



Case report

Immunoglobulin G4-related dacryoadenitis presenting as bilateral chorioretinal folds from severely enlarged lacrimal glands

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ABSTRACT

Purpose: To describe a case of immunoglobulin G4 (IgG4)-related dacryoadenitis presenting as bilateral chorioretinal folds from eyeball compression by massively enlarged lacrimal glands.

Observations: A 51-year-old woman with severely enlarged bilateral lacrimal glands was diagnosed as having IgG4-related dacryoadenitis. The glands strongly compressed the globes, forming chorioretinal folds resembling those found in orbital malignancy. Eventual treatment with oral prednisolone dramatically reduced the volume of the lacrimal glands and released globe compression on magnetic resonance imaging. However, the chorioretinal folds remained in the right fundus and symptoms of blurred vision improved but persisted.

Conclusions and importance: This is the first account of chorioretinal fold formation by severely enlarged lacrimal glands appearing in IgG4-related dacryoadenitis. Chorioretinal fold formation by an enlarged lacrimal gland occurring bilaterally may represent a basis for suspecting IgG4-related dacryoadenitis. Prompt treatment is recommended for patients presenting with very large lacrimal glands to avoid visual impairment.

1. Introduction

Since Hamano's first description of high serum immunoglobulin G4 (IgG4) in patients with sclerosing pancreatitis (autoimmune pancreatitis type 1) in 2001¹, there have been numerous reports of high serum IgG4 concentration and infiltration of IgG4-positive plasmacytes into multiple organs resulting in enlargement and sclerosis. Such manifestations are now collectively termed IgG4-related disease (IgG4-RD),^{2–4} the epidemiology of which remains poorly understood.^{3,5} IgG4-RD involves organs such as the pancreas, kidney, lung, prostate, and lymph nodes. The disease also afflicts the ocular adnexa and orbital tissues as IgG4-related ophthalmic disease (IgG4-ROD)^{2,4,6,7} in the lacrimal glands (IgG4-related dacryoadenitis),² extraocular muscles (IgG4-related orbital myositis),² and branches of the trigeminal nerve.^{8,9} The lacrimal glands are the most frequently involved ocular adnexa in IgG4-ROD,^{9–16} having previously been referred to as (IgG4-related) Mikulicz's disease with enlargement of the parotid glands.^{4,6,12} Abnormally large lacrimal glands often cause disorders of eye movement.^{13,14}

Orbital tumors and other diseases are known to compress the sclera of the globe, resulting in the formation of chorioretinal folds.^{15–22} To

our knowledge, severe compression of the globes in IgG4-related dacryoadenitis has not been reported. We herein describe a case of IgG4-related dacryoadenitis causing bilateral chorioretinal folds via globe compression by severely enlarged lacrimal glands that behaved more like an orbital malignancy to result in irreversible changes of the choroid and retina. This highlights the importance of frequent fundus examination in patients with IgG4-related dacryoadenitis, especially during conservative treatment.

2. Case report

A 51-year-old woman presented with blurred vision in her right eye and diplopia at upper gaze after feeling discomfort in the right superior eyelid for two years. Her best corrected visual acuity (BCVA) was 1.2 OD and 1.5 OS. Her bilateral upper eyelids were swollen and eye movement was bilaterally restricted at upper gaze. Hard masses were palpable under the bilateral temporal upper lids. Intraocular pressure (IOP) was 17 mmHg in both eyes. Chorioretinal folds involving the macula were detected in the right fundus by ophthalmoscopy and optical coherence tomography (OCT) (Fig. 1A). The left fundus was

