

Figure 2. Fundus photographs of the right eye. A, Mild optic nerve fullness and fine macular striae. B, Inferotemporal retina of the right eye showing a large chorioretinal scar with hyperpigmented edges and central retinitis with adjacent retinitis associated with overlying vitreous debris secondary to toxoplasmosis. C, After antibiotic therapy, the area of chorioretinitis decreased in size and developed pigmented edges.

mented edges. The anterior scleritis resolved. Prednisone treatment was slowly tapered once improvement was seen in the vitritis, and the patient reported a resolution of her symptoms.

Comment. Toxoplasmosis is believed to be the most common cause of posterior uveitis.² Scleritis in association with toxoplasmic retinochoroiditis is an uncommon entity.¹ Accordingly, in a review of 243 patients with scleritis, no patient was reported to have toxoplasmic infection as a cause; furthermore, 37% had a systemic rheumatologic disorder, only 7% had an infection, and 44% had an associated medical disorder. Herpes zoster virus was the most commonly reported infectious cause and rheumatoid arthritis was the most common rheumatic disease.⁴

In cases of toxoplasmic scleritis, the inflammatory response is believed to extend from the active retinochoroiditis to involve the overlying sclera. Accordingly, pathologic specimens from eyes that were enucleated secondary to severe toxoplasmic scleritis displayed granulomatous inflammation of the retina, uvea, and episclera with associated retinal thickening. In many cases, the entire sclera extending outward from the retinitis was inflamed; however, in some cases, a region of uninflamed sclera separated the active scleritis from the underlying retinitis.¹

Isolated toxoplasmic retinochoroiditis can rapidly spread and lead to severe permanent vision loss when treated with steroids alone.⁵ In our patient with scleritis, a dilated fundus examination revealed an area of typical toxoplasmic retinochoroiditis, allowing for prompt diagnosis and treatment with appropriate antibiotic therapy. The patient improved and maintained excellent visual acuity. This patient's course underscores the importance of a complete examination in cases of scleritis, including a dilated fundus examination, to rule out an infectious retinochoroiditis in association with the scleritis.

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Unusual Paraneoplastic Cause of Vision Loss: Combined Paraneoplastic Cone Dystrophy and Optic Neuropathy

Paraneoplastic cone dystrophy is a very rare condition with only a few cases reported in the literature.¹⁻⁵ Paraneoplastic optic neuropathy is also a rare cause of cancer-associated visual disturbance. We describe a patient with subacute bilateral vision loss resulting from combined optic neuropathy and cone dystrophy of paraneoplastic origin (occult lung small cell carcinoma). The patient's serum contained antibodies reactive with a novel 42-kDa retinal antigen.

Report of a Case. A 55-year-old man had slight photophobia, photopsias, progressive loss of color perception, and slightly diminished visual acuity in both eyes over a month. Seven days before his initial visit, he developed massive painless vision loss in both eyes over a few hours. His medical history revealed active smoking and alcohol abuse as well as the removal of in situ epidermoid oropharyngeal carcinoma 1 month earlier.

On examination, best-corrected visual acuity was counting fingers at 2 ft OU. Color vision was abolished in both eyes. Slitlamp biomicroscopy showed 2+ cells in the vitreous of both eyes. Fundus examination revealed slightly pallid, swollen optic discs and narrowed arteries in both eyes (**Figure 1A**). Lumbar puncture revealed an elevated level of cells (9/ μ L) and proteins (855 mg/L) in cerebrospinal fluid with normal opening pressure. Brain magnetic resonance imaging

