

Uncoating Coats

Management of advanced disease may include surgery.

BY MANISH NAGPAL, MS, DO, FRCS(UK); AND PRAVIN JAIN, FRF, DNB

Coats disease is a developmental retinal vascular anomaly characterized by leaking telangiectatic and aneurysmal retinal vessels associated with hemorrhage, subretinal lipid exudation, and exudative retinal detachment without appreciable vitreo-retinal traction. It commonly presents unilaterally with strong male preponderance. Coats disease has reportedly occurred in patients ranging from 4 months to 70 years of age, with the peak incidence occurring toward the end of the first decade of life in otherwise healthy boys. George Coats, a Scottish ophthalmologist, first reported the association of retinal telangiectasis with hemorrhage, exudates, and retinal detachment in 1907.¹ He described these abnormal telangiectatic vessels as nonfamilial, dilated, and incompetent.

PRESENTATION AND DIAGNOSIS

Initially described as separate entities, the milder (Leber miliary aneurysms) and more severe (Coats disease) forms of this retinal vascular anomaly are now considered by most authorities to be variable expressions of the same disease and are therefore currently grouped under the common name of Coats disease. It is one of the important differential diagnoses of childhood leukocoria, and differentiating it from some forms of ocular malignancy can be difficult.

A common presentation is leukocoria with severely reduced vision, but this may go unnoticed because of a normally functioning fellow eye. When the macula is involved, fixation is lost, and strabismus is therefore a common finding. Fundus examination reveals retinal vascular abnormalities, most commonly the retinal telangiectasias that are limited to the arteriolar circulation of only one quadrant initially. Minor vascular leakage can be cleared by the pumping action of retinal pigment epithelium, leaving yellow exudate to resolve slowly. In the late stages, however, massive leakage from these abnormal vessels can result in a total bullous exudative retinal detachment. This subretinal fluid has

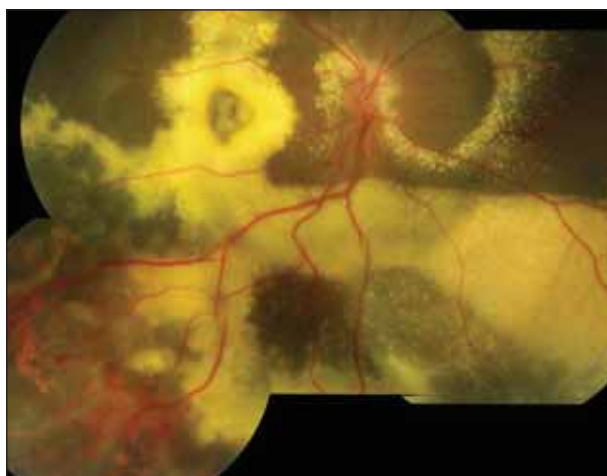


Figure 1. Extensive subretinal exudates with telangiectatic vessels are seen in the right eye with submacular scarred membrane and inferior retinal detachment.

a higher specific gravity than vitreous, and it causes a shifting retinal detachment.²

Its natural history is variable but typically progressive and can ultimately result in development of scarred, nonviable retina and subretinal nodules. Visual prognosis, if untreated at earlier stages, is always guarded because of development of stimulus-deprivation amblyopia and of secondary complications, such as neovascular glaucoma, which often require enucleation or evisceration, or phthisis bulbi.

TREATMENT

The goal of treatment is to control exudation by the pathologic retinal vessels and to prevent further progression to visually threatening complications. In early stages, this can be achieved by retinal ablative procedures such as laser photocoagulation and transscleral cryopexy. It is important to treat the entire area of abnormal vessels. Fluorescein angiography may aid in visualizing the full extent of vascular disease. Multiple