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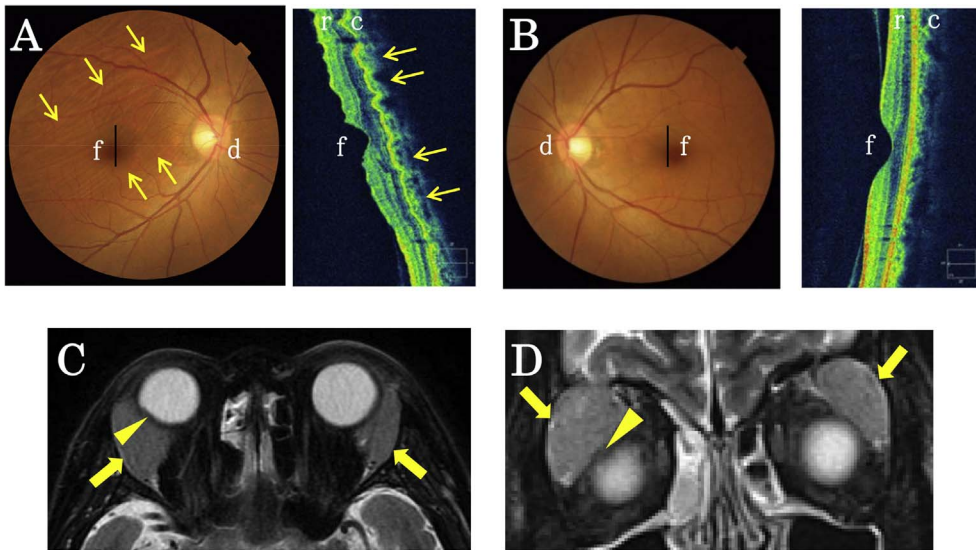


Fig. 1. Images of fundus and orbital magnetic resonance imaging (MRI) at the initial examination. A and B: Fundus photographs and cross sectional images of the central fundus obtained by optical coherence tomography (OCT). Chorioretinal folds are seen in the right fundus (A, arrows) but not in the left fundus (B). f, fovea centralis; d, optic disc; r, retina; c, choroid; black lines, lines scanned by OCT. C and D: T2 fat-suppressed axial (C) and coronal (D) images of orbital MRI. Enlarged bilateral lacrimal glands (arrows) are seen. The intensity of the lacrimal glands is slightly higher than that of the extraocular muscles. The right globe appears deformed by the enlarged lacrimal gland (arrowheads).

apparently normal (Fig. 1B). Goldmann perimetry testing of the right eye demonstrated decreased sensitivity in the lower field. Orbital magnetic resonance imaging (MRI) revealed enlargement of bilateral lacrimal glands, especially the right gland, without alterations in extraocular muscles or infraorbital nerves (Fig. 1C and D). Laboratory testing disclosed normal blood cell count and negative serum C-reactive protein but elevated serum IgG and IgG4 (1916 and 450 mg/dL, respectively). Serum interleukin-6 and angiotensin converting enzyme were normal. Thyroid hormones were also normal and thyroid stimulating hormone receptor antibody was negative. Anti-nuclear antibody, proteinase 3-anti-neutrophil cytoplasmic antibody, myeloperoxidase anti-neutrophil cytoplasmic antibody, and anti-SSA/SSB antibody were negative as well. Whole-body computed tomography (CT) revealed enlargement of bilateral lacrimal and submandibular glands and segmental enlargement of the pancreatic tail. Images of 2-[¹⁸F]-fluoro-2-deoxy-D-glucose positron-emission tomography/CT displayed increased uptake in bilateral lacrimal and submandibular glands.

Pathological study of a biopsy specimen obtained from the right lacrimal gland disclosed marked infiltration of lymphocytes and

plasmacytes along with mild fibrosis (Fig. 2A and B). Obliterative phlebitis and storiform-type fibrosis were absent (Fig. 2A and B). No atypical lymphocytes or non-necrotizing epithelioid granulomas were apparent (Fig. 2B). Immunohistological analysis revealed abundant infiltrating IgG- and IgG4-positive plasmacytes (Fig. 2C and D). The number of IgG4-positive cells was 80/high-power field (HPF), with an IgG4-/IgG-positive cells ratio of 55%. Analysis by the polymerase chain reaction of paraffin-embedded biopsy sections disclosed no evidence of monoclonal immunoglobulin gene rearrangement.

