J Clin Cell Immunol 2017, 8:3(Suppl)
DOI: 10.4172/2155-9899-C1-037

Case series: Association between elevated circulating plasma blasts and their gated flocytometric picture in a cohort of diversity of clinical presentations of IGG4-RD cases

H A El-Saadany
Kobry-El Qobba Medical Complex, Egypt

Background: Immunoglobulin G4-related disease (IgG4-RD), is an immune-mediated disorder with certain clinical, serological, and histopathological features. The affected tissues and organs show sclerosis with dense lymphoplasmacytic infiltrate rich in IgG4-positive plasma cells usually associated with high level of serum IgG4 and elevated circulating plasmablasts.

Objectives: To report a retrospective analysis of case series of patients diagnosed as IgG4-RD who were seen in Kobri El-Kobba military complex, Cairo, Egypt since January 2015 till June 2016. Reporting includes the relationship between elevated circulating plasmablasts with their gated flow cytometric picture and the presented diversity of clinical presentation of patients

Methods: 22 patients of IgG4-RD with different clinical presentations are included. The diagnosis was made based on the clinical manifestations, detecting elevated circulating plasmablasts, imaging studies, flow cytometry by gating with CD138, CD38, CD19LOW, CD20-, and CD27, and the appropriate tissue biopsy characteristic to the diagnosis of the disease.

Results: The presented manifestations were as follows: biliary diseases (4) 18.1%, orbital diseases (6) 20.7%, interstitial lung disease (4) 18.1%, thyroid disease (4) 18.1% and salivary gland disease (4) 18.1%. Elevated circulating plasmablasts were found in all cases (100%) irrespective of their count. All patients had imaging studies related to diagnosis of the disease corresponding to the affected organ. All patients had immunophenotyping on peripheral blood by flow cytometry with gating to CD138, CD38, CD19LOW, CD20 -, and CD27 all of them are indicative to the disease. All the results were same for different clinical presentations of the IgG4-RD cases included.

Conclusions: Our retrospective case study provides data on a variety of clinical presentation of a cohort of cases of IgG4-RD. All the presented cases with their diversity show common results in having a high level of circulating plasmablasts and a picture of flow cytometry consistent with the disease especially positive CD138, CD38, Dim CD20 which support the previous results of the criteria of the diagnosis of the disease. In addition, combination of elevated circulating plasmablasts and the above results of flow cytometry are present in all varieties of clinical presentations.

hanyelsadany@yahoo.com