Autoimmunity in moroccan patients with common variable immune deficiency

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Background

Common variable immunodeficiency (CVID) is the most frequent symptomatic primary immune deficiency in adults. It is characterized by various degrees of hypogammaglobulinemia. Similar to many immunodeficiency diseases, autoimmunity is a common feature along with infectious complications. Autoimmunity and auto inflammation can present as an autoimmune cytopenias, a sarcoidosis-like disorder or a rheumatological like disease.. CVID can be associated with a wide range of autoimmune diseases.

Purpose:

to identify autoimmune features and their frequencies in the moroccan registry of common variable immune deficiency (CVID).

Methods:

It is a cross-sectional study that was conducted in July 2021 in Morocco. We have used the moroccan registry of CVID to collect demographic, clinical and biological data of the patients. We have compared between two groups, one group with autoimmune manifestations and the other group was free from any autoimmune or inflammatory involvement. For data analysis, we have used SPSS 25 software, P value was considered significant when p<0.05. The patients consent was obtained.

Results:

26 patients were diagnosed with CVID. Mean age of diagnosis was at 25,38 ± 15,4, and a mean age of onset was at 18,8 ± 15,3. Sex ratio was 0,73. Consanguinity was present in 46,5% of the cases. The revealing sign was sinopulmonary involvement in 61,5% of the patients, followed by diarrhea (7,7%) and autoimmune manifestations (7,7%). During the disease course, half of our patients presented with autoimmune features, lymphoproliferation was a complication in 42,3% of the cases. The main autoimmune features were: immune thrombocytopenic purpura, autoimmune hemolytic anemia, vitiligo, granulomatosis, arthritis, and lupus. Two patients presented with coeliac-like disease, three other patients had a Crohn-like disease, and four more patients were diagnosed as having sarcoidosis-like manifestations. Autoimmune complications were prevalent in women without a statistic significance.

Patients with no consanguinity were having more autoimmunity, p value was non significant. Lymphoprolifrative disease was associated with autoimmunity (p=0,005). Arthritis was prevalent in patients with autoimmunity (p=0,01). Immunoglobuline therapy was used in 92,3% of patients, corticosteroids were added in 42,3% of cases along with immunosuppressors in 11,5%. Disease course was good in 69,2%.

Conclusion:

Autoimmunity is frequent in patient with CVID, especially autoimmune cytopenia. It should be searched for because the treatment depends on it, to enhance life quality of patients and to avoid severe and lethal complications.