

## Intraocular IgG4 disease masquerading as nodular scleritis

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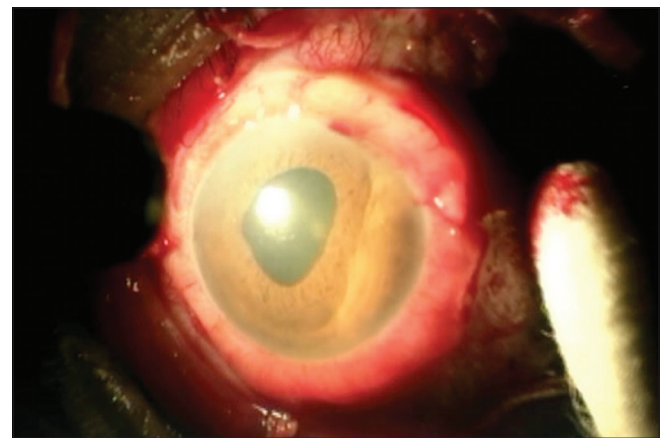
**Key words:** Excision, eye, histopathology, IgG4 disease, immunohistochemistry, malignancy, sclerouvectomy, surgery, tumor

A 37-year-old gentleman presented with complaints of redness in left eye of 4 months duration and blurring of vision of 1 month duration. He was referred with a diagnosis of scleritis and was being treated with topical and oral steroids since 3 months but remained symptomatic. He did not have any systemic comorbidities. Best corrected visual acuity was 6 / 6. Anterior segment examination revealed diffuse scleritis with anterior uveitis. Posterior segment examination showed exudative retinal detachment with choroidal detachment, with overlying vitreous haze in the inferotemporal quadrant corresponding to the area of scleritis. Rheumatoid factor was negative and antinuclear antibody by indirect immunofluorescence was negative. C-reactive protein was <6 mg/L, erythrocyte sedimentation rate was 10 mm/h. As the scleritis was not responding to steroids he was started on oral methotrexate 15 mg/week with folic acid. After 2 months of immunosuppressive therapy, there was no improvement and the vision worsened to 3 / 60. Examination revealed a retroiridal tumor in the temporal half with distorted pupil, shallow anterior chamber, circumcorneal congestion, panuveitis with scleritis, and increased intraocular pressure [Fig. 1]. Ultrasound biomicroscopy revealed a homogenous reflective, echoeic ciliary body mass, measuring 4.6 × 3.7 mm, extending from 2 to 6 O' clock position with overlying episcleral thickening and nasally subluxated lens [Fig. 2]. Positron emission tomography revealed no extraocular spread or metastasis.

Fine needle aspiration biopsy showed clumps of oval cells with high nuclear to cytoplasmic ratio, mixed inflammatory cells, and atypical cells. He underwent partial lamellar sclerouvectomy (PLSU). Histopathology revealed fibrosis with infiltration by mixed inflammatory cells in episcleral tissue, sclera, and ciliary body. Ciliary body tumor showed focal collection of small lymphocytes forming lymphoid follicle and numerous plasma cells [Figs. 3 and 4]. Immunohistochemistry showed presence of 50–60 immunoglobulin 4 (IgG4) plasma cells per high power field (plasma cells were positive for CD138, kappa and lambda), although serum IgG4 level was normal at 1.02 g/L. Postoperatively scleral thinning was noted at the site of PLSU and hence scleral patch graft was done [Fig. 5]. Tapering course of oral steroids was given for a period of 6 weeks postoperatively. The patient developed cataract on follow-up but refused to undergo further surgery.

### Discussion

IgG4-related disease is associated with formation of typical mass lesions which show dense lymphoplasmacytic infiltration, fibrosis, IgG4 positive plasma cells, and often elevated serum IgG4 concentration.<sup>[1]</sup> Our patient did not have elevated serum IgG4 levels. The most common ocular structures involved include lacrimal gland, orbital soft tissue, and extraocular muscles.<sup>[2,3]</sup> Rare reports of intraocular IgG4 disease with



**Figure 1:** External photograph reveals retroiridal tumor extending from 2 to 6 O'clock position with distorted pupil, shallow anterior chamber and circumcorneal congestion

