

Treatment and outcomes in SPANISH PATIENTS WITH IgG4-related disease

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Objective:

To describe the treatments used in a series of patients diagnosed with IgG4-Related Disease (IgG4-RD) in Spain and to review the outcomes.

Method:

Clinical data were obtained from the Spanish Registry of IgG4-RD (REERIGG4) from October 2013 to January 2016, including 14 centers. Outcomes were assessed by a self-made response scale and the IgG4 responder index (RI). We categorized the outcomes as a total response (disappearance of the pseudotumoral lesions and absence of symptoms), partial response (<50% regression of the tumefactive lesions or persistence of inflammation without symptoms) and no response if no changes were noticed. Treatment failure was considered if an increase of the activity, mass size or reappearance of symptoms were noticed among patients under treatment.

Results:

Sixty-eight patients were included. Twenty-six (38%) were females, mean age 53.4 years. Thirty-six patients (52.9%) had systemic IgG4-RD involving >1 tissue. The most commonly involved tissues were: retroperitoneum (33%), orbital pseudotumor (28%), and maxillary and paranasal sinuses (24%). The main treatments used were: steroids (90%), surgery (45%) and azathioprine (19%).

All treatments were successful in achieving complete or partial response. The mean pre and post-treatment RI values were 6.7 (SD 4.6) and 1.9 (SD2.6) respectively. There were no differences between systemic and non-systemic disease regarding the chosen treatments and the outcomes. The combination azathioprine-steroids was used in 12 patients. Fourteen percent of them relapsed (considering relapse as an increase of the inflammation, mass size or reappearance of symptoms, since the first month after the treatment withdrawal). The treatment failed in 28.6% of them. The combination steroids-rituximab was indicated in 6 patients, showing no relapses and 1 treatment failure. The majority of patients treated with azathioprine or rituximab combined with steroids had a systemic disease (6.6 and 80%, respectively). Nearby all of them had already failed other previous treatments.

Conclusions:

In our series, IgG4-RD has been treated with a myriad of drugs and procedures. The outcomes have been acceptable but the disease tended to relapse (21%) and the treatment failures were common (27%), probably due to the lack of well-defined treatment schemes supported by solid studies. Steroids were still the cornerstone of the treatment. Rituximab results were promising in our study but the number of patients was limited. Azathioprine, in combination with steroids, may be an accessible alternative treatment for IgG4-RD that should be explored. The RI correlated with the treatment outcomes and will have an important role monitoring future studies on IgG4-RD therapies.