usually treated with a combination of high-dose steroids and an immunosuppressive agent. Therapy with intravenous cyclophosphamide is the agent best studied for SLE. Intravenous immunoglobulin has also proven to be beneficial in selected patients. The role of methotrexate in the treatment of SLE remains unproven, but its safety record makes it an attractive alternative, especially for the control of some steroid-refractory manifestations of SLE.

Plasmapheresis could, by rapidly removing circulating immune complexes, provide acute relief in the severely ill patient in whom cyclophosphamide is not effective (case 1) or is undesirable (case 2). However, it could not have a lasting therapeutic effect unless it is combined with an immunosuppressive agent to retard the re-accumulation of immune complexes.

Although randomized controlled clinical trials failed to document a generalized benefit of plasmapheresis when added to standard immunosuppressive therapy, patients with SLE who are in crisis seem to benefit from the concomitant use of plasmapheresis with systemic prednisone and sequential intravenous cyclophosphamide.

To the best of our knowledge, this is the first report of patients with SLE retinal vasculitis who were successfully treated with a combination of plasmapheresis and immunosuppressive chemotherapy.

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Ocular Findings in a Patient With Hemophagocytic Syndrome

Hemophagocytic syndrome is a rare disease characterized by fevers, hepatosplenomegaly, and pancytopenia. Additionally, there is increased proliferation and activation of macrophages, with hemophagocytosis seen histologically in the liver, spleen, and bone. Only 3 case reports of retinal findings associated with hemophagocytic syndrome have appeared in the literature. Two reports describe retinal hemorrhages, disc edema, and perivenous white patches, and the other describes acute posterior multifocal placoid pigment epitheliopathy–like findings. We report a case of hemophagocytic syndrome with ocular findings that, to our knowledge, have not been previously described in the literature.

Report of a Case. A 31-year-old African American woman admitted for fevers, 18-kg weight loss, hepatosplenomegaly, and ascites over a 6-month period was referred to the Department of Ophthalmology, University of Chicago, Chicago, Ill, for bilateral visual blurring of 1 week’s duration. The patient was being treated with intravenous ceftazidime, fluconazole, and acyclovir for a suspected systemic infection. She denied symptoms of hair loss, headache, neck pain, tinnitus, or skin changes. Laboratory examination values were as follows: hemoglobin level, 93 g/L; white blood cell count, 1.7 × 10^9/L; platelet count, 15 × 10^9/L; lactate dehydrogenase level, 385 IU/L; and reticulocyte count, 5.7%. On ocular examination, her best-corrected visual acuity was 20/60 OD and 20/40 OS. Anterior segment examination results and intraocular pressures were normal. Funduscopic examination revealed multiple bilateral serous pigment epithelial detachments with macular edema. Fluorescein angiography showed corresponding pinpoint areas of leakage with late staining at the level of the pigment epithelium (Figure 1). Optical coherence tomography revealed a macular thickness of 845 μm OD and 439 μm OS. Consideration was given to treating the patient with systemic corticosteroids, although owing to concern of possible systemic infection, no treatment was instituted from the ocular standpoint.

A week later, the patient underwent a diagnostic and therapeutic splenectomy with hepatic and regional lymph node biopsies. Surgical pathological examination revealed a diagnosis of hemophagocytic syndrome exemplified by marked erythrophagocytosis and histiocytosis in the spleen, liver, and lymph nodes without clonal expansion of T or B cells (Figure 2). Results of a follow-up ocular examination performed 2 weeks after the initial consultation were unchanged.

After stabilization of her condition, the patient was discharged to a physical rehabilitation center with the intention to start treatment for hemophagocytic syndrome as an outpatient. However, she was soon readmitted to intensive care at a different hospital for recurrent fevers, worsening ascites, and gastrointestinal bleeding. The patient developed multiple organ failure and died a month later. An autopsy could not be arranged.

Comment. Hemophagocytic syndrome results from uncontrolled T-lymphocyte activation with hyperproduction of TNF proinflammatory cytokines, including interferon-γ, tumor necrosis factor α, interleukin 2, interleukin 10, and interleukin 12, causing macrophage activation.
Macrophage proliferation and activation result in histiocytic infiltration of the lymph node sinuses and medullary cords, liver sinusoids and portal areas, splenic red pulp, and bone marrow with a high degree of hemophagocytosis seen on histopathological examination.1

The ocular features in our patient closely resemble those in ocular Vogt-Koyanagi-Harada disease, an inflammatory condition characterized by bilateral multiple retinal pigment epithelial detachments thought to be caused by an autoim-

Figure 1. Color fundus photographs and fluorescein angiography transiting the right eye. In the right eye, there are diffuse macular edema and multiple serous pigment epithelial detachments in the posterior pole (A) as well as the nasal retina (B). C, Similar findings are seen in the left eye. Early (D) and late frames of the angiogram show pinpoint areas of leakage with eventual staining at the level of the retinal pigment epithelium with macular edema (E) in the right eye, involving the nasal retina as well (F). G, These findings are also seen in the left eye.
mune reaction to melanocytes. It is typically found in darkly pigmented women aged 30 to 39 years. It can also be associated with systemic features including meningismus, hearing changes, and skin manifestations.6,7

To our knowledge, this is the first case report of a patient with hemophagocytic syndrome with features resembling ocular Vogt-Koyanagi-Harada disease. It is possible that the hyperinflammatory state of hemophagocytic syndrome causes a uveal inflammatory reaction with histiocytic infiltration resulting in ocular features similar to ocular Vogt-Koyanagi-Harada disease. An alternative explanation would be that the patient had 2 separate disease processes, although this would seem to be less likely.

In summary, we describe another possible ophthalmic manifestation of hemophagocytic syndrome. This can be added to the previously described abnormalities resembling leukemic retinopathy and acute multifocal placoid pigment epitheliopathy.

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Stage 1 Macular Hole as a Complication of Laser Iridotomy

Laser peripheral iridotomy (LPI) is performed in patients with narrow anterior chamber angles at risk for angle-closure glaucoma. Argon or green diode and Nd:YAG lasers are often used sequentially in dark irides to create an iridotomy with a minimum amount of laser energy. Posterior segment complications of LPI are uncommon and generally related to direct laser-induced damage.1 Herein we report the development of a stage 1 macular hole following LPI and its