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CASE REPORT

The tarsal plate manifestation of IgG4-related disease

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Abstract
Purpose To describe a clinical case of bilateral biopsy-proven IgG4-related disease confined to the tarsal plate.
Method Interventional case report.
Results A 58-year-old woman presented with a tarsal swelling in the lateral part of the upper eyelids, with focal ulceration and mucus. Histology revealed fibrotic inflammation with increased IgG4-positive plasma cells fulfilling the criteria of IgG4-related disease (IgG4-RD). Serum IgG4 levels were increased, and pathological fluorodeoxyglucose uptake at positron emission tomography/CT scanning was restricted to the upper eyelids. After treatment with oral and topical prednisone, the tarsal lesions markedly regressed.
Conclusions Periorbital IgG4-RD may be confined to the tarsal plate. Treatment with systemic and topical steroids may induce significant regression.

Keywords IgG4-related disease · Orbital · Eyelids · Tarsus

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Introduction

IgG4-related disease (IgG4-RD) is an increasingly recognized fibro-inflammatory condition leading to tumour-like infiltrations of the affected tissues [1]. It is characterized by a lymphoplasmacytic infiltrate enriched in IgG4-positive plasma cells, storiform fibrosis, obliterative phlebitis and often a mild-to-moderate tissue eosinophilia [2]. IgG4-RD predominantly affects the salivary glands, pancreas, orbital tissue and the retroperitoneal cavity, either singly or systematically; however, manifestations in almost all parts of body have been described [3, 4]. In the case of IgG4-related orbital disease, patients mostly present with lacrimal complaints, periorbital masses or, less frequently, scleritis or uveitis [5, 6]. Here, we describe a rare case of bilateral tarsal IgG4-RD.

Case presentation

A 58-year-old woman, with a history of rosacea, presented with painless bilateral swelling of the eyelids, mild hyperaemia of the conjunctiva and mucopurulent discharge since 3 months. She reported no improvement on topical antibiotics prescribed by her general practitioner. On clinical examination, tarsal swelling was observed, more apparent in the lateral part of the upper eyelid, right more than left, with focal ulceration and mucus (Fig. 1a).

Histopathological examination from a tarsal biopsy demonstrated a storiform fibrosing inflammation with increased numbers of IgG4-positive plasma cells (> 200 per high-power field, with IgG4/IgG ratio > 0.80), fulfilling the criteria for IgG4-RD (Fig. 2).

General blood testing showed no irregularities except for elevated serum IgG4 concentration at 439 mg/dL (normal < 135 mg/dL). FDG-PET/CT scans showed increased accumulation in the bilateral upper eyelids without signs of systemic manifestation of the disease.

For the management of the tarsal lesions, oral prednisolone was administered at a 60 mg/day initial dose and was slowly tapered down within 6 months. In addition, topical steroids were prescribed. During the first 6 months after initiation of therapy, there was marked regression leading to residual tarsal scarring (Fig. 1b). No signs of recurrence, systemic involvement, or progression to lymphoma occurred after 10 months.

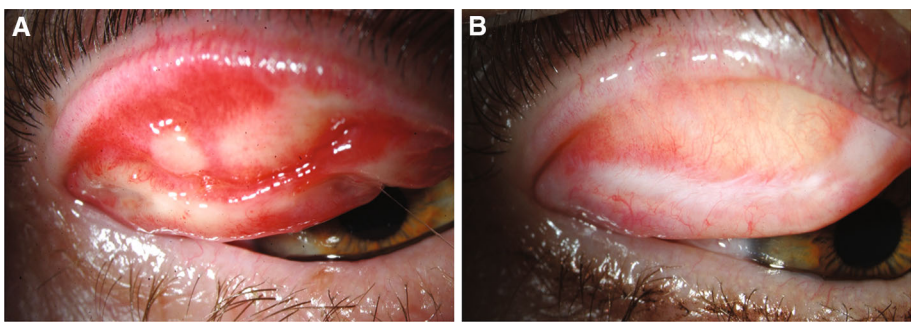

Fig. 1 Photograph demonstrating tarsal IgG4-related disease before (**a**) and after treatment (**b**) 6 months of treatment with systemic and topical steroids

