Ocular Immunoglobulin G4-Related Disease. A Case Series.

Introduction
Immunoglobulin G4-related disease has emerged over the last decade as a systemic, chronic, inflammatory disorder that can present to the Ophthalmologist.

Its pathophysiology is not, yet, completely understood. Molecular mimicry with cross-reactivity between antibodies against bacterial peptide sequences and the body’s autologous proteins in a genetically predisposed individual may be explanatory.

Objective
Through the adjacent case descriptions we aim to describe some of the common features of Immunoglobulin G4-related disease.

Case Series
We present the cases of three patients with Immunoglobulin G4-related orbitopathy.

Table 1. The cases of three patients thought to have Immunoglobulin G4-related ocular disease are presented. The key features of their clinical examination and extracocular manifestations of their disease, and the findings at orbital imaging and the histological analysis of orbital tissue samples are given.

<table>
<thead>
<tr>
<th>Age and Gender</th>
<th>Presenting feature</th>
<th>Medical history</th>
<th>IgG4</th>
<th>Additional findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>Case 1</td>
<td>46 years Male</td>
<td>Proptosis Eyelid fat prolapase</td>
<td>Lymphadenopathy</td>
<td>1.68g/L</td>
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<tr>
<td>Case 2</td>
<td>28 years Male</td>
<td>Proptosis Eyelid swelling</td>
<td>Asthma Pulmonary nodules Sinusitis Myocardial infarct</td>
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<tr>
<td>Case 3</td>
<td>31 years Female</td>
<td>Eyelid swelling</td>
<td>Optic neuritis</td>
<td></td>
</tr>
</tbody>
</table>

Spontaneous remissions have been reported. Watchful waiting with close follow up examinations may be an option in patients without organ dysfunction and with disease in locations unlikely to cause complications.

When vital organs are involved aggressive treatment is necessary as IgG4 related disease can rapidly lead to organ dysfunction and failure.

Glucocorticoids are the mainstay of treatment and maintenance therapy.

Rituximab is a very specific agent which interferes with the immunological processes thought to underlie IgG4 disease.

Relapses do occur.

Conclusions
Immunoglobulin G4-related disease is being diagnosed with increased frequency. It should be considered in the differential diagnosis of anyone presenting with orbital pathology.

The work-up requires clinical examination, serological analysis, imaging and histology from affected tissue. It must be distinguished from infections, malignancy and other systemic diseases.

Interdisciplinary collaboration is important for proper diagnosis and treatment.