



Surgical repair of an inflammatory macular hole associated with ocular syphilis

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ABSTRACT

Purpose: To describe a case of an inflammatory macular hole associated with ocular syphilis and its successful surgical closure.

Observations: A 55-year-old man presented with count fingers vision and was found to have unilateral ocular syphilis with panuveitis and a foveal subretinal lesion. The inflammation quickly resolved with treatment, but he developed a full-thickness macular hole on day 5. A pars plana vitrectomy with membrane peel and intraocular gas was performed three months later. The macular hole was successfully closed, and the patient's vision improved to 20/200 18 months after repair. Foveal outer retinal atrophy limited final vision improvement.

Conclusions and importance: Ocular syphilis can rarely be associated with inflammatory macular holes. Surgical intervention can offer successful anatomic results; however, final visual outcomes may be limited by retinal scarring or atrophy from the original inflammation.

1. Introduction

Syphilitic uveitis can occur at any stage of acquired syphilis. Due to a diverse range of presentations and ocular structures affected, syphilitic uveitis is often considered a “great masquerader” to be included in the differential diagnosis of most forms of ocular inflammation.¹ The literature reveals only a single case report of a syphilitic macular hole that eventually closed with conservative management.^{2,3} The following describes the first reported case of a syphilitic macular hole necessitating surgical repair.

2. Case report

A 55-year-old Cape Verdean man presented with 4 days of pain, decreased vision, and enlarging central scotoma in his right eye. His visual acuity was count fingers at 1 foot in the right eye and 20/25 in the left. Intraocular pressure was 16 mmHg in the right eye and 15 mmHg in the left eye. Examination of the right eye revealed fine keratic precipitates, anterior chamber cell and pigment, and diffuse vitritis. Fundus examination showed a white, elevated lesion in the fovea with adjacent intraretinal hemorrhage as well as scattered moderate-sized peripheral intraretinal hemorrhages and temporal vascular sheathing (Fig. 1A) with no anterior chamber cell, vitritis, or posterior segment

inflammation or lesions.

Spectral-domain optical coherence tomography (SD-OCT) and autofluorescence (AF) were performed using Heidelberg Spectralis (Heidelberg Engineering, Heidelberg, Germany), while fluorescein angiography (FA) was performed with Optos 200Tx (Optos plc, Dunfermline, Scotland). SD-OCT showed subretinal hyperreflective material at the fovea with overlying subretinal fluid in the right eye (Fig. 1B). Autofluorescence images showed hypoautofluorescence secondary to blockage in the area of the macular lesion and intraretinal hemorrhages. Fluorescein angiography images revealed blockage of the macular lesion with peri-lesion leakage and diffuse vascular leakage. All imaging for the left eye was unremarkable.

There was high suspicion for bacterial or fungal infection, and the patient underwent immediate empiric intravitreal injections of vancomycin 1 mg/0.1 cc, ceftazidime 2.25 mg/0.1 cc, and voriconazole 50 µg/0.1 cc. A vitreous tap was sent for bacterial and fungal cultures. Further serologic testing was performed, which demonstrated positive serum syphilis IgG/IgM, negative rapid plasma reagin, and positive *Treponema pallidum* particle agglutination. Blood cultures, fungal cultures, and all further workup for other active infectious or inflammatory conditions were negative or within normal limits. The patient was admitted the following day and started on intravenous aqueous crystalline penicillin G 24 million units daily for 10 days, followed by 2 weekly intramuscular

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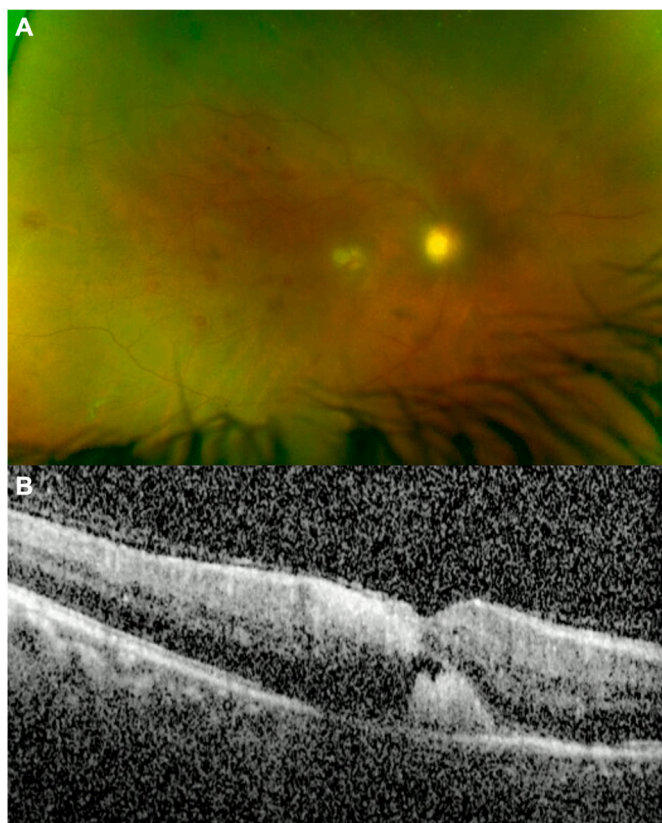