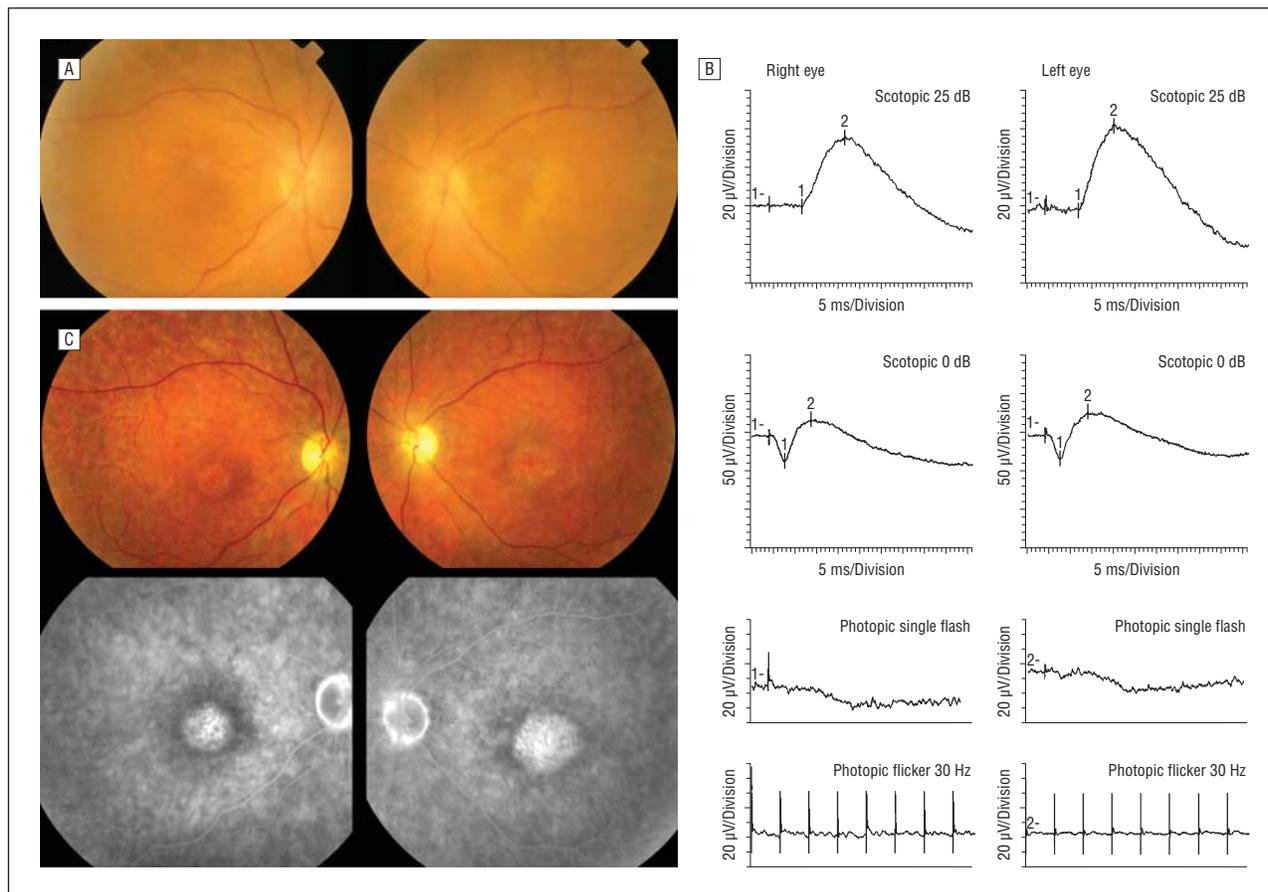


Figure 1. Findings at the initial visit and 8 months after the onset of symptoms. A, Fundus photographs at the initial visit. Narrowed retinal arteries and swollen optic discs were obvious despite vitreous haziness. B, At the initial visit, full-field electroretinographic studies (International Society for Clinical Electrophysiology of Vision Standards) revealed normal rod electroretinographic results (first and second rows) but nonrecordable cone electroretinographic results (third and fourth rows) in both eyes. C, Eight months after the onset of symptoms, fundus photography and fluorescein angiography showed bilateral macular atrophy with a bull's-eye appearance.

was unrevealing. Full-field electroretinography showed bilateral nondetectable cone function but normal rod function (Figure 1B). Paraneoplastic retinopathy was suspected and a chest computed tomographic scan disclosed parahilar lymph nodes. Biopsy revealed metastasis of a neuroendocrine lung small cell carcinoma. Radiotherapy and systemic chemotherapy were initiated.

On follow-up visits, photophobia remained severe, visual acuity was counting fingers OU, and the visual field was markedly constricted under photopic conditions bilaterally. However, under very dim illumination, visual acuity improved to 20/400 OU and the visual field expanded in each eye. Vitreous cells disappeared and bilateral optic disc atrophy ensued. After 8 months, bilateral macular atrophy with a bull's-eye appearance was obvious in both eyes (Figure 1C). The patient's condition remained stable for 4 years with unchanged visual acuity, visual fields, and full-field electroretinographic results. He eventually died of recurrence of his primary lung cancer. No autopsy was performed.

Findings on serum analyses were negative for all of the paraneoplastic antibodies known at the time (1996), including antirecoverin antibody. Serum was sent for further analysis (by C.E.T.), and antibody activity directed

toward a 42-kDa retinal antigen was detected. When incubated with fresh rhesus monkey retina, these antibodies specifically labeled the retinal cone outer segments (Figure 2), providing evidence for a mechanism explaining the cone dysfunction.

Comment. Within 1 month, the patient had experienced a sudden acute and profound bilateral vision loss defined by swollen discs, vitreous cells, and inflammatory spinal fluid related to a paraneoplastic optic neuropathy.