The patient satisfied the criteria for definitive IgG4-ROD and was diagnosed as having IgG4-related dacryoadenitis (Table 1).⁶ Other diseases were ruled out by systemic examination, blood testing, pathological study, and analysis of clonality (Table 1).⁶ The patient declined treatment of oral prednisolone (PSL) due to the mildness of her symptoms and concern about side effects.

Five months after the initial examination (two months after refusing treatment), the patient returned with deteriorated blurred right eye vision. Her right BCVA was unchanged, but IOP had increased to 23 mmHg in both eyes. Hess charting revealed the restriction of not

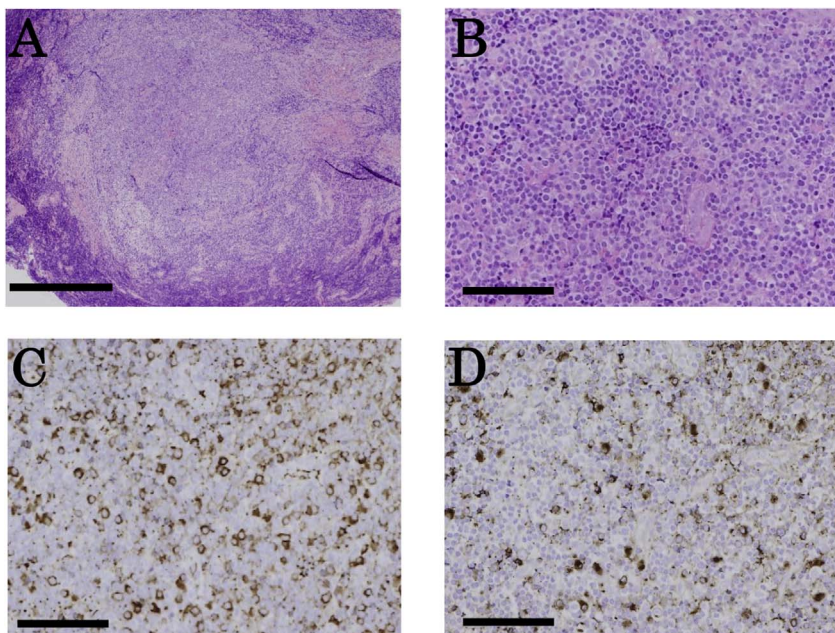


Fig. 2. Pathological studies. A and B: Lymphoplasmacytic infiltration is evident in the right lacrimal gland (hematoxylin and eosin; A, low magnification, scale bar: 500 μ m; B, high magnification, scale bar: 100 μ m). Storiform fibrosis is not apparent. C: immunoglobulin G (IgG)-positive plasmacytes (immunoperoxidase, scale bar: 100 μ m). D: IgG4-positive plasma cells (immunoperoxidase, scale bar: 100 μ m). The ratio of IgG4/IgG-positive plasma cells is 55%.

Table 1
Diagnostic criteria for IgG4-ROD (2015)⁶ (Reproduced with permission from Japanese Journal of Ophthalmology).

(1) Imaging studies show enlargement of the lacrimal gland, trigeminal nerve, or extraocular muscle as well as masses, enlargement, or hypertrophic lesions in various ophthalmic tissues.
(2) Histopathologic examination shows marked lymphocyte and plasmacyte infiltration, and sometimes fibrosis. A germinal center is frequently observed. IgG4+ plasmacytes are found and satisfy the following criteria: ratio of IgG4+ cells to IgG+ cells of 40% or above, or more than 50 IgG4+ cells per high-power field (×400).
(3) Blood test shows elevated serum IgG4 (≥135 mg/dl).
Differential diagnosis for IgG4-ROD ⁶ (Reproduced with permission from Japanese Journal of Ophthalmology)
Sjögren's syndrome
Lymphoma
Sarcoidosis
Granulomatosis with polyangiitis (Wegener's granulomatosis)
Thyroid-related orbitopathy
Idiopathic orbital inflammation
Dacryoadenitis or orbital cellulitis caused by bacteria or fungi