Fig. 1. A. Optos ultra-widefield imaging of right eye at presentation demonstrating vitritis, central white macular lesion, and scattered intraretinal hemorrhages. B. SD-OCT of the right eye at presentation demonstrating hyperreflective subretinal material at the fovea with overlying subretinal fluid. The OCT quality reflects the presence of anterior and posterior inflammation.

penicillin G benzathine 2.4 million units as an outpatient. Intraocular inflammation was treated with topical prednisolone acetate 1%. Periocular, intraocular, and systemic corticosteroids were avoided at presentation due to high suspicion of unknown infection.

Within two days of presentation, the anterior chamber inflammation and vitritis began to improve, and the foveal subretinal lesion began decreasing in size. However, by day 5, the patient was noted to have developed a full-thickness macular hole with a mild epiretinal membrane (Fig. 2A). The intraocular inflammation quickly resolved, and the topical steroids were tapered; however, the macular hole continued to enlarge, and by 3 months, the patient was noted to have developed a posterior vitreous detachment (Fig. 2B). At this time surgical intervention was discussed with the patient, who agreed to proceed with 25-gauge pars plana vitrectomy with indocyanine green dye assisted internal limiting membrane peel and C3F8 gas tamponade.

At post-operative month 1, the macular hole was closed with residual subretinal fluid (Fig. 3A). Vision at this time remained count fingers. The subretinal fluid continued to improve with complete resolution by post-operative month 6 (Fig. 3B). This result was maintained through post-operative month 18 (Fig. 3C), at which time visual acuity had improved to 20/200.

3. Discussion

Inflammatory macular holes are an uncommon sequelae of uveitis and have been associated with several etiologies, including Behçet's disease, Bartonella henselae infection, ocular toxoplasmosis, Vogt-Koyanagi-Harada syndrome, and idiopathic intraocular inflammation, among others.² Although a few macular holes secondary to ocular

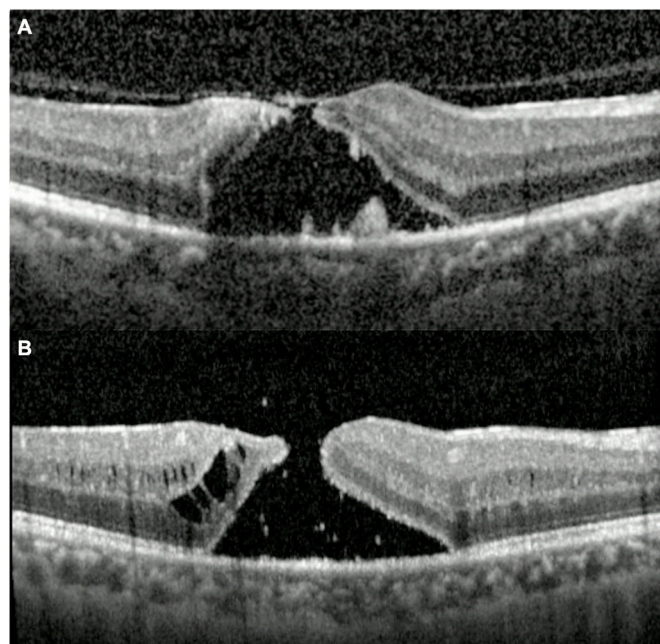


Fig. 2. A. SD-OCT of the right eye 5 days after presentation demonstrating full-thickness macular hole with near resolution of the subretinal lesion. B. SD-OCT of the right eye 3 months after presentation demonstrating full-thickness macular hole with new posterior vitreous detachment, further resolution of the subretinal lesion and improved vitritis.

