximum of 7 days. Of the 64 patients with clinical and histologic evidence of SJS/TEN included in the study, 12 received conservative treatment, 35 received IVIG, 15 received cyclosporin, and 2 received both IVIG and cyclosporin. Based on the estimated mortality according to the SCORE of Toxic Epidermal Necrosis (SCORTEN) scale, the authors concluded that those patients who received IVIG had a higher than expected mortality. In contrast, those who received cyclosporin had a lower mortality.

A study conducted by Paradisi et al.⁶ included 10 patients whose diagnosis of TEN was based on the presence of blisters and erosions on more than 30% of the body surface area. In addition to the corresponding support measures, patients received a single etanercept dose

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(50 mg subcutaneously) within 6 h of hospital admission. All patients responded satisfactorily to treatment and attained complete reepithelization within a mean of 8.5 days (range, 7-20 d). No patient experienced adverse effects attributed to etanercept. To date, the use of etanercept had only been reported in 2 patients with TEN. In addition, isolated cases had been reported of infliximab administration with good response. This is the largest study published to date and demonstrates the beneficial role of anti-TNF agents in patients with TEN. Although this study lacks a control arm, it nevertheless seems reasonable to consider etanercept as a new therapeutic possibility.

Discontinuation of the causative drug and support measures, such as wound dressing, fluid administration, nutritional support, pain management, and prevention and treatment of infections remains the mainstay of treatment. Patients with extensive involvement and/or SCORTEN scores ≥ 2 should be referred to large burns units.

There is certain controversy regarding management of SJS/TEN, particularly with regards the use of systemic corticosteroids and IVIG. However, these recent studies seem to shed some light on the best approach as they support the use of cyclosporin and open up the possibility of using anti-TNF agents as a promising therapy in what is an extremely serious disease.

Ethical Responsibilities

Protection of human and animal subjects. The authors declare that the procedures followed are in line with the corresponding ethics committee and the Helsinki Declaration of the World Medical Association.

Confidentiality of data. The authors declare that they have followed their hospital's protocol on the publication of data concerning patients.

Right to privacy and informed consent. The authors obtained the informed consent of patients and/or subjects mentioned in this article. The informed consent form is located in the archives of the corresponding author.

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