Diagnosis is classified as definitive when (1), (2), and (3) are satisfied, probable when (1) and (2) are satisfied, and possible when (1) and (3) are satisfied. As MALT lymphoma may also contain IgG4+ cells, careful differentiation is necessary.

only supraduction, but also of infraduction and abduction, in the right eye (Fig. 3). The chorioretinal folds had become more apparent in the right fundus and had also appeared in the left fundus (Fig. 4A and B). Oral PSL of 0.6 mg/kg/day (40 mg/day) was commenced with her consent and an improvement in bilateral lid swelling was noted after one week. PSL was tapered every two weeks, during which time IOP normalized. Her restricted eye movement improved and diplopia disappeared. Her serum IgG4 level had decreased to 132 mg/dL. The chorioretinal folds in the left fundus had disappeared by two months of PSL but were detectable in the right fundus even at three months after the commencement of therapy (Fig. 4C and D). Orbital MRI performed three months after starting treatment revealed decreased bilateral lacrimal gland volume (Fig. 4E and F). Ten months later, the patient displayed no signs of relapse on repeated orbital MRI with PSL maintenance doses of 10 mg/day. However, the chorioretinal folds in her right fundus persisted. Ultimately, BCVA was increased to 1.5 OD and her complaint of blurred vision in the right eye became slightly improved. The chorioretinal folds in her right fundus remained without relapse on MRI (Fig. 5A and B) and serum IgG4-level was 238 mg/dL after two years of follow-up with PSL maintenance therapy of 5 mg/day.

3. Discussion

We encountered a rare case of strong compression of the globes by severely enlarged lacrimal glands in IgG4-related dacryoadenitis that caused bilateral chorioretinal folds. Painless swelling of bilateral lacrimal and parotid glands along with infiltration of IgG4-positive plasmacytes and high serum concentration of IgG4 was initially termed IgG4-related Mikulicz's disease.^{4,5,12,23} Afterwards, specific recommendations for IgG4-related organ system nomenclature were introduced.² IgG4-ROD was adopted as the general term for periocular manifestations of the disease,^{2,4} and the classification of IgG4-related dacryoadenitis was subsequently proposed for lacrimal lesions instead of IgG4-related Mikulicz's disease.²

The causes of chorioretinal folds vary but can be classified into two main patterns. One is a result of such ocular disorders as age-related macular degeneration,^{19,21,22} scleritis,^{19,21,22} uveitis,^{19,21,22} hypotony,^{19,21,22} and uveal effusion.^{19,21} The other is caused by compression of the globes by thyroid ophthalmopathy^{19,21,22} or by orbital tumors such as fibrous dysplasia,¹⁶ osteosarcoma,¹⁷ maxillary sinus sarcoma,¹⁷ cavernous hemangioma,¹⁷ and metastatic cancer.¹⁶ Lacrimal benign mixed tumors were also reported to produce chorioretinal folds by globe compression.²⁰ To our knowledge, however, severe lacrimal gland compression of the globes to the extent of forming chorioretinal folds has not been reported in IgG4-related dacryoadenitis. In the present case, chorioretinal folds were present in the right fundus at the initial examination and later appeared in the left fundus despite the absence of malignancy (Fig. 1A, B, 4A and B). IgG4-RD causes enlargement in various organs, which often exhibit accompanying sclerosis. The nomenclature of several IgG4-RDs derive from the sclerosis appearing in the lesions, for instance, sclerosing mesenteritis,² IgG4-related sclerosing cholangitis,² and IgG4-related chronic sclerosing dacryoadenitis.²⁴ It was therefore possible that not only enlargement, but also sclerotic change, of the lacrimal glands caused the compression and even deformity of the globes in the present case of IgG4-related dacryoadenitis. As chorioretinal fold formation is unilateral in many cases of orbital and lacrimal tumors,^{6–8,10,12} bilateral chorioretinal fold formation by lacrimal glands may be a clue in diagnosing IgG4-related dacryoadenitis.

