



Published in final edited form as:

Retin Cases Brief Rep. 2016 ; 10(4): 341–344. doi:10.1097/ICB.0000000000000254.

Severe Panuveitis, Retinal Vasculitis, and Optic Disc Granuloma Secondary to Sarcoidosis

Michael Patterson, DO¹, Chris S. Bergstrom, MD/OD², W. Lloyd Clark, MD^{1,3}, Hans E. Grossniklaus, MD², and John F. Payne, MD^{1,3}

¹Department of Ophthalmology, University of South Carolina, Columbia, SC

²Department of Ophthalmology, Emory Eye Center, Atlanta, GA

³Palmetto Retina Center, West Columbia, SC

Abstract

Purpose—To report a case of panuveitis, retinal vasculitis, and optic disc granuloma due to sarcoidosis.

Methods—Case report and literature review.

Results—A 26 year-old previously healthy African American male presented with four months of gradual progressive visual decline in the right eye. Clinical examination revealed severe panuveitis, retinal vasculitis, and large optic nerve mass lesion. Diffuse supraclavicular lymphadenopathy was also present. Histopathologic examination of the lymph node biopsy revealed granulomatous inflammation with some areas of caseous necrosis, consistent with sarcoidosis.

Conclusion—Sarcoidosis is a common cause of uveitis and retinal vasculitis. In rare cases, an optic disc granuloma may occur and can be treated with immunosuppressive therapy.

Keywords

Optic Disc Granuloma; Retinal Vasculitis; Sarcoidosis; Uveitis

Sarcoidosis is an idiopathic, multisystem granulomatous disorder with ocular involvement occurring in 25–60% of cases.¹ The histopathologic hallmark of the disease is noncaseating granuloma formation.^{2,3} Clinically, optic nerve involvement can present as papilledema, papillitis, and/or granulomatous infiltration of the optic disc.^{4,5} We report a case of sarcoidosis which presented with panuveitis, retinal vasculitis, and optic nerve granuloma.

Corresponding author and reprint requests to: John F. Payne, MD, Palmetto Retina Center, 124 Sunset Court, West Columbia, SC 29169, Phone: (803) 931-0077, Fax: (803) 931-0076, jpayne@palmettoretina.com.

The information contained in this manuscript was presented at the 2013 annual meeting of The Retina Society in Beverly Hills, CA.

None of the authors have any financial or proprietary interest in any material mentioned in this manuscript.

All of the authors had full access to the data in this study. The first author and corresponding author take responsibility for the integrity of the data and the accuracy of the data analysis.

Case Report

A 26 year-old previously healthy African-American male presented with four months of painless vision loss in the right eye. Review of systems was positive for floaters and progressive lymphadenopathy. Visual acuity was hand motions right eye and 20/25 left eye. A relative afferent pupillary defect was present in the right eye, and there was marked bilateral submandibular and supraclavicular lymphadenopathy. Slit lamp biomicroscopy of the right eye showed trace anterior segment inflammation and a large vascularized optic disc granuloma (Figure 1). There was also a peripapillary exudative retinal detachment, diffuse vascular sheathing, and a mid-peripheral choroidal granuloma. Ultrasonography revealed a 6.6 mm elevated optic nerve mass with low internal reflectivity. The left eye contained one focal area of vasculitis, and mild late disc leakage on fluorescein angiography.

The patient was admitted for further evaluation and lymph node biopsy. Angiotensin-converting enzyme and lysozyme levels were markedly elevated, while Quantiferon testing for tuberculosis was negative. Additional serologic testing for syphilis, human immunodeficiency virus, toxoplasmosis, and fungemia were negative. Chest computed tomography demonstrated bilateral hilar lymphadenopathy and diffuse nodular densities. Cervical lymph node biopsy revealed granulomatous inflammation with some areas of caseous necrosis (Figure 2). Acid-fast bacilli and gomori methenamine silver stains were negative for tuberculosis and fungus, respectively, and flow cytometry showed no immunophenotypic evidence of lymphoma.

The patient's clinical presentation as well as radiographic and histopathologic examinations were consistent with advanced sarcoidosis and he was started on intravenous corticosteroids. He was transitioned to a slow oral prednisone taper, and because of the severity and extent of his disease, he was started on subcutaneous adalimumab therapy. There was significant improvement in the lymphadenopathy and subjective visual improvement within the first few weeks of treatment. At 21 months follow-up, his vision remained hand motions, but the optic nerve granuloma had decreased to 2.2 mm in thickness (Figure 3). The left eye remained 20/20 and showed no signs of clinical activity.

