Editorial

Treatment of Churg-Strauss syndrome: options for the future

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Treatment of Churg-Strauss syndrome (CSS) is quickly effective and patients respond immediately to steroids: a decrease in eosinophil count is observed and respiratory clinical symptoms improve. In the most severe forms, an immunosuppressant is needed to control disease activity (1, 2). The initial response rate exceeds 80% and the 10-year survival rate 75%.

However, treatment is not so effective in the long term and at least one third of patients relapse. Asthma and eosinophilia remain present in the majority of patients and are controlled by a permanent steroid treatment. Therapeutic difficulties reflect our poor understanding of disease pathogenesis. All studies underline the heterogeneity of pathogenic mechanisms in CSS. Three main components of CSS pathophysiology have been highlighted by previous studies: TH2 lymphocytes, as the main known actors in asthma and allergic manifestations; eosinophils, with their proteins' cellular cytotoxicity; and anti-neutrophil cytoplasmic antibodies (ANCA). The respective roles of each of those players may vary during the course of the disease and in each individual, and other more subtle and/or yet unidentified immunological abnormalities are certainly involved.

Several new drugs targeting cytokines or other mechanisms are now available to treat patients but an appropriate prescription should ideally be based on identified mechanisms, which need to be investigated and analysed, may be case by case and according to disease stage. Flares of Churg-Strauss syndrome have been described when patients have been treated with omalizumab or leukotriene-receptor antagonists. The mechanism of flares is controversial but complicates treatment choice. Rituximab was also reported to be beneficial in a few CSS patients who were mainly ANCA-positive and who had short-term improvement in their clinical features and eosinophil counts. Conversely, rituximab was reported to be ineffective and even incriminated as having provoked immediate and severe bronchospasm in rare ANCA-negative patients. Mepolizumab, a monoclonal antibody against interleukin-5, a cytokine involved in eosinophil maturation, proliferation, and survival, has been shown effective in Churg-Strauss patients, with not only a sustained reduction in eosinophil levels but also a significant corticosteroid-sparing effect (3).

Time for new prospective studies on Churg-Strauss syndrome has come. New drugs have to be evaluated *versus* immunosuppressants. Steroids will probably be necessary for most patients but new therapeutic schemes focusing on steroids tapering have to be established. Combinations of drugs, steroids, cytotoxic agents and biotherapies could also be evaluated concentrating on their benefit/risk ratio.

The future is promising but has to be coordinated and based on a common objective of clinicians and basic researchers to treat patients and to cure the disease.

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