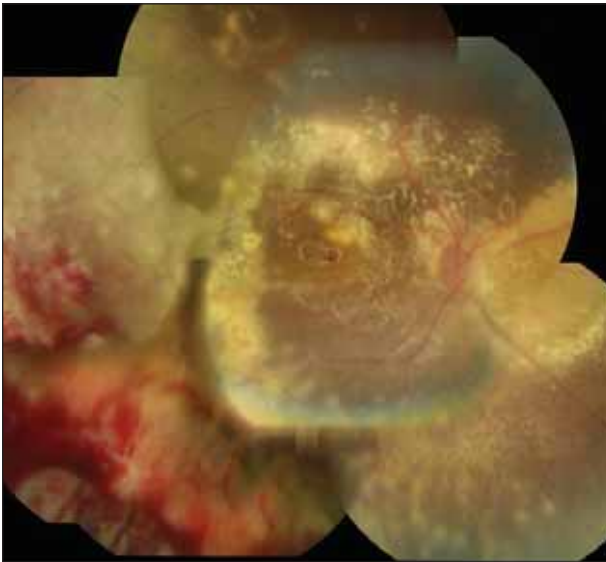


Figure 2. Right eye postoperative week 1. The macula is substantially clear of exudates and scarred membrane.

treatment sessions may be necessary to assure complete vascular closure and a confluent ablation of the ischemic retina. Regression of the lipid deposits may take weeks to months, even after successful coagulation therapy. In areas of thick exudation, there may be inadequate absorption of laser energy; in such areas, the triple freeze-thaw method of transscleral retinal cryopexy may be more effective. Careful follow-up is required to document regression of the abnormal vessels, as well as to detect new areas of telangiectasis.^{2,3}

Intravitreal anti-vascular endothelial growth factor (anti-VEGF) agents have recently been used in treating Coats disease.⁴ We have attempted combination of intravitreal bevacizumab and ranibizumab (Avastin and Lucentis, Genentech, Inc.) in addition to laser ablation in a small number of cases, resulting in reduction of exudation and improvement in visual acuity.

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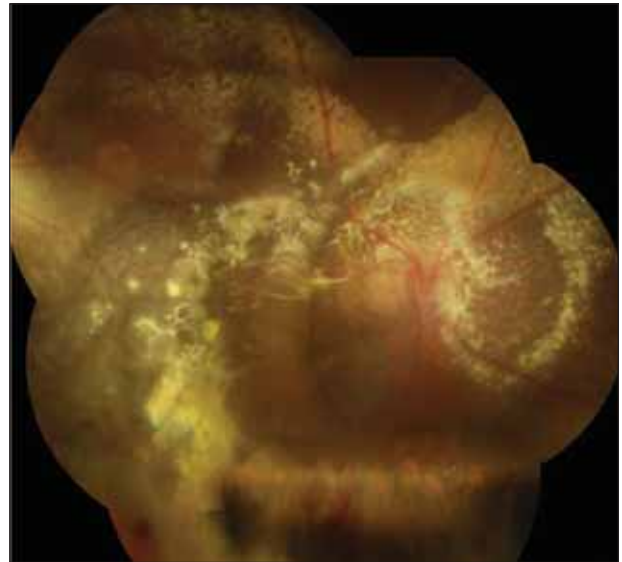


Figure 3. Right eye postoperative week 6. The exudates and telangiectatic vessels have resolved significantly.

In advanced Coats disease associated with exudative retinal detachment, cryotherapy or laser alone are less effective.² These may require surgical drainage of subretinal fluid and exudate to allow treatment of abnormal retinal vessels with either photocoagulation or cryopexy. This has been attempted either externally with or without placement of a scleral buckle, or internally in conjunction with pars plana vitrectomy with or without encircling buckle, and endodiathermy to leaking telangiectasias followed by tamponade with intravitreal gas or silicone oil.^{5,6}

CASE REPORT

Here we discuss a case in which we planned a vitrectomy to remove subretinal lipid exudates and preexisting scars.

Presentation. An 8-year-old boy first presented to our office for evaluation of poor visual acuity and strabismus in his right eye of 3 to 4 months duration as noted by his parents. Medical history and family history were unremarkable. Visual acuity was hand movements in the right eye and 20/20 in the left eye. Slit-lamp examination revealed quiet anterior segments in both eyes. Indirect ophthalmoscopy of the right eye revealed multiple telangiectatic vessels in the temporal retina and extensive subretinal lipid exudation involving the macula with submacular scarred membrane. A lesser amount of exudation was seen in the nasal retina. An exudative retinal detachment extended from 3 o'clock to 10 o'clock peripherally (Figure 1). The left fundus was normal. A clinical diagnosis of Coats disease was made.

