
Conclusion: Disease severity scores should be used for treatment choice, evaluation of disease prognosis and classify patients according to disease severity in clinical trials. In the current study, we found a difference between the classifications of disease severity, which could be a result of each scoring system uses different parameters of the disease.

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CLINICAL PHENOTYPES OF IGG4-RELATED DISEASE IN SPAIN

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Background: Recently, several clinical phenotypes in IgG4-related disease have been described in a multinational and ethnically variant cohort1.

Objectives: To assess the clinical presentation of IgG4-related disease (IgG4-RD) in Spanish patients and assess the distribution among different clinical phenotypes.

Methods: Clinical data were obtained from the Spanish IgG4-RD registry (REERIGG4) from October 2013 to December 2018, including 9 centers. We reviewed demographic data and organ involvement. The assignment of clinical phenotypes was done by 2 experts, based on organ involvement and clinical manifestations, following Wallace et al.1 subsets. The phenotypes were: pancreato-hepato-biliary (HBP), retroperitoneum and aorta (RA), head and neck limited (HNL) and Mikulicz and systemic (MS). A fifth group designated not defined (ND), included the patients that did not fit in the previous phenotypes.

Results: One-hundred patients were included. Thirty-four (34%) were females, median age at diagnosis was 54.8 years (IQR 20.7). The ethnicity of the participants was: Caucasian 83%, Hispanic 12% and North-American/Middle-East 5%. Ninety-two percent were diagnosed with a biopsy. Regarding the diagnostic criteria, 85% met consensus pathology criteria and 94% comprehensive criteria.

Fifty-one patients (51%) had systemic IgG4-RD involving >1 tissue. The most commonly involved tissues were: retroperitoneum (35%), lymph nodes (19%), orbit pseudotumor (18%), salivary glands (16%) and pancreas (14%). Forty-two patients (42%) had elevated serum IgG4.

The representation of each clinical phenotype was: HBP 14%, RA 25%, HNL 26%, MS 20%, ND 15%. Patients were equally distributed from the perspective of ethnicity. Men were predominant in all groups (71, 84, 70, 73%) except in HNL (61% for women). Systemic disease was present in all the MS patients, but in 1/3 of the other groups. Serum IgG4 was elevated in 86% of the cases in HBP, 28% in RA, 19% in HNL, 60% in MS and 40% in ND.

Conclusion: The Spanish IgG4-RD population was mainly ethically Caucasian. Few patients had serum IgG4 elevation. The most frequent phenotype was HNL, followed by RA. The HBP phenotype was less frequent than in previous reports. The influence of race could modify the clinical expression of IgG4-RD. Knowing the regional phenotypes of IgG4-RD may help clinicians improve disease management.

REFERENCES:

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