

**Case Report** 

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# A Case of IgG4-Related Sclerosing Disease Presenting at the Tarsal Conjunctiva

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#### Abstract

IgG4-related disease (IgG4-RD) is a relatively newly-described disease entity, increasingly recognized for its involvement of the orbital tissues and lacrimal glands. We report a 66-year-old male patient with unusual presentation of IgG4-related sclerosing disease involving the right upper eyelid tarsal conjunctiva only. The lesions subsided after local treatments with no recurrence thus far.

**Keywords:** IgG4-related sclerosing disease; Tarsal conjunctiva

### Introduction

IgG4-related disease (IgG4-RD) is a systemic immune-mediated disease that presents as localized or multi-focal mass lesions in the body. It can affect almost every organ in the body including the orbit and ocular adnexa, [1] which was first reported in 2007 [2,3]. Later, the term IgG4-related ophthalmic disease (IgG4-ROD) was introduced [4]. The exact prevalence of IgG4-ROD is largely unknown [5]. Various foci of IgG4-ROD have been described, but unilateral and isolated involvement of the eyelid conjunctiva is rarely seen. We present a case of unilateral eyelid conjunctival lesions, which was subsequently biopsied and diagnosed as IgG4-related sclerosing disease. No other concurrent ophthalmic or systemic involvements were detected. The lesions responded well to local treatment alone and are maintained in remission.

#### **Case Presentation**

A 66-year-old man presented with 9-months history of right eye discomfort and found to have large and firm conjunctival nodules over the right upper eyelid tarsal conjunctiva, the largest size was  $13 \times 8$  mm (Figure 1). No treatment was given preoperatively. An incisional biopsy performed showed pathological features of IgG4-related sclerosing disease (Figures 2 and 3) with IgG4 plasma cells over 100 HPF and IgG4:IgG ratio over 80%. The patient had normal serum IgG level but elevated IgG4 of 2.552 g/L (N=0.168-1.000 g/L). CT orbit showed no orbital mass and there was no systemic involvement. Topical antibiotic and steroid ointment was given post-operatively for 40 days. Residual nodules resolved, with no recurrence seen at 43 months.



**Figure 1:** Clinical photo of conjunctival nodules over the right upper eyelid tarsal conjunctiva, the largest size was  $13 \times 8$  mm.

#### Discussion

IgG4-related disease is a relatively newly-described disease entity. Little is known about the prevalence of IgG4-ROD, an organ-specific subset of IgG4-related disease. Some investigators have estimated the incidence to be 0.28-1.09 per 1,000,000 individuals. [5] However, several retrospective studies have found that a portion of orbital lesions previously diagnosed as idiopathic orbital inflammation and some other diseases in fact satisfy the criteria for IgG4-ROD, [1] suggesting that IgG4-ROD may be an under-recognized condition in clinical practice.



**Figure 2:** IgG4 stain: Magnification 400x showing increased in IgG4 cell count and increased IgG4 to IgG ratio.



Figure 3: H&E stain: Magnification 400x showing plasma cells in abundance.

Various foci of IgG4-ROD have been described. Common sites of involvement include the lacrimal gland, orbital fat and extraocular muscles [6]. Involvement of the conjunctiva is rare [6] with only a few cases reported, [7-11] suggesting a possible underlying pathological mechanism distinct from other ophthalmic tissues. The case we present resembles the previous cases of conjunctival IgG4-RD in the unilaterality and non-specific clinical features of the lesions. In contrast, the location of the involved conjunctiva and absence of other concurrent ophthalmic or extra-ophthalmic involvements make this case distinct from the others. Two cases of unilateral and isolated conjunctiva involvement have been reported but both lesions are on the bulbar conjunctiva [9,11]. On the other hand, IgG4-RD on the tarsal conjunctiva is usually accompanied by the presence of other disease foci [8,10]. The case of IgG4-RD with unilateral and isolated lesions on the eyelid conjunctiva we present is rarely described in the literature, which contributes further to our understanding of the

disease process. Besides, the case highlights that such lesions can be the sole initial presentation of IgG4-RD, which may be difficult to distinguish from other nodular pathology on the eyelid conjunctiva clinically. In fact, Leivo et al reported one such lesion of IgG4-RD misdiagnosed as chalazion with concurrent extra-ophthalmic skin lesions on presentation [10].

Local treatments were employed in this case, as opposed to systemic glucocorticoids commonly used as the first-line treatment in IgG4-ROD [12]. All the lesions subsided spontaneously in our case with no signs of recurrence thus far. Hence, we propose that localized treatments alone can be used as initial treatments to eradicate such localized lesions with no orbital or systemic involvement, although Philippakis et al. reported that previous experience in a similar lesion was unsatisfactory [10]. This can help to minimize the systemic side effects of steroid and other immunosuppressive agents commonly used to treat IgG4-ROD.

Even though the case is maintained in remission for some time, it is still followed up and monitored for the possibility of relapse [5] and other extra-ophthalmic disease that may present in the future [12]. Moreover, there is a possible association between IgG4-RD and B-cell lymphomas [5]. Nevertheless, there are currently no recommendations on how to follow up stable patients in remission, as the exact course of IgG4-RD is still poorly understood [13].

## Conclusion

Unilateral and isolated tarsal conjunctival mass without other concurrent ophthalmic or systemic involvements may be the initial presentation of IgG4-related disease, among a wide range of ocular manifestations. Localized treatments such as topical steroid and surgical excision can potentially be effective first line treatments for confined localized disease. There is currently no guideline on followup for stable disease.

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