The characteristic pathological findings of general IgG4-RD are marked infiltration of polyclonal lymphocytes and plasmacytes, especially IgG4-positive plasmacytes, storiform-type fibrosis, and obliterative phlebitis.^{3,5} According to the pathological items in the comprehensive diagnostic criteria for IgG4-RD 2011 released by Umehara et al., remarkable lymphocyte and plasmacyte infiltration, fibrosis, and infiltration of IgG4-positive plasma cells are required for diagnosis.⁴ However, some findings are not apparent in certain organs; in the

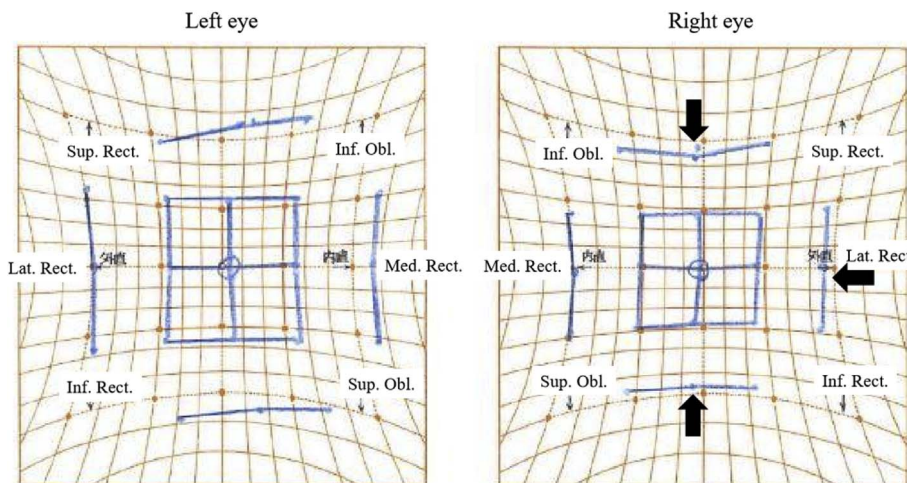


Fig. 3. Hess chart one day before starting prednisolone treatment. Supraduction, abduction, and infraduction of the right eye were slightly restricted (arrows). Sup. Rect; superior rectus muscle, Inf. Obl; inferior oblique muscle, Lat. Rect; lateral rectus muscle, Med. Rect; medial rectus muscle, Inf. Rect; inferior rectus muscle, Sup. Obl; superior oblique muscle.