Discussion

Sarcoidosis is most frequently a multisystem granulomatous disorder thought to result from an exaggerated immune response to a variety of antigens.¹ The exact pathogenesis remains unclear. The disease most often affects the lungs and thoracic lymph nodes, skin, and eyes, with anterior and posterior uveitis being the most frequent ocular manifestations.¹ The diagnosis remains one of exclusion and is based on clinical, radiographic, and histopathologic findings.¹ Radiographic testing typically reveals hilar lymphadenopathy and is often associated with elevated angiotensin-converting enzyme and lysozyme levels, all of which were present in our patient.¹

This patient's lymph node biopsy revealed tightly formed granulomas with some areas of caseous necrosis. While noncaseating granulomas are the hallmark histopathologic finding in sarcoidosis, Mitchell and colleagues and others reported that minor degrees of caseous

necrosis can still be compatible with the diagnosis.³ Although many of the clinical and histopathologic findings in sarcoidosis resemble those in patients with infectious diseases, no pathogenic organism has been implicated in its cause. In this case, our patient demonstrated negative serology and histopathology for syphilis, tuberculosis, toxoplasmosis, bartonella, cryptococcus, histoplasmosis and human immunodeficiency virus.

Table 1 illustrates the nine reported cases of optic disc granuloma secondary to sarcoidosis since 1980. Our patient was slightly younger than the average age of these patients, which was 35 years. Almost all patients were African American and all had unilateral optic disc granulomas. Our patient appears to have the largest granuloma of those reported. The presenting visual acuity varied among the patients with four eyes having 20/30 vision and four eyes having 20/200 vision. Oral and/or intravenous corticosteroids were the initial treatment of choice and visual outcomes were mixed.

The list of differential diagnoses for a mass lesion on or around the optic nerve head in the setting of uveitis is relatively broad. Neoplastic, inflammatory, and vascular lesions are among the most common etiologies. Specific diseases include peripapillary choroidal hemangiomas or metastasis, amelanotic choroidal melanomas, sarcoid or tuberculous granulomas, and possibly Blau syndrome. Blau syndrome is a rare, autosomal dominant condition typically characterized by early-onset granulomatous arthritis, uveitis and skin lesions.¹² While optic disc granulomas have not been specifically described in this condition, the granulomatous uveitis can be severe and multifocal choroiditis has been reported.¹²

Central nervous system involvement has been estimated to occur in 5–12% of all cases of sarcoidosis.¹³ Optic neuritis, myelopathy, and facial nerve palsy are among the most common presentations of those with neurosarcoidosis.¹³ The prognosis is variable but particularly poor when the optic nerve is involved.¹³ The development of an optic disc granuloma is exceedingly rare. Farr reported a case of acute vision loss secondary to a sarcoid optic disc granuloma and good visual recovery with oral corticosteroid use.⁵ Our patient responded to immunosuppressive therapy but suffered significant vision loss because of the severity and chronicity of disease at presentation.

Supplementary Material

Refer to Web version on PubMed Central for supplementary material.

References

1. Umur KA, Tayfun B, Oguzhan O. Different ophthalmologic manifestations of sarcoidosis. *Curr Opin Ophthalmol*. 2012; 23:477–84. [PubMed: 23014267]
2. Birnbaum AD, Oh FS, Chakrabarti A, Tessler HH, Goldstein DA. Clinical features and diagnostic evaluation of biopsy-proven ocular sarcoidosis. *Arch Ophthalmol*. 2011; 129(4):409–13. [PubMed: 21482866]
3. Mitchell DN, Scadding JG, Heard BE, Hinson KF. Sarcoidosis: histopathological definition and clinical diagnosis. *J Clin Pathol*. 1977; 30(5):395–408. [PubMed: 325017]
4. Katz JM, Bruno MK, Winterkorn JMS, Nealon N. The Pathogenesis and Treatment of Optic Disc Swelling in Neurosarcoidosis. *Arch Neurol*. 2003; 60:426–430. [PubMed: 12633155]