Typical features of an acquired cone dystrophy (decreased visual acuity better with sunglasses, loss of color vision, photopsias, and suppressed photopic response on electroretinography) were initially present. Later, a bull's-eye maculopathy developed and abnormal antibody activity involving the cone pedicles appeared in the patient's serum.

The unique combination of paraneoplastic cone dystrophy and optic neuropathy in our patient was associated with the presence of antibody activity involving a 42-kDa retinal antigen. Indirect fluorescent antibody assays on sectioned rhesus monkey eye associated this antibody activity with the cone outer segments, representing a previously unknown paraneoplastic reaction. The nature of the 42-kDa antigen is not known.

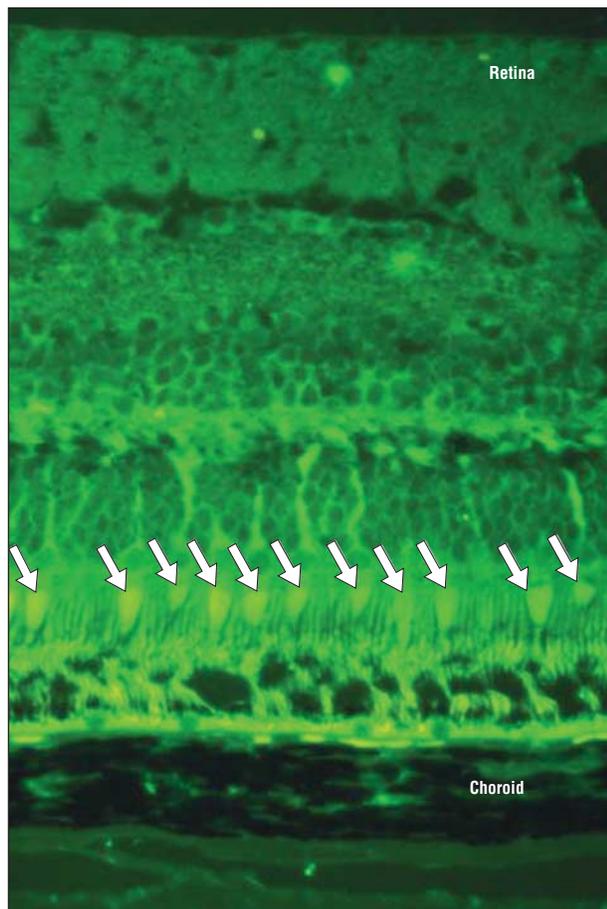


Figure 2. Rhesus monkey retina incubated with the patient's serum. The cone outer segments are specifically labeled by the patient's antibodies (arrows) directed against a single neuronal protein of 42 kDa.

Combined paraneoplastic vision loss (retinopathy and optic neuropathy) has recently been reported in 5 of 16 patients with collapsin response-mediated protein 5 antibodies.⁶ Nine of the 16 patients exhibited vitreous cells, and 15 of the 16 had swollen optic discs.

This case expands the spectrum of clinical entities capable of producing paraneoplastic vision loss.

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Orbital Sarcoma in a Young Patient With Li-Fraumeni Syndrome

We describe the first reported case, to our knowledge, of a myxoid liposarcoma in a young man with a family history of a p53 gene abnormality.

Report of a Case. A 32-year-old man had diplopia and slowly progressive right proptosis for 2 months. He had a family history of Li-Fraumeni syndrome (LFS),¹ with tumor-related death of 2 first-degree and 1 second-degree relatives all before age 30 years.

Visual functions were normal (unaided visual acuity 6/5 OU). Mild right optic disc swelling and macular choroidal folds were evident. There was 5-mm right relative proptosis and a right esotropia. Computed tomography revealed a large, well-defined intracanal mass lying lateral to the right optic nerve and extending to the orbital apex (**Figure 1**).

Biopsy was performed through a lateral canthotomy; the tumor showed a gelatinous gross appearance during handling (**Figure 2A**). Histopathological analysis showed features of a myxoid liposarcoma (**Figure 2B**). There was focal nuclear immunoreactivity with S-100 protein, but other markers including AE1/AE3, glial fibrillary acidic protein, desmin, and CD34 were negative.

Given the family history of LFS, the use of either radiotherapy or alkylating chemotherapy was considered to carry an unacceptable risk of secondary tumor formation.² The patient underwent skin-sparing orbital exenteration with multiple staging biopsies.

Histopathological analysis showed an extensively infiltrating sarcoma with vacuolated cells and lipoblasts. Analysis demonstrated cells with 3 to 5 paired *CHOP* and *FUS* signals, but no cells with a split signal were seen. The multiple signals support the presence of an abnormal clone, but the lack of a split signal is evidence against a translocation of *CHOP* or *FUS*, which would typically be seen in this type of tumor.