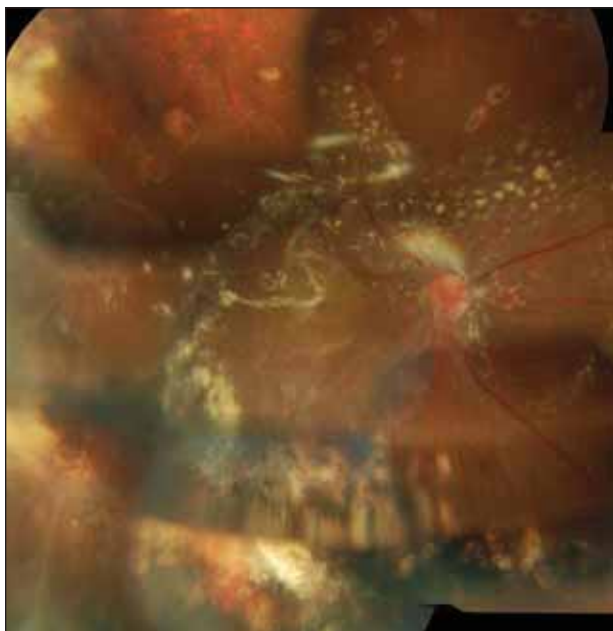


Figure 4. Right eye postoperative year 1. The retina is reattached, and the exudates and regressed telangiectatic vessels are completely resolved.

Surgical Intervention. We scheduled the patient for surgery. A 2.5-mm silicone band encirclage was placed at the equator to aid in extensive base excision of vitreous and to support the peripheral retina. This was followed by 23-gauge, three-port pars plana vitrectomy. Complete removal of the posterior hyaloid face (which was firmly adherent to the retina) was done after staining with 0.1 cc triamcinolone acetonide and inducing posterior vitreous detachment using high suction. At this stage, perfluorocarbon liquid (PFCL) was injected to stabilize the posterior pole. Endodiathermy was applied to large vessels in the temporal periphery to prevent bleeding.

A retinectomy was made in the periphery on the buckle from 7 to 11 o'clock using the cutter. At this stage, some of the heavy liquid was aspirated, and the temporal retinal flap was everted gently with forceps. Some of the exudates were released into the surrounding fluids and were aspirated. A bulk of thick exudates was seen partly on the undersurface of retina and partly on the underlying choroid. Active suction with a cutter was used to aspirate these exudates. The exudates on the undersurface of the retina were strongly adherent.

The retinectomy was extended a bit further inferiorly (2 clock hours), to allow more eversion of the flap to get access to the macular area. PFCL was further injected in the subretinal space to keep the flap retracted and to disperse the sticky exudates from the choroid. At this stage the submacular scar was visible, and this was gently sepa-

rated from its surrounding adhesions and removed with intraocular forceps. After aspirating the exudates, PFCL was injected to flatten the retina. Endolaser was carried out to barrage the retinectomy as well as to coagulate the telangiectatic vessels. This was followed by air-fluid exchange and injection of silicone oil.

Follow-up. Postoperative follow-up at weeks 1 and 6 revealed a gradual regression of the overall activity of abnormal vessels and clearing of residual macular exudates (Figures 2 and 3). At 1 year, the retina was attached with almost complete resolution of exudates and telangiectasis and with improvement in vision to counting fingers at 1 foot (Figure 4). At this time, we advised that the silicone oil should be removed.

CONCLUSION

Although visual function may be severely limited, particularly in such cases of advanced Coats, these surgical maneuvers may allow faster regression and flattening of the macula, thereby stabilizing the eye and preventing the subsequent development of phthisis, painful neovascular glaucoma, and the need for enucleation.

This extensive procedure, however, should be limited to only those cases which have extensive exudation in which external laser or cryotherapy are less effective. In this case, there was a subretinal scarred membrane that could also be removed during the surgery. ■

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