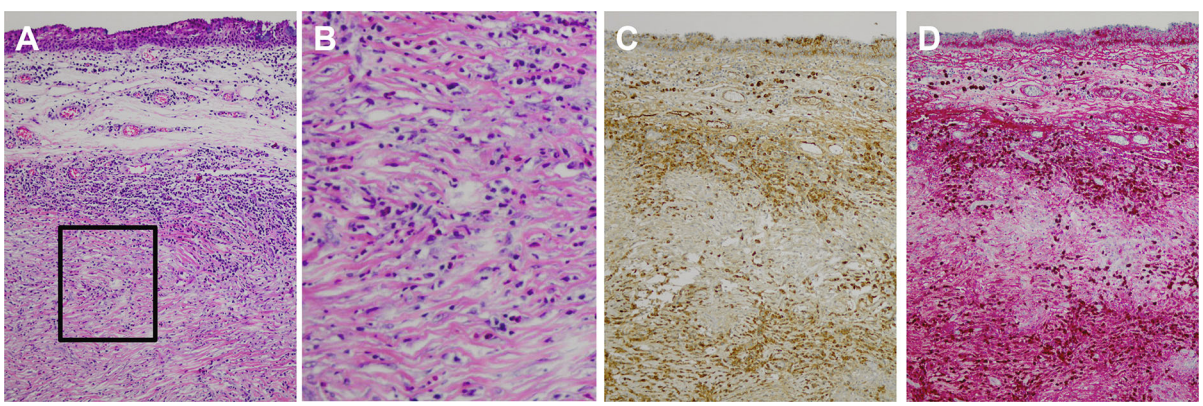

Fig. 2 **a** Histopathology showing a lymphoplasmacellular infiltration with occasional eosinophilic granulocytes, atrophy of the tarsal glands and fibrosis visualized with haematoxylin and eosin stain. **b** Magnified view of (**a**), showing storiform fibrosis; the pattern is characterized by loosely arranged whorls of elongated spindled fibroblast-like cells, representing spokes of the wheel. **c** Increased numbers of IgG4-positive plasma cells (> 200/HPF) visualized with IgG4 immunohistochemical staining in red. **d** Almost equal numbers of IgG-positive plasma cells (IgG4/IgG ratio 0,8) visualized with IgG immunohistochemical staining in brown. All images $\times 100$ original magnification

Discussion

We here demonstrate a case of bilateral biopsy-proven periorbital IgG4-RD confined to the tarsal plate successfully treated with local and systemic glucocorticoids.

IgG4-RD may primarily present in the orbit and periorbital tissue. IgG4-RD is increasingly recognized as part of the spectrum of orbital diseases [6]. Recognition of IgG4-RD is important because the disease can present as a multisystem inflammatory disorder. Early recognition may avoid permanent organ dysfunction and disability [7]. The present case demonstrates the importance of pathologic confirmation. The differential diagnosis includes other causes of inflammation (infectious and autoimmune) and tarsal gland carcinoma (especially in unilateral disease).

To our knowledge, two cases of tarsal involvement have been reported previously [8, 9]. The clinical presentation in these patients differed in that these lesions were more nodular and in one case mimic a chalazion. In these cases, treatment was locally and included either excision or local injection of triamcinolone acetonide. The ulceration in our case necessitated a combination of topical and systemic steroids yielding a complete regression.

We monitor the patient for signs of recurrence, systemic involvement, and potential progression to lymphoma.

In conclusion, IgG4-RD may manifest in the tarsal plate and may require systemic immunosuppressive treatment.

Compliance with ethical standards

Informed consent Informed consent was obtained from the patient for publication of the case report and any accompanying images.

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