IgG4-related orbital disease (ROD) - a clinicopathologic study of 22 patients

L. Holbach, A. Bergua, C. Rummelt, J. Weller, F. E. Kruse
Department of Ophthalmology and Eye Hospital, University Erlangen-Nürnberg, Erlangen, Germany

Aim: To determine diagnostic criteria and treatment outcomes in 22 patients with biopsy-proven IgG4-ROD.
Patients and Methods: Retrospective analysis of 22 patients (18 females, 4 males; mean age at presentation 46 years, range 26-85 years) who underwent an incisional biopsy and surgical debulking. 16 lesions were unilateral, six bilateral. The mean duration of disease was 6.7 years (range 2-18). One organ was involved in 9 patients, more than one in 13 patients. The mean follow-up was 6.7 years (range 0.4-14).
Results: Common clinical symptoms and/or signs included eyelid swelling, proptosis, downward displacement of the globe, blepharoptosis, yellowish eyelid infiltrates, palpable mass and/or clinical evidence of preseptal inflammation. Prominent histopathologic features were lymphoplasmacytic infiltrates, sclerosing inflammation, lymphoid hyperplasia and xanthogranulomatous inflammation. The mean IgG4: IgG ratio in the biopsy specimens was 0.8 (range 0.4-1.0), the mean IgG4 serum concentration 1003 mg/dl (range 13-6620). Systemic steroids showed a prompt and efficient response in all patients, recurrences after tapering in 7 patients. Methotrexate, azathioprine and mycophenolate were associated with recurrences and incomplete remission. Rituximab and steroids showed in 10 patients sustained improvement and few side effects.
Conclusions: An early diagnosis of IgG4 - ROD based on careful clinicopathologic correlation may avoid severe organ damage and disease durations over many years. Rituximab appears to be efficient and safe in patients who are steroid dependent or resistant.