ment with both ceftriaxone sodium, 2 g twice daily, and metronidazole, 1 g twice daily, his neurologic condition improved from being unable to walk (0/5 strength) to 4/5 lower-extremity strength. He was discharged for rehabilitation. Intravenous treatment with ceftriaxone was continued for 12 weeks. On follow-up examination 7 months after his initial visit to the ophthalmology clinic, best-corrected visual acuity measured 20/200. A 2+ posterior subcapsular cataract was noted in the left eye, along with a central macular scar (Figure 3).

Comment. *Streptococcus constellatus* is generally a commensal organism found in the mouth, oropharynx, and gastrointestinal tract. This organism has been cultured from dental caries and periodontal disease but has also been isolated from brain abscesses, gastrointestinal perforations, and obstetric infections. To our knowledge, it has not previously been reported as a cause of endogenous endophthalmitis; however, cases of orbital abscesses and cavernous sinus thrombosis after dental work have been reported. *Streptococcus constellatus* is a difficult organism to classify and is commonly misidentified. Although it has been cultured from blood in cases of endocarditis, it grows mainly via abscesses; therefore, an abscess is the best culture source. Many antibiotics eradicate these organisms, but surgical intervention is usually needed for absolute treatment.1

Lena V. Chheda, MD
Warren M. Sobol, MD
Bruce M. Buerk, MD
Paul A. Kurz, MD

Author Affiliations: Department of Ophthalmology, The Ohio State University, Columbus (Drs Chheda, Sobol, Buerk, and Kurz), and Retina Physicians and Surgeons (Drs Sobol and Buerk) and Dayton Veterans Affairs Medical Center (Drs Sobol, Buerk, and Kurz), Dayton.

Correspondence: Dr Kurz, Department of Ophthalmology, The Ohio State University, Havener Eye Institute, 915 Olentangy River Rd, Columbus, OH 43212 (paul.kurz@osumc.edu).

Financial Disclosure: None reported.


Rosai-Dorfman Disease Simulating Nodular Scleritis and Panuveitis

Scleritis is a potentially sight-threatening inflammatory condition of the sclera that may be associated with keratitis, uveitis, glaucoma, and exudative retinal detachments.1 Sinus histiocytosis with massive lymphadenopathy, or Rosai-Dorfman disease, is a rare histiocytic disorder characterized by massive, painless lymphadenopathy.2,3 While extranodal involvement is common, eye involvement is infrequent and most often seen in the orbit or eyelid.4 Direct ocular involvement is exceedingly rare.5 We report a case of Rosai-Dorfman disease that simulated nodular scleritis and panuveitis.

Report of a Case. A 20-year-old woman with diabetes, hypertension, and hypothyroidism had a 3-day history of severe pain and vision loss in the left eye. She also noticed a slowly enlarging nodule on the left eye over the previous 5 months. Review of systems was negative for fever. Visual acuity was 20/20 OD and counting fingers OS. Intraocular pressure was 43 mm Hg OS. Examination showed an elevated perilimbal subconjunctival nodule, large keratic precipitates, 3+ anterior segment inflammation, 2+ vitreous haze, and subretinal exudate (Figure 1 A and B). Ultrasonography revealed diffuse scleral thickening. Laboratory testing revealed no leukocytosis, a normal angiotensin-converting enzyme level, a nonreactive rapid plasma reagin test result, and a slightly elevated erythrocyte sedimentation rate (25 mm/h). Tuberculosis skin testing and chest radiography results were normal. Results of testing for antinuclear antibodies, anticytoplasmic nuclear antibodies, and HLA-B27 were negative.

She was diagnosed as having nonnecrotizing nodular scleritis with panuveitis and was treated with oral prednisone, topical prednisolone acetate, and topical antihypertensive eyedrops. She experienced frequent relapses over the following 6 months and began treatment with oral methotrexate. She was lost to follow-up for 4 years. During this time, a systemic evaluation for pelvic lymphadenopathy and rheumatologic disease was negative. She returned to the eye clinic with recurrent left eye pain and vision loss. She was restarted on a course
of oral prednisone and subsequently underwent 2 subconjunctival injections of triamcinolone acetonide. Her ocular inflammation subsided and remained inactive for 3 years, but her vision was ultimately limited by a mature cataract. Progressive enlargement of the mass eventually led to severe globe displacement and an inability to close her eyelids.

Partial resection of the epibulbar mass was performed with scleral patch grafting (Figure 1C). Histopathologic examination showed histiocytes exhibiting emperipolesis and S-100 protein positivity (Figure 2), which are diagnostic for Rosai-Dorfman disease. At last follow-up, 1 year after resection, significant tumor regrowth had occurred (Figure 1D).

