Surgical Drainage of Large Macular Cysts in Coats Disease

Consider this approach in cases of retinal edema after ineffective laser or pharmacologic therapies.

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Coats disease, a retinal vascular condition, is typically marked by unilateral retinal telangiectasias and abnormal microaneurysm formation without any known systemic disease. The management goals generally aim to preserve vision by limiting or eliminating telangiectatic vessels, leaking aneurysms, and subretinal exudates. However, advanced or refractory cases often result in poor outcomes. Peripheral lesions can often be managed with laser photocoagulation or retinal cryopexy, whereas macular pathology often requires laser treatments and intravitreal pharmacotherapies, such as anti-VEGF agents or steroids.

Recently, we encountered a young man with Coats disease who exhibited very large macular cysts and dense subretinal exudates that were previously refractory to both laser and intraocular injections. We decided to attempt surgical drainage of the retinal cysts.

THE CASE

A 30-year-old man with Coats disease in his left eye presented with worsening vision over several years. Diagnosed at age 9, he received multiple rounds of laser photocoagulation to the peripheral lesions. However, as a young adult, he began to develop macular cysts and exudates that were unsuccessfully managed, which led to his evaluation in our clinic.

At presentation, his VA was 20/400 in the affected eye with large macular cysts seen on fundus examination (Figure 1A). The cysts were located mostly in the superotemporal macula and dense exudates extending through the fovea. Fluorescein angiography (B) showed telangiectasias and large microaneurysms associated with the large cysts, as well as laser photocoagulation scars in the temporal macula from previous treatment.

Figure 1. The ultrawide fundus image (A) showed large cysts in the superotemporal macula and dense exudates extending through the fovea. Fluorescein angiography (B) showed telangiectasias and large microaneurysms associated with the large cysts, as well as laser photocoagulation scars in the temporal macula from previous treatment.
superotemporal macula and accompanied by retinal telangiectasias and light-bulb aneurysms, which leaked on fluorescein angiography (Figure 1B). Dense subretinal exudates extending through the fovea were seen on OCT imaging (Figure 2A), which accounted for the severity of his vision loss.

We began treatment with additional photocoagulation and multiple rounds of anti-VEGF therapy, but the intraretinal fluid did not abate. The vascular lesions showed no laser uptake despite increasing the energy and duration of the laser, likely due to the thick layer of retinal fluid insulating the retinal vasculature from the laser’s energy uptake at the level of the retinal pigment epithelium. His macular edema worsened despite aggressive treatment.

After extensive discussion about the risks, benefits, and alternatives, we decided to pursue pars plana vitrectomy with surgical drainage of the intraretinal cysts and laser photocoagulation.

THE SURGICAL DRAINAGE

Using 23-gauge instrumentation, we performed a standard three-port pars plana vitrectomy. Initially, we intended to use a 41-gauge subretinal cannula to perform the drainage with intraoperative OCT guidance. However, the tip of the 41-gauge cannula was clogged by the thick, viscous contents of the chronic cysts, preventing us from effectively aspirating the fluid. We switched to endodiathermy, and created a small retinotomy that allowed for aspiration using a 25-gauge soft-tip cannula. After fluid-air exchange, we aspirated the large macular cysts, with visible extrusion of the yellowish exudates through the retinotomy. This enabled us to perform endolaser to ablate the aneurysms and vascular lesions.

We left the patient with 25% SF₆ gas and noted no residual fluid at the 1-month postoperative visit. Over the subsequent year, the exudates gradually receded (Figure 2B), although his VA was 20/200.

THE RESULTS

Our case demonstrates a proof of concept of the possibility of surgically draining large macular cysts in cases that are refractory to standard laser photocoagulation or intravitreal therapies. Although the patient did not regain most of his vision, we attributed this to the prolonged duration of the subretinal exudates that were inadequately treated, resulting in photoreceptor degeneration and loss of the outer retinal layers, as seen on the postoperative OCT scans. Had he undergone surgical management at an earlier time, more of his vision might have been recovered, evidenced by the rapid resolution of the retinal fluid and dense exudates.

Generally, we believe that surgical intervention, especially in a young patient with a formed vitreous, should be left as a last-resort intervention. However, this case suggests that surgically draining large retinal cysts could be considered in select cases of retinal edema for which laser or pharmacologic therapies are ineffective.

Our experience also taught us that chronic intraretinal fluid, as with subretinal fluid in longstanding retinal detachments, can be challenging to remove and requires the use of a drainage retinotomy. We chose a slightly more expansile concentration of a short-acting gas to tamponade and compress the retinal tissue, although the effectiveness of such an approach is unclear.

We hope that sharing this unique case of surgically draining macular cysts in a patient with Coats disease helps to guide other vitreoretinal surgeons who are considering this approach in clinically appropriate cases.