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【 PICTURES IN CLINICAL MEDICINE 】

IgG4-related Ophthalmic Disease with Proptosis

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Key words: IgG4-related ophthalmic disease, proptosis, lymphadenopathy, hypertrophic pachymeningitis, ophthalmic disease

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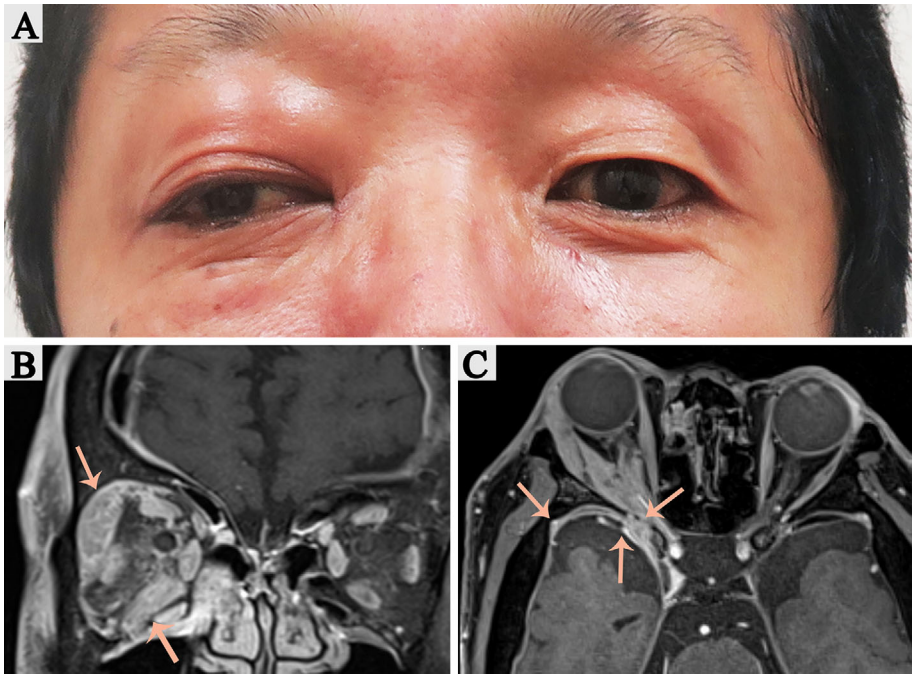

Picture.

A 49-year-old man presented to the general medicine outpatient clinic with a 3-month history of right eye proptosis. He had a 12-month history of right cervical lymphadenopathy, which was reactive according to the results of a biopsy performed 4 months previously. At presentation, he had an itchy eye without ocular pain, neuralgia, or diplopia. A physical examination revealed a swollen upper eyelid and proptosis (Picture A) and nodal masses in the cervical region. An ophthalmic examination revealed slightly limited eye movement. Tissue thickening was revealed on contrast-enhanced magnetic resonance imaging. Homogeneous continuity of enhancement was observed in the lacrimal gland and extraocular muscles (Picture B), and the enhancement spread to the pterygopalatine fossa, middle cranial fossa, and cranial dura mater (Picture C, arrow). His erythrocyte sedimentation rate was 19 mm/h and his C-reactive protein concentration was 0.21 mg/dL. His serum IgG4 level was elevated at 515 mg/dL (normal, <117 mg/dL). Immunostaining of the previous lymph node biopsy specimen revealed abundant IgG4-positive plasma cells. Based on these findings, the patient was diagnosed with IgG4-related ophthalmic disease (1). Treatment with oral glucocorticoids was initiated, after which the patient reported a significant improvement of proptosis and itchiness. Orbital imaging 4 months later showed recovery.

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Reference

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