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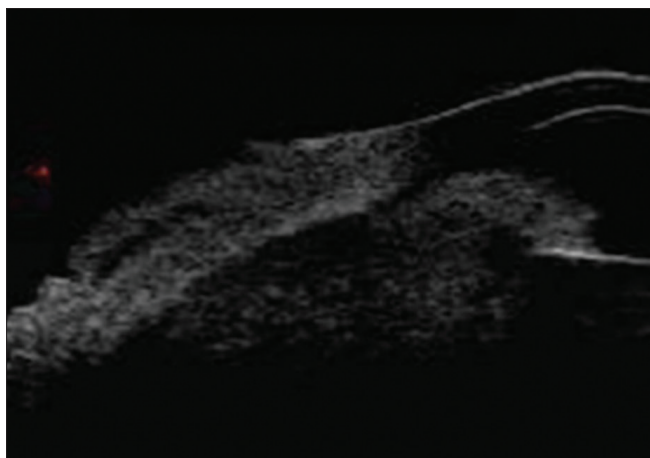
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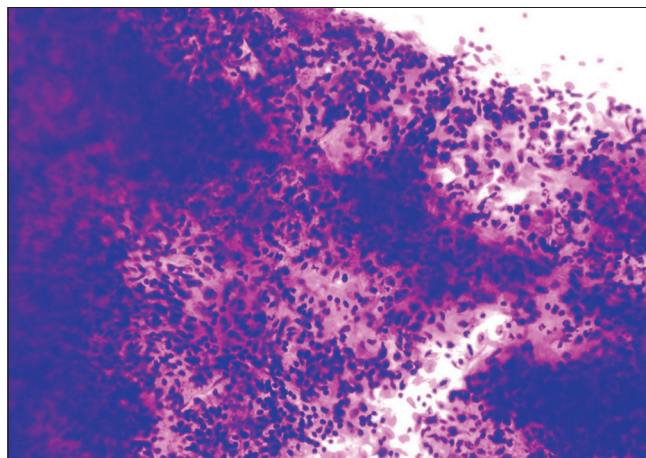
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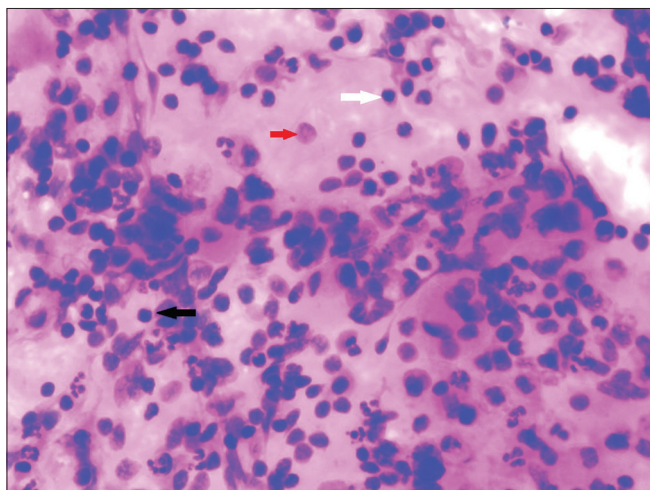
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**Figure 2:** Ultrasound biomicroscopy shows a homogeneously reflective ciliary body tumor measuring 4.6 × 3.7 mm extending from 2 to 6 O'clock position with adjoining episcleral thickening



**Figure 3:** Microphotograph (H&E, 10×) shows mixed inflammatory cells



**Figure 4:** Microphotograph (H&E, 40×) shows mixed inflammatory cells composed of numerous plasma cells (white arrow), small lymphocytes (black arrow) and few neutrophils (red arrow)

scleritis, uveitis, or pseudomelanoma have been reported.<sup>[4,5]</sup> We report a rare case of intraocular IgG4 disease.

#### Declaration of patient consent

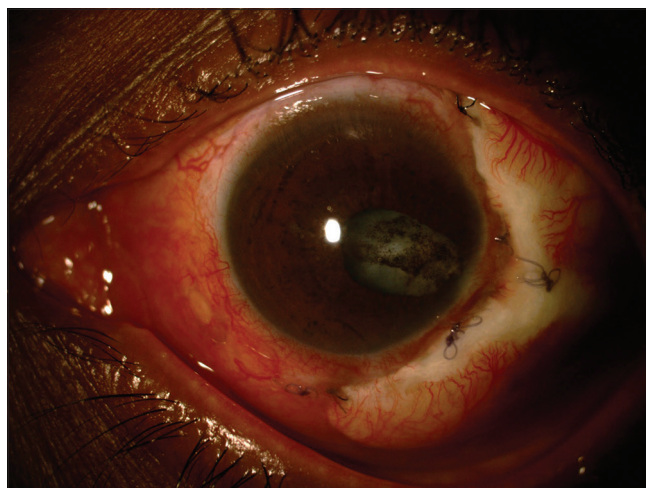
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

#### Conflicts of interest

There are no conflicts of interest.



**Figure 5:** Postoperative slit-lamp photograph showing scleral patch graft in the area of partial lamellar sclerouvectomy

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