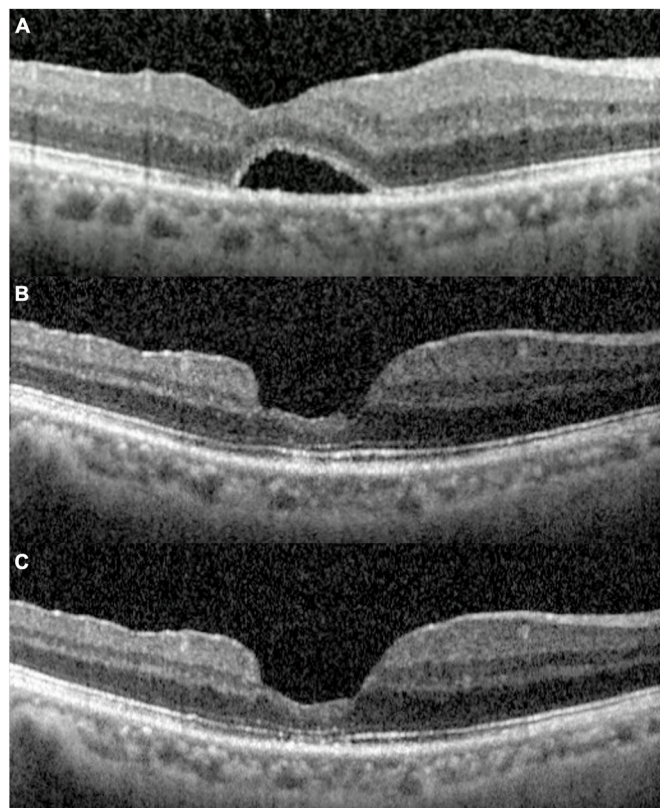


Fig. 3. SD-OCT of the right eye at A. post-operative month 1, B. post-operative month 6, and C. post-operative month 18.

syphilis have been reported in literature, there is only one published report of a macular hole secondary to ocular syphilis with OCT and outcome data.^{3–5} In that case, however, the patient did not have a PVD and showed subtotal closure of the hole without surgical intervention after 2 years of follow up, with improvement in visual acuity from count fingers to 20/40.

Inflammatory macular holes are thought to develop due to several factors. Inflammation induced changes to the vitreous body contributes to posterior vitreous detachment and antero-posterior traction. The development of an epiretinal membrane can also cause tangential traction. These forces, in combination with the fragilization of retinal tissue due to focal chorioretinitis or macular edema can produce a macular hole. While there are case reports of spontaneous closure with the resolution of inflammation and release of vitreous traction, patients may require surgical intervention.^{6–8} Outcomes data from a retrospective review of patients with macular holes secondary to Behçet's disease in 2011 demonstrated poor closure rates (1/6 eyes) and no improvement in vision with surgical intervention; however, more recent data suggests a role for surgery.⁹ Callaway et al. published a multicenter case series of 16 eyes in 15 patients that underwent pars plana vitrectomy with indocyanine green dye assisted internal limiting membrane peel for macular hole secondary to various inflammatory etiologies. Macular hole closure was reported in 81% of eyes and visual acuity improvement in 75%.¹⁰

Our patient presented with a foveal subretinal lesion that rapidly improved after intravitreal antibiotics, IV penicillin, and topical corticosteroids. However, he quickly developed a full-thickness macular hole likely secondary to initial inflammatory changes, a mass effect from the subretinal lesion, and treatment. The ocular inflammation may have contributed to the formation of an epiretinal membrane that exerted tangential traction on already thinned foveal retinal tissue from a mass effect caused by the subretinal lesion. Additionally, the intravitreal injection of 0.3 cc of antimicrobials may have caused rapid shifts in the vitreous body, thereby exerting vertical vitreomacular traction in the setting of an attached posterior hyaloid. These mechanical forces in conjunction with a fragilization of retinal tissue after rapid recovery of the subretinal lesion may all have contributed to the formation of the full-thickness macular hole on presentation day 5.

Due to the persistence of the macular hole despite the development of a PVD and good inflammatory control by 3 months, the decision was made to move forward with surgical repair. After pars plana vitrectomy with membrane peel and intraocular gas, the macular hole was closed, and vision improved to 20/200. This final vision was limited by retinal atrophy, which was likely due to the presenting subretinal lesion and additional stress on the retinal tissues from a rapid recovery, thereby preventing full recovery at the fovea.

It was interesting how quickly after intravitreal injection of ceftazidime, this patient was noted to have rapid resolution of the subretinal lesion, despite a delay in starting IV penicillin. A similar rapid clinical improvement in bilateral ocular syphilis was reported in a patient who received intravitreal ceftazidime injections in both eyes, while undergoing penicillin desensitization for penicillin allergy.¹¹ While definitive treatment for ocular syphilis remains intravenous aqueous crystalline penicillin G 18–24 MU daily for 10–14 days to maintain adequate treponemacidal levels in the cerebrospinal fluid, intravitreal ceftazidime may be considered an adjunct to promote early recovery. Further research is necessary to better understand the role for intravitreal

antibiotics in syphilitic uveitis.

4. Conclusions

In summary, this is the first reported case of a macular hole secondary to syphilis which necessitated surgical repair. In this case, favorable surgical and vision outcomes were achieved, although vision was ultimately limited by macular atrophy due to the foveal focus of the initial subretinal lesion.

Patient consent

Consent to publish the case report was not obtained. This report does not contain any personal information that could lead to the identification of the patient.

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Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Declaration of competing interest

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