5. Farr AK, Jabs DA, Green WR. Optic disc sarcoid granuloma. *Arch Ophthalmol*. 2000; 118(5):728–9. [PubMed: 10815174]
6. Lustgarten JS, Mindel JS, Yablonski ME, Friedman AH. An unusual presentation of isolated optic nerve sarcoidosis. *J Clin Neuroophthalmol*. 1983; 3(1):13–18. [PubMed: 6222071]
7. Graham EM, Ellis CJ, Sanders MD, McDonald WI. Optic neuropathy in sarcoidosis. *J Neurol Neurosurg Psychiatry*. 1986; 49(7):756–763. [PubMed: 3746306]
8. Asensio Sanchez VM, Corral Azor A, Bartolome Aragon A, De Paz Garcia M. Sarcoidosis of the optic nerve. *Arch Soc Esp Oftalmol*. 2003; 78(3):165–168. [PubMed: 12677494]
9. Frohman LP, Guirgis M, Turbin RE, Bielory L. Sarcoidosis of the anterior visual pathway: 24 new cases. *J Neuroophthalmol*. 2003; 23(3):190–197. [PubMed: 14504590]
10. Koczman JJ, Rouleau J, Gaunt M, et al. Neuro-ophthalmic sarcoidosis: the University of Iowa experience. *Semin Ophthalmol*. 2008; 23(3):157–168. [PubMed: 18432542]
11. Moschos MM, Guex-Crosier Y. Anterior segment granuloma and optic nerve involvement as the presenting signs of systemic sarcoidosis. *Clin Ophthalmol*. 2008; 2(4):951–953. [PubMed: 19668451]
12. Kurokawa T, Kikuchi T, Ohta K, Imai H, Yoshimura N. Ocular manifestations in Blau syndrome associated with a CARD15/Nod2 mutation. *Ophthalmology*. 2003; 110(10):2040–2044. [PubMed: 14522785]
13. Pawate S, Moses H, Sriram S. Presentations and outcomes of neurosarcoidosis: a study of 54 cases. *QJM*. 2009; 102(7):449–460. [PubMed: 19383611]

Summary Statement

The patient presented with profound painless vision loss in the right eye and was found to have severe panuveitis, retinal vasculitis and optic disc granuloma due to sarcoidosis. The disease was managed with immunosuppressive therapy consisting of corticosteroids and adalimumab.

Author Manuscript

Author Manuscript

Author Manuscript

Author Manuscript

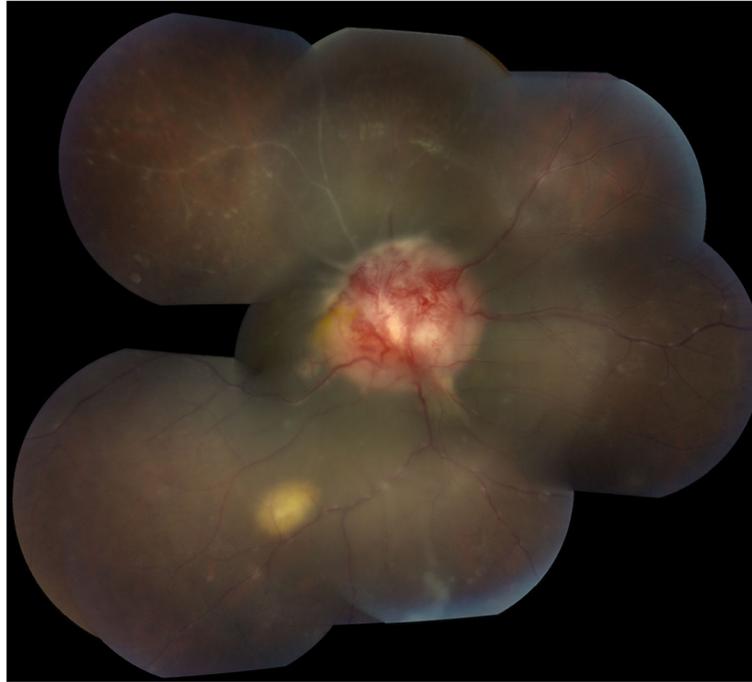


Figure 1. Montage fundus photograph of the right eye showing a very large vascularized optic disc granuloma with peripapillary exudative retinal detachment, and diffuse vasculitis. A mid-peripheral choroidal granuloma is visible in the inferotemporal quadrant.

