**270. Neurological Manifestations of IgG4 Related Disease: A Descriptive Study**

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**Background:** Neurological manifestations of IgG4-related disease (IgG4-RD) is rare compared to other involvements. It may be in four different forms: pachymeningitis, orbital disease, hypophysitis and parenchymal disease. This study is aimed to describe clinical features, diagnostic parameters, and therapeutic management of patients with a neurologic spectrum.

**Methods:** Patients diagnosed with IgG4-RD from January 2005 – September 2021 were included in the study. Demographic data, clinical features, involved organs, disease course, laboratory parameters, and therapeutic responses were analysed.

**Results:** Among 99 IgG4-RD patients, 76 patients with regular follow-up visits were included in the study. The female to male ratio was 1:1, and the mean diagnostic age (years) was 47.38 ± 14.9 SD. Median follow-up period was 27.5 months and median serum IgG4 level was 184.75 mg/dL (min 14 - max 765 mg/dL). Neuro-IgG4RD was present in 16 (21%) of the patients (10 had orbital disease, 5 had pachymeningitis, one had hypothalamohypophyseal axis involvement). In the neuro-IgG4RD group, the female to male ratio was 1.3:1 and the mean diagnostic age was 47.5 ± 17.7 SD. Neurological involvement is the presenting symptom in 77% of the patients (n: 13), while in the remaining 23%, it was additive to another organ involvement (n: 3). Median serum IgG4 level was 128.43 mg/dL (min 14 – max 349 mg/dL) in this group. 60% of the patients with the orbital disease were male and the most frequent presenting symptoms were headache, loss of vision, and proptosis. The most frequent radiological findings of orbital disease were thickening of extraocular muscles, staining in the retrobulbar area and compression or inflammation around optic nerve. 80% of the patients with pachymeningitis were male, and the most frequent presenting symptoms were headache and accompanying cranial nerve involvement. In this group, skull base was most affected and the most frequent site of involvement was tentorium cerebelli. Main choice of treatment were corticosteroids in neuro-IgG4RD; but in 43.75% of the patients, rituximab was needed to be added to the treatment (n: 7). In all patients receiving rituximab, neuroradiological findings were either stabilized or regressed.

**Conclusions:** Neurological involvement was observed in one out of every five patients, either at presentation or during follow-up. Orbital disease and pachymeningitis are the most frequent neurological manifestations in this cohort. Therapeutic options include corticosteroids and conventional immunosuppressive drugs. Rituximab might be an effective treatment option in resistant cases.

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