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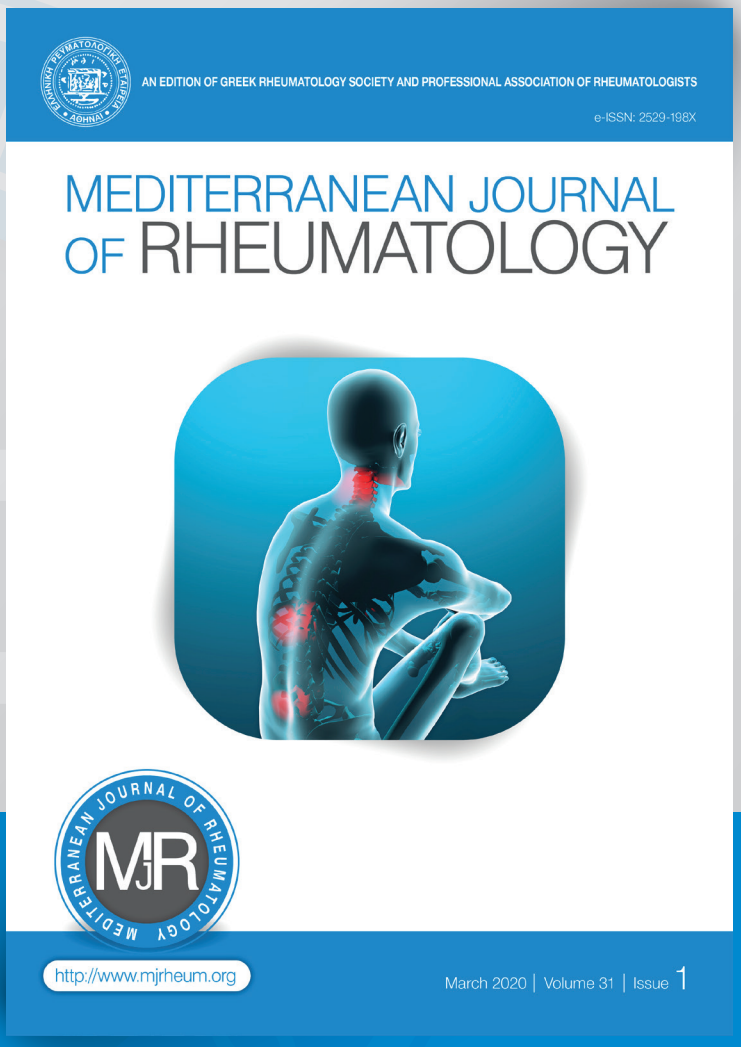

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LETTER TO THE EDITOR

Overlapping between IgG4-RD and Behçet's Disease

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To the editor:

Alazani MB et al.[1] reported for the first time a case of IgG4-Related disease (IgG4-RD) retroperitoneal mass in a patient with Behçet's Disease (BD), and they found only one other similar patient with BD, and in that case, an IgG4-RD laryngeal mass.[2] Similarly, we have previously described another patient with recurrent aphthous stomatitis and IgG4-related laryngitis with the suspected diagnosis of overlapping with BD.[3]

Some conditions that were previously thought to be unique clinical syndrome are now recognized as a clinical manifestation of IgG4-RD. IgG4-RD physiopathological basis is the presentation of autoantigens by plasmablasts or B cells to CD4 cytotoxic T cells, which produce pro-fibrotic cytokines such as IFN-γ, IL1-ß and TGF- ß.[4] Although BD shares some common features with autoimmune and autoinflammatory diseases, is ultimately caused by disturbance of T-cell homeostasis, especially Th1 and Th17 expansion, as well as Tregs response suppression. Neutrophil activity is increased at the earliest stage of inflammation in the affected organs, as well as the presence of HLA-B*5 and increased IL-17, which appears to play a major role in neutrophil activity.[5] IgG4-RD is a rare, multiorgan condition characterized by histologic features, and accurate clinical diagnosis can be challenging due to the atypical manifestations or the presence of overlapping autoimmune diseases like BD or antineutrophil cytoplasmic antibody-associated vasculitides.[6]

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CONFLICT OF INTEREST

The authors declare no conflict of interest.

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