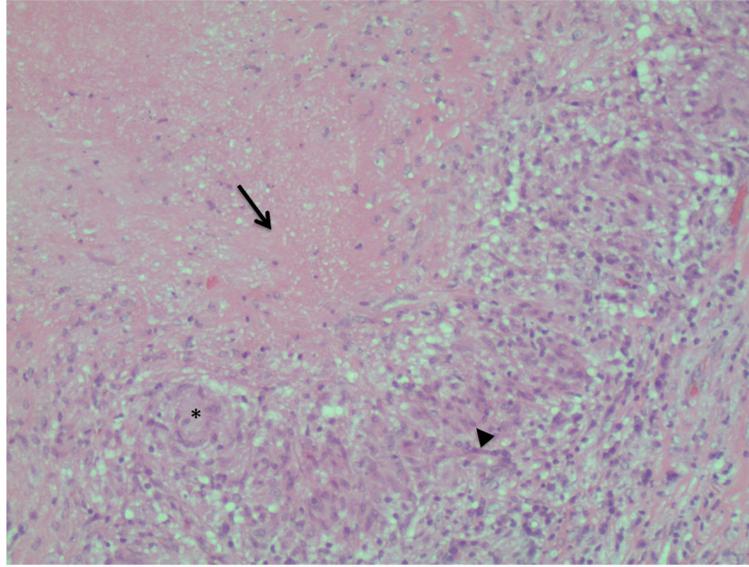


Figure 2. Histopathologic examination reveals granulomatous inflammation including palisading epithelioid histiocytes (arrowhead) and giant cells (*) surrounding an area of caseous necrosis (arrow). (Hematoxylin-eosin, original magnification 10x)

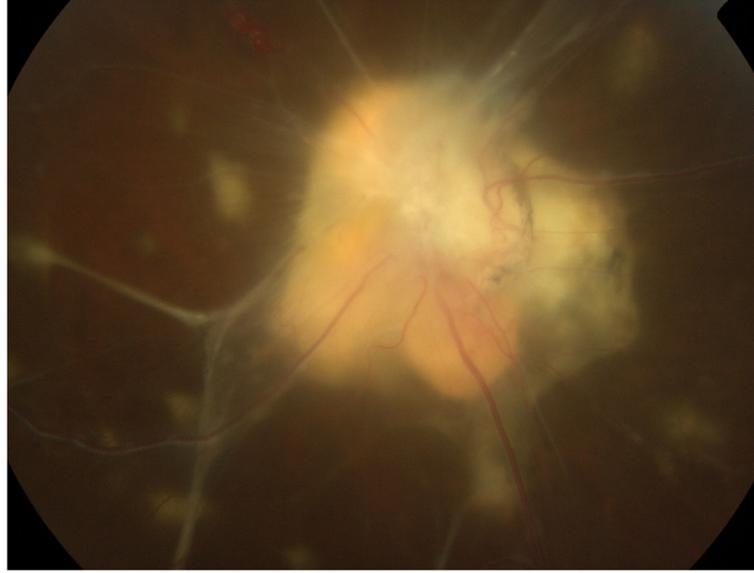


Figure 3. Color fundus photograph of the right eye 21 months after presentation. The optic disc margins are partially visible and the granuloma is much smaller in size. There is extensive prepapillary and subretinal fibrosis and diffuse sclerosis of the retinal vessels.

Table 1

Clinical characteristics and visual outcomes in patients with optic disc granuloma secondary to sarcoidosis since 1980.

| Publication | Age/Sex (Years) | Race | Laterality of Granuloma | Initial VA | Treatment/Clinical Course | Length of Follow-up | VA at Last Follow-up |
|------------------------------|-----------------|------------------|-------------------------|------------|--|---------------------|----------------------|
| Lustgarten ⁶ | 32/Female | African American | Unilateral (Left eye) | 20/30 | Vision declined over several weeks to 20/200, unresponsive to oral corticosteroids. Sudden vision loss to NLP several weeks later due to optic disc granuloma. | 13 months | NLP |
| Graham ⁷ | 43/Female | West Indian | Unilateral (Left eye) | NLP | Oral corticosteroids failed to improve vision | 2 years | NLP |
| Farr ⁵ | 31/Female | African American | Unilateral (Left eye) | 20/25 | 2 months after presentation, vision declined to 20/400. Oral corticosteroids started with 6 month taper | 18 months | 20/25 |
| Asensio Sanchez ⁸ | 59/Male | Not reported | Unilateral (Left eye) | 20/200 | Oral corticosteroids started | 1 year | 20/20 |
| Frohman ⁹ | 50/Female | Not reported | Unilateral (Left eye) | NLP | Not reported | Not reported | Not reported |
| Frohman ⁹ | 32/Female | Not reported | Unilateral (Left eye) | 20/70 | Not reported | Not reported | Not reported |
| Koczman ¹⁰ | 24/Male | African American | Unilateral (Left eye) | 20/25 | Oral/IV corticosteroids and methotrexate | Not reported | 20/15 |
| Moschos ¹¹ | 19/Male | African American | Unilateral (Left eye) | 20/25 | Not reported | Not reported | Not reported |
| Current Study | 26/Male | African American | Unilateral (Right eye) | HM | IV corticosteroids followed by slow oral corticosteroid taper and adalimumab | 22 months | HM |

HM: Hand motions; IV: Intravenous; NLP: No light perception; VA: Visual acuity