Toxic Epidermal Necrolysis and Stevens Johnson Syndrome: Our Current Understanding

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Stevens-Johnson syndrome (SJS) and toxic epidermal necrolysis (TEN, Lyell's syndrome) are now considered to be distinct clinical entities within a spectrum of adverse cutaneous drug reactions of increasing severity based on their surface of skin detachment. Within this spectrum, SJS which can be considered as a minor form of TEN is characterized by less than 10% body surface area of skin detachment, and an average reported mortality of 1—5%, whereas TEN is characterized by more than 30% skin detachment, and an average reported mortality 25—35%.

Both SJS and TEN are characterized morphologically by the rapid onset of keratinocyte cell death by apoptosis, a process that results in the separation of the epidermis from the dermis. Recent evidence is supportive of a role for inflammatory cytokines and the death receptor Fas and its ligand FasL in the pathogenesis of keratinocyte apoptosis during TEN. This Fas-mediated keratinocyte apoptosis that is the last step culminating in epidermal detachment in TEN can be inhibited in vitro by antagonistic monoclonal antibodies to Fas, and by intravenous immunoglobulins (IVIG) which have been shown to contain natural anti-Fas antibodies.

Consequently, over the last few years, numerous case reports and 9 non-controlled clinical studies containing 10 or more patients have analyzed the therapeutic effect of IVIG in TEN. Taken together, although each study has its potential biases, 7 of 9 such studies point towards a benefit of IVIG used at doses greater than 2 g/kg on the mortality associated with TEN. These studies should serve as the basis for designing an appropriate prospective trial or for conducting a metaanalysis in the near future, in order to determine the therapeutic efficacy of IVIG in TEN.

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