Comment. Rosai-Dorfman disease is a rare, idiopathic, benign proliferative histiocytic disorder originally described in 1969. It is most common in children and adolescents and is typically characterized by massive cervical lymphadenopathy, fevers, leukocytosis, and elevated erythrocyte sedimentation rate. Our patient was atypical in that she lacked cervical lymphadenopathy and leukocytosis and denied having fevers. Ocular involvement, which may be the initial and only manifestation of the disease, is atypical and seen in only 11% of patients. To our knowledge, there are only 2 other reported cases of Rosai-Dorfman disease with subconjunctival nodules and uveitis.

Rosai-Dorfman disease can be difficult to diagnose and often requires histopathologic examination. Characteristic histology includes a polymorphous infiltrate with a predominance of proliferating histiocytes. The histiocytes usually contain prominent vesicular nucleoli and abundant pale, eosinophilic cytoplasm and exhibit lymphophagocytosis or emperipolesis. Immunocytochemical analysis will show positive staining for S-100 protein.

Prognosis is generally good and usually self-limited. Typically, eventual spontaneous regression or stabilization occurs. While there is no effective cure, treatments include surgical resection, local irradiation, and systemic chemotherapy. Our patient’s disease continued to progress despite local and systemic immunosuppression and re-
section. Although rare, Rosai-Dorfman disease can be confused with nodular scleritis and should be considered in patients with epibulbar masses and uveitis.

John F. Payne, MD
Sunil K. Srivastava, MD
Jill R. Wells, MD
Hans E. Grossniklaus, MD

Author Affiliations: Department of Ophthalmology, Emory Eye Center, Atlanta, Georgia (Drs Payne, Wells, and Grossniklaus); and Department of Ophthalmology, Cole Eye Institute, Cleveland, Ohio (Dr Srivastava).

Correspondence: Dr Grossniklaus, Department of Ophthalmology, Emory Eye Center, 1365 B Clifton Rd, Room BT428, Atlanta, GA 30322 (ophtheg@emory.edu).

Author Contributions: All of the authors had full access to the data in this study. Dr Payne takes responsibility for the integrity of the data and the accuracy of the data analysis.

Financial Disclosure: None reported.

Funding/Support: This work was supported in part by a grant to Emory Eye Center from Research to Prevent Blindness, Inc, New York, New York.


Granulomatous Choroiditis in Wegener Granulomatosis

Wegener granulomatosis (WG) is characterized classically as the triad of necrotizing granulomatous lesions of the upper and lower respiratory tract, focal segmental glomerulonephritis, and necrotizing vasculitis of small arteries and veins.1 Ophthalmological disease is the manifesting feature of WG in 8% to 16% of patients1 but develops in an estimated 50% to 60% of patients.2 Orbital disease is the most common ophthalmological manifestation of WG, uveal involvement is uncommon,2 and granulomatous sclerouveitis is rare.1-5 I describe the first histological documentation, to my knowledge, of granulomatous choroiditis in WG in the absence of scleritis.

Report of a Case. A 71-year-old man developed protracted nausea, a 6.75-kg weight loss, acute renal failure, and pulmonary hemorrhage. He had positive results on a perinuclear antineutrophil cytoplasmic autoantibody assay and an antineutrophil cytoplasmic enzyme-linked immunosorbent assay (level=130.8 U; positive >20.0 U) and negative results on an antiprotease 3 enzyme-linked immunosorbent assay (level=3.9 U; positive >20.0 U). Although he did not have the typical pattern of antineutrophil cytoplasmic antibodies with cytoplasmic staining, his findings were considered most compatible with WG. His renal failure did not resolve with hemodialysis, high-dose methylprednisolone sodium succinate, cyclophosphamide, and plasmapheresis. He had progressively decreasing strength, mental status, and ability to tolerate tube feeding and died approximately 5 months after his initial symptom of nausea developed. A complete autopsy confirmed the diagnosis of WG with necrotizing granulomatous and fibrinous vasculitis with neutrophils and karyorrhectic debris involving the kidneys, testes, appendix, liver, spleen, lungs, pancreas, lymph nodes, small and large intestines, trachea, aorta, pericardium, myocardium, and both orbits (listed in order of decreasing histological severity).

The posterior choroid of both eyes had many foci of granulomatous inflammation similar to those in other organs with mostly epithelioid cells accompanied by lymphocytes and a few multinucleated giant cells (Figure 1). In multiple areas, the choriocapillaris was infiltrated by inflammatory cells and the capillaries were stenotic or occluded by inflammatory cells (Figure 2). Rare minute foci of fibrinoid necrosis with occasional neutrophils and karyorrhectic debris were in the choroid just beneath the choriocapillaris (Figure 2). The choroidal vessels were surrounded by the dense inflammatory infiltrate, but only a rare artery appeared to have its wall infiltrated by lymphocytes without necrosis. Degeneration of the neurosensory retina and scleral inflammation were not seen. Microorganisms were not detected using histochemical stains.

Comment. Choroidal involvement in WG may manifest clinically as uveitis, choroidal folds, retinal epithelial pigmentary changes, choroidal arterial occlusion, or choriocapillaritis.1,2 Only 1 histological description of isolated choroidal involvement by WG exists, to my knowledge.6 The patient described by Cutler and Blatt6...