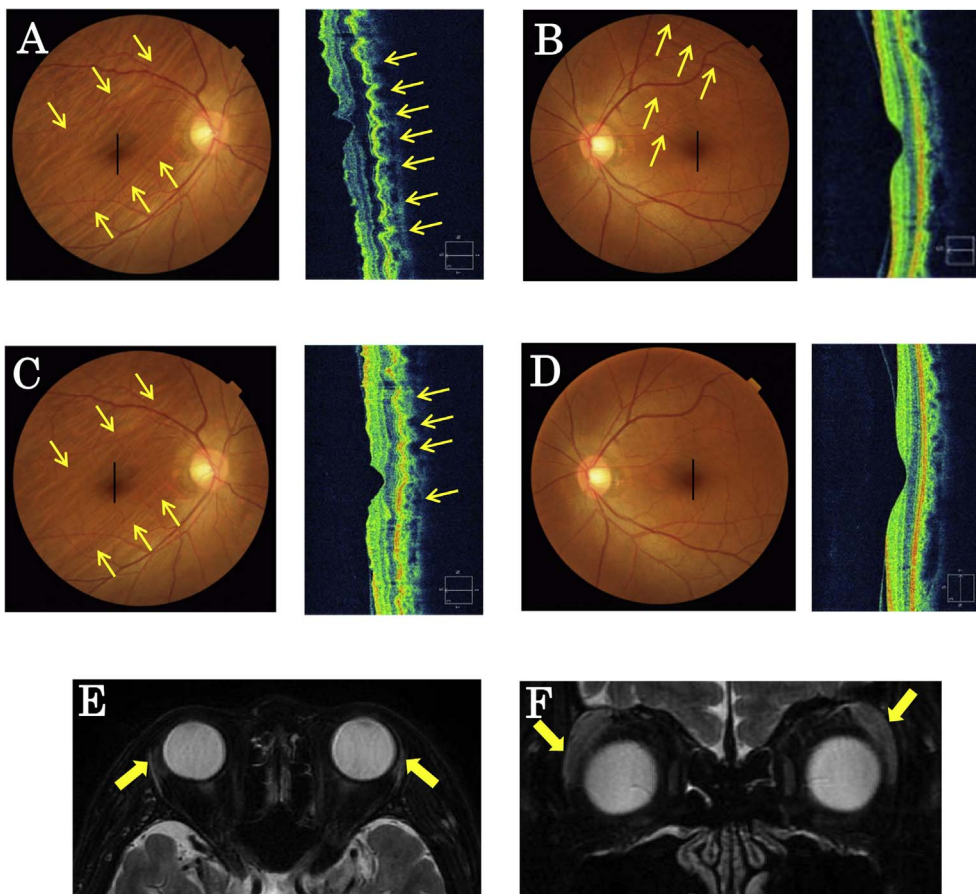


Fig. 4. Fundus photographs and optical coherence tomography (OCT) images just before treatment (A and B) and three months afterwards (C and D) along with magnetic resonance imaging (MRI) findings three months after therapy commencement (E and F). Chorioretinal folds are more obvious than those in the initial examination for the right fundus (A, arrows) and have appeared in the left fundus (B, arrows; since the folds do not reach the macula, none are seen in the OCT image). Three months after starting oral prednisolone, the chorioretinal folds are resolved in the left fundus (D) but remain in the right fundus (C, arrows) despite a markedly decreased volume of the bilateral lacrimal glands and no apparent compression of the globes on MRI (E and F, arrows).

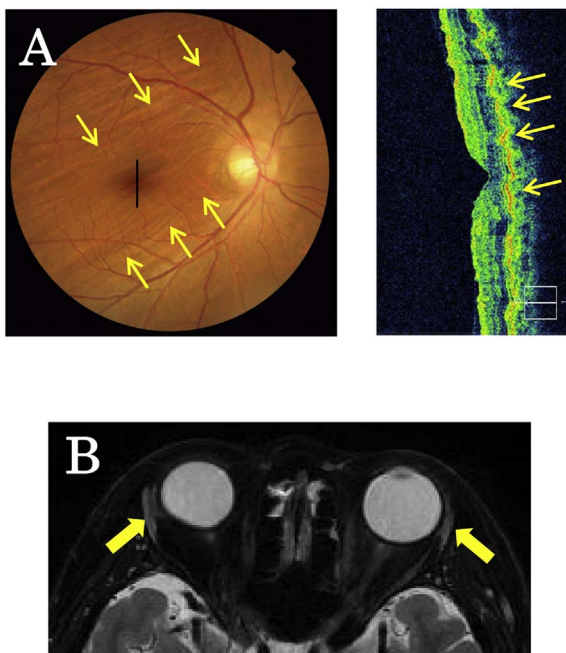


Fig. 5. Right fundus photographs and images of optic coherence tomography and orbital magnetic resonance imaging (MRI) two years after starting treatment. Chorioretinal folds were not altered (A, arrows) regardless no relapse on MRI (B, arrows).

lacrimal glands, storiform-type fibrosis and obliterative phlebitis are often inconspicuous or absent.^{5,11,23,25} Therefore, specific diagnostic criteria are required for each organ. In 2015, the diagnostic criteria for ROD were released by Goto and a Japanese study group of IgG4-ROD.⁵

