was noted superiorly in the left eye and felt to be consistent with a laser burn. Follow-up examinations through 6 months of age confirmed resolution of the retinopathy with no change in the lenticular opacity.

At 7 months of age the patient developed conjunctival injection in his left eye. Examination showed moderate corneal haze, iritis, miosis, and the sectoral cataract superiorly. The intraocular pressure was 46 mm Hg and was unresponsive to medical management. A trabeculotomy with iridectomy and lysis of posterior synchiae with removal of an anterior chamber membrane was performed. A red reflex was noted inferiorly through the miotic pupil. During the procedure, a thin, milky-white fluid flowed from the iridectomy and out through the limbal incision. A C. albicans abscess was suspected and the white material was cultured.

The intraocular pressure normalized postoperatively but the cultures grew C. albicans on the second postoperative day. The patient was taken back to the operating room and examination after placement of iris hooks revealed a creamy-white lenticular opacity underlying the previous focal cataract. A cataract extraction with anterior vitrectomy and intravitreal injection of amphotericin B was performed. An infectious workup revealed a nidus in the patient’s right kidney, consistent with a Candida abscess, that resolved itself during a 28-day course of systemic fluconazole. The postoperative course was uneventful with treatment consisting of a Silsoft aphakic contact lens and part-time occlusion. He had a strong right fixation preference but was able to fix and follow with his left eye on his most recent follow-up examination at 13 months of age.

Comment. To our knowledge these are the fourth and fifth reported cases of a Candida intralenticular abscess with a white plaque in the pupill after treated candidemia in a premature, low-birth-weight infant. Similar to a previous case, our patients responded well to aggressive treatment that included cataract extraction, anterior vitrectomy, and intravitreal injection of amphotericin B. Case 1 is unique in that intralenticular opacities were noted in the affected eye at the time of initial candidemia. The opacities resolved with antifungal treatment. Of interest is the fact that all reported cases of endogenous intralenticular fungal abscess have occurred in premature infants. We believe that in such eyes, the persistent tunica vasculosa lenticis serves as the route for lenticular seeding by fungal organisms at the time of Candida septicemia. As the tunica regresses, Candida organisms become sequestered adjacent to and within the lens, resulting in inadequate treatment by systemic antifungal therapy. The lenticular abscess then slowly activates, appearing months later as a focal lens opacity accompanied at first by relatively little intraocular inflammation. Case 1 represents the first infant treated aggressively who developed good visual function and cosmesis. Case 2 is the first case to present as glaucoma.

Our cases and review of the literature suggest that premature neonates who have been successfully treated for systemic candidiasis remain at risk for developing sequestration of fungal organisms within the lens, which later appears as a focal lens opacity that evolves into a white papillary plaque up to 6 months following treatment. Close ophthalmologic follow-up and parental education regarding this presentation is recommended for this group of patients. As demonstrated by these cases, a high index of suspicion and aggressive therapy can result in an excellent clinical outcome.

The authors have no relevant financial interest in this article.

Correspondence: Dr Singh-Parikshak, Simpson Eye Associates, 650 Springhill Ring Rd, West Dundee, IL 60118 (ritasing10@hotmail.com).


Vitreous Opacities and Retinal Vascular Abnormalities in Gaucher Disease

Gaucher disease is an autosomal recessive lipid-storage disease. Deficiency in the enzyme glucosylceramidase, normally present in macrophage lysosomes, leads to accumulation of glucosylceramide in scavenger macrophages and subsequent deposition in the organs of the reticuloendothelial system (liver, spleen, and bone marrow). Enlarged macrophages with a foamy cytoplasm, likened to “crinkled tissue paper,” with an eccentrically placed nucleus (Gaucher cells) are abundant, leading to organomegaly with resultant pancytopenia. In the past, many affected pa-
Patients were treated with splenectomy at an early age to eliminate splenomegaly and resultant splenic sequestration of platelets. During the past decade, enzyme replacement therapy with intravenous infusions of glycosylceramidase has proven safe and effective for treating the visceral manifestations of Gaucher disease.\(^2\)

**Report of a Case.** A 20-year-old white woman had best-corrected visual acuities of 20/160 OD and 20/60 OS. There was a small-angle esotropia in the right eye. Although she had long complained of floaters, her visual acuity had been correctable to 20/25 OU until 2 years earlier. Results of anterior segment examination were normal. The posterior segment examination showed moderate to dense vitreous opacities in both eyes, overlying the optic nerve head, vessels, and vitreous (Figure 1). There was a conspicuous absence of inflammatory cells in the vitreous bilaterally. Fluorescein angiography showed normal dye transit, blockage of fluorescence due to the vitreal opacities, and marked vascular tortuosity (Figure 2).

