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IgG4-related disease
Bolest povezana s IgG4

Emanuel Della Torre [1]

[1]*San Raffaele Hospital, Milan, Italy*

IgG4 related disease (IgG4-RD) is an immune mediated condition presenting with mass forming lesions that lead to permanent organ injury and death if left untreated. Abundant IgG4 positive plasma cells in affected tissues and fibrosis represent hallmark pathological features of this disorder. IgG4-RD was first described in 2003, when conditions regarded as unrelated entities for decades – such as type I autoimmune pancreatitis (AIP), sclerosing cholangitis, retroperitoneal fibrosis, hypertrophic pachymeningitis, Mikulicz's disease, and Riedel's thyroiditis – were shown to occur simultaneously in a proportion of patients and to share common histological findings. Since then, IgG4-RD has been recognized with
increasing frequency by both generalists and specialists worldwide. However, awareness of the diagnostic and therapeutic tools for the management of patients with IgG4-RD remains confined to tertiary care centers, and the disease is still misdiagnosed as neoplastic, inflammatory, and infectious conditions. In the past decade, international collaborations
have produced consensus documents for the pathological diagnosis and treatment of IgG4-RD, providing clinicians with useful guidelines for improving the management of these patients. The 2019 American College of Rheumatology (ACR) and European League Against Rheumatism (EULAR) classification criteria for IgG4-RD have been released, and disease phenotypes have been identified, paving the way for a new era of personalized therapeutic approaches. IgG4 related disease is highly treatable and responds promptly to glucocorticoids, but it can lead to end stage organ failure and even death if unrecognized. Prolonged courses of corticosteroids are often needed to maintain remission because the disease relapses in most patients. Rapid advancement in our understanding of the pathophysiology of IgG4 related disease is leading to the identification of novel therapeutic targets and possible personalized approaches to treatment.
In this lecture, the latest advances in diagnosis and management of IgG4-RD, will be outlined touching on diagnostic and therapeutic guidelines for IgG4-RD, established and novel potential disease biomarkers, and emerging treatment options based on the most recent acquisitions in disease pathophysiology.

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E-mail adresa prezentirajućeg autora: dellatorre.emanuel@hsr.it
Izjava o sukobu interesa: nema sukoba interesa