In these criteria, the pathological findings of fibrosis and obliterative phlebitis are not necessary and the importance of excluding mucosa-associated lymphoid tissue (MALT) lymphoma is emphasized (Table 1).⁶ In the present case, abundant infiltration of lymphocytes and plasmacytes, including IgG4-positive plasmacytes, were seen (Fig. 2A–D). The number of IgG4-positive cells was greater than 50/HPF and the ratio of IgG4/IgG-positive cells was greater than 40% (Fig. 2C and D). Only mild fibrosis was observed and obliterative phlebitis was absent (Fig. 2A and B). Atypical lymphocytes were not seen and monoclonal immunoglobulin gene rearrangement was negative. These findings collectively satisfied the pathological criteria for IgG4-ROD (Table 1).⁶ Along with elevated serum IgG4 level and enlargement of the lacrimal glands, our patient met the criteria for definitive IgG4-ROD (Table 1).⁶

Sclerosis following the enlargement of organs in IgG4-RD is generally believed to be caused by fibrosis.³ A major determinant of treatment is the degree of fibrosis in affected organs since long-standing and highly fibrotic lesions tend to respond poorly to PSL.^{3,23} In the present case, the volume of the lacrimal glands diminished dramatically by oral PSL, indicating that the cause of enlargement and sclerosis was most likely abundant plasmacyte and lymphocyte infiltration into the glands rather than fibrosis. The mild fibrosis pathologically witnessed in the patient supports this hypothesis.

The chorioretinal fold worsening during watchful waiting was clearly apparent in OCT images of the right fundus, where the folds had developed in the central retina (Fig. 4A). Although the chorioretinal folds in the left fundus formed in the absence of PSL and disappeared after commencing steroid treatment (Fig. 4B and D), those in the right fundus remained in spite of compression release from the bilateral lacrimal glands on MRI (Fig. 4C, E, F and Fig. 5A, B). Wolter reported the case of a large cavernous hemangioma wherein retinal folds remained even at three years after complete tumor removal and speculated that

prolonged indentation of the globe culminated in permanent shrinkage of the sclera.¹⁷ Apart from that, we could find only the present case of long-term residual chorioretinal folds after release of globe compression. The patient had been experiencing discomfort in the right eyelid for two years prior to presentation. Thus, our fundus findings also suggested that long-standing, intensive compression of the globes by IgG4-related dacryoadenitis caused irreversible changes not only to the sclera, but also to the retina and choroid. Glucocorticoids are the first-line agent for both IgG4-RD and IgG4-ROD.^{3,11,23} However, some patients refuse or halt oral PSL due to side effects, which results in worsening of ocular dysfunction. Not all manifestations in IgG4-RD need immediate treatment.²³ For example, patients with IgG4-related dacryoadenitis who are asymptomatic apart from lid swelling or those with asymptomatic lymphadenopathy and mild submandibular gland enlargement often require simple observation.³ However, considering the present case, early treatment before irreversible chorioretinal changes may lead to a more favorable prognosis in IgG4-related dacryoadenitis. Frequent fundus examination is advised for chorioretinal folds in patients under observation for enlarged lacrimal glands.

4. Conclusions

The number of reports on IgG4-RD and IgG4-ROD is steadily increasing. Although nomenclature and diagnostic criteria have been established for IgG4-ROD,^{2,6} its pathogenesis remains unclear. Representing more than 20% of orbital lymphoproliferative disorders in Japan, IgG4-ROD is not a rare disease^{9,26} and enlarged lacrimal glands are often seen in this condition.⁹ As far as we know, however, IgG4-related dacryoadenitis causing deformity of the globes and subsequent chorioretinal folds has never been described. Our patient's extremely enlarged, palpable lacrimal glands were apparently caused by infiltration of inflammatory cells rather than fibrosis. Frequent fundus examination and immediate treatment are recommended to avoid irreversible changes and obtain good visual outcomes in patients presenting with chorioretinal folds in IgG4-related dacryoadenitis. Bilateral chorioretinal fold formation by an enlarged lacrimal gland may represent a basis of suspecting IgG4-related dacryoadenitis.

Patient consent

Consent to publish this case was obtained orally from the patient. This report does not contain any personal identifying information.

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Conflicts of interest

The following authors have no financial disclosures: TK, TM, HH, TU, SM, and TM.

Authorship

All authors attest that they meet the current ICMJE criteria for

authorship.

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