The patient underwent standard 20-gauge pars plana vitrectomy in the right eye. A vitreous aspirate specimen obtained before infusion instillation was stained with hematoxylin-eosin (Figure 3). The material was paucicellular, with some degenerated and a few typical Gaucher cells. The entire vitrectomy specimen was sent for centrifugation and analyzed via thin-layer chromatography, which showed a large amount of glucosylceramide (Figure 4).

Postoperative photographs of the operated-on eye showed a few remaining preretinal white dots studding the retina and around the optic nerve head. A macular pucker was evident (Figure 5). Visual acuity was 20/40 at 6 months of follow-up.

**Comment.** Gaucher disease often occurs in Ashkenazi Jews but is panethnic.\(^2\) Type 1 Gaucher disease is characterized by an absence of central nervous system involvement, in contrast to types 2 and 3, which have primary central nervous system disease. Classic descriptions of this disease mention multiple ocular and neurologic associations, such as conjunctival pterygia, strabismus, and trismus with retroflexion of the neck. Vitreous opacities in this dis-

---

**Figure 1.** Fundus photographs of both eyes showing prominent vitreous opacities.

**Figure 2.** Fluorescein angiograms of both eyes in transit phase showing blockage of fluorescence caused by vitreal opacities, and vascular tortuosity.
order were first noted by Cogan et al3 and Gass.4 The incidence of vitreous opacities was found to be approximately 3% in a series of 80 consecutive patients with type 1 Gaucher disease.5 Only those who have undergone splenectomy have a tendency to form vitreous aggregates. Perhaps this is due to the occurrence of more circulating glucosylceramide in these patients, resulting in manifestations in

Figure 3. Vitreous aspirate showing a Gaucher cell (hematoxylin-eosin, original magnification ×250).

Figure 4. Thin-layer chromatography of lipids from vitreous and control lipids. Phospholipids were removed by alkaline methanolysis, dried under nitrogen gas, and resolved on borate-impregnated silica gel plates. Note intense staining of the glucosylceramide band in the patient.
unusual systemic locations, such as the eye. Our case is, to our knowledge, both the most severe that has been reported and the first noting retinal vascular tortuosity. The corkscrew vascular pattern is reminiscent of that in Fabry disease, a related disorder of sphingolipid metabolism. In our case, there were Gaucher cells in the vitreous cavity, while the vitreous gel contained large amounts of glucosylceramide.

The pathophysiologic mechanism of glucosylceramide deposition in the vitreous cavity is unclear. The material is a by-product of breakdown of myelin, leukocytes, red blood cells, and endothelial cells; myelin may be deposited within the eye when oligodendroglial cells migrate through the lamina cribrosa. Alternatively, deposition may occur by leakage from the vasculature. The latter is less likely, as glucosylceramide is not deposited within neurons in type 1 Gaucher disease, although Gaucher cells do accumulate in the periventricular macrosphages of the brain.¹

We have shown that vision may be improved with vitrectomy in patients with Gaucher disease. It is unknown whether vitreous opacities will recur in our patient, as has been reported in patients who have undergone vitrectomy for vitreous opacities in familial amyloidotic polyneuropathy syndrome.² The relationship of long-term enzyme replacement therapy to the vitreous opacities in our patient is speculative.

Eric M. Shrier, DO
Charles C. Barr, MD
Gregory A. Grabowski, MD

The authors have no relevant financial interest in this article.

This study was supported by the Kentucky Lions Eye Foundation, Louisville, and in part by a grant from Research to Prevent Blindness Inc, New York, NY.

Correspondence: Dr Barr, Department of Ophthalmology and Visual Sciences, University of Louisville School of Medicine, 301 E Muhammad Ali Blvd, Louisville, KY 40202 (ccbarr@pol.net).