

Clinical aspects of IgG4-related optic neuropathy

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Objective:

The triad symptoms of IgG4-related ophthalmic disease (IgG4-ROD) are enlargement of lacrimal glands, trigeminal nerves and extraocular muscles. On the other hand, the most severe symptom in IgG4-ROD is optic neuropathy presenting visual loss. We term such cases IgG4-related optic neuropathy and reviewed our experiences.

Methods:

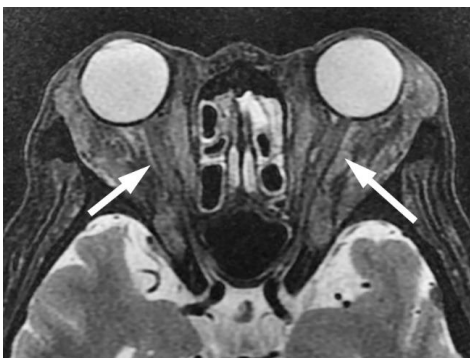
Among 41 consecutive cases (21 in men and 20 in women) with IgG4-ROD pathologically diagnosed from November, 2004 through December, 2015 in Kanazawa University hospital, six patients presented mass lesions compressing the optic nerve in magnetic resonance images, and four of them showed symptoms of optic neuropathy. Clinical aspects and steroid therapies of these four cases were reviewed.

Results:

The four cases with IgG4-related optic neuropathy were three males aged 52, 67 and 71 years and one 87 year-old female at diagnosis. The serum IgG4 levels before treatment ranged from 717 to 2340 mg/dL (normal level less than 135 mg/dL), whose median value of 1520 mg/dL was significantly higher ($p = 0.011$ by a Mann-Whitney U-test) than that of 534 mg/dL (range 22 - 1750 mg/dL) in the group of 37 cases without optic neuropathy. Swelling of the lacrimal glands and extraocular muscles were accompanied in every four cases, and the trigeminal (infraorbital and supraorbital) nerves enlargement were present in three of them. All the patients showed deteriorated visual acuities (no light perception in the worst case) and/or visual field defects consistent with optic neuropathy. Two patients were initially diagnosed and treated as having glaucoma. These four patients underwent systemic steroid therapy, and overall, their visual functions responded to the steroid therapy at least in part, but the recoveries were limited.

Conclusions:

IgG4-related optic neuropathy might be misdiagnosed as glaucoma. Vigilance is necessary to avoid visual loss because of optic neuropathy in IgG4-related disease, especially in patients with high serum IgG4 levels.



MRI showed lesions around the optic nerves (arrows) in a patient with IgG4-related ophthalmic disease