Denouement and Comment

Anterior Chamber Cholesterolosis in Coats Disease

The patient was diagnosed with anterior chamber cholesterolosis secondary to Coats disease. Coats disease is an idiopathic sporadic condition typified by retinal vascular telangiectasia and subretinal lipid exudation, which can lead to secondary retinal detachment. It occurs more commonly in males (76%) and is almost always unilateral (95%).[1] Coats disease can occur at any age but the majority of cases are diagnosed in the first 2 decades of life.[1] There are no documented systemic associations with classic Coats disease. In the Figure, A and B, the retina is detached and can be seen through the pupil just posterior to the lens. Inferiorly, the characteristic vascular telangiectasia is visible. In this case, the diagnosis of Coats disease was made at the initial presentation based on the presence of vascular telangiectasia and the absence of intraocular calcification.

Cholesterolosis is believed to result from the breakdown and crystallization of intraocular red blood cells as well as transvascular exudation of lipids. Anterior segment cholesterolosis has been documented after severe trauma, chronic inflammation, and chronic retinal detachment.[2] It can also occur as a result of hyphema or vitreous hemorrhage, neither of which were found in our patient.[3] In a large series of 158 eyes with Coats disease, only 3% had anterior segment cholesterolosis.[1] In Coats disease, the subretinal exudation characteristically contains lipid-laden macrophages and cholesterol clefts.[1] The exudation can escape into the vitreous and anterior chamber by either transudation or through undetected retinal holes, where it is seen as small, freely mobile, yellow refractile crystals (Figure, C and D)[4,5]

Although Coats disease is relatively rare, it is the most common condition in the differential diagnosis of suspected retinoblastoma.[6] The presenting signs of Coats disease typically include decreased visual acuity (43%), strabismus (23%), and leukocoria (20%).[1] Treatment is determined by the stage and severity of the disease. Laser photocoagulation and cryotherapy are used to ablate peripheral telangiectasias associated with exudative retinal detachment in an effort to minimize foveal involvement and maximize visual acuity. Patients with mild disease and minimal exudation are observed and patients with markedly advanced disease typically show visual acuity loss despite successful therapy.

Intraocular bevacizumab has been used off label in adults for numerous neovascular and exudative retinal diseases. Abnormal retinal blood vessels in Coats disease promote ischemia and leakage of lipid and protein through breakdown of the blood-retina barrier. This is thought to be mediated in part by vascular endothelial growth factor A. Bevacizumab is a monoclonal antibody that blocks vascular endothelial growth factor A and preliminary reports suggest that it may be useful in reducing exudation in children with Coats disease.[7] If untreated, Coats disease can lead to neovascular glaucoma and a blind, painful eye requiring enucleation (16%).[8] Visual prognosis is poor and limited by complications including retinal detachment, subfoveal exudation, and amblyopia, with more than half of patients demonstrating visual acuity of 20/200 or worse.[8]

Coats disease is only one of a number of causes of anterior segment cholesterolosis. Clinicians who see this condition should also evaluate the patient for trauma, ocular inflammation, and other causes of retinal detachment including retinopathy of prematurity and retinoblastoma.

Accepted for Publication: March 10, 2011.
Correspondence: Carol L. Shields, MD, Ocular Oncology Service, Wills Eye Institute, 840 Walnut St, Ste 1440, Philadelphia, PA 19107 (carol.shields@shieldsoncology.com).
Author Contributions: *Acquisition of data:* Patel and Murphy. *Analysis and interpretation of data:* Patel, Murphy, and Shields. *Drafting of the manuscript:* Patel and Murphy. *Critical revision of the manuscript for important intellectual content:* Shields. *Study supervision:* Murphy and Shields.
Financial Disclosure: None reported.
Funding/Support: Support was provided by the Retina Research Foundation of the Retina Society, Cape Town, South Africa (Dr Shields), and the Eye Tumor Research Foundation, Philadelphia, Pennsylvania (Dr Shields).

REFERENCES

1. Shields JA, Shields CL, Honavar SG, Demirci H. Clinical variations and complications of Coats disease in 150 cases: the 2000 Sanford Gifford Memorial Lecture. *Am J Ophthalmol.* 2001;131(5):561-571.
2. Eagle RC Jr, Yanoff M. Cholesterolosis of the anterior chamber. *Albrecht Von Graefes Arch Klin Exp Ophthalmol.* 1975;193(2):121-134.
3. Hemady R, Foster CS. Anterior chamber crystals following hyphema. *Arch Ophthalmol.* 1990;108(1):14-15.
4. Patel HK, Augsburger JJ, Eagle RC Jr. Unusual presentation of advanced Coats' disease. *J Pediatr Ophthalmol Strabismus.* 1995;32(2):120-122.
5. Shields JA, Eagle RC Jr, Fammartino J, Shields CL, De Potter P. Coats' disease as a cause of anterior chamber cholesterolosis. *Arch Ophthalmol.* 1995;113(8):975-977.
6. Shields JA, Shields CL. *Intraocular Tumors: An Atlas and Text.* Philadelphia, PA: Lippincott Williams & Wilkins; 2007.
7. Lin CJ, Hwang JF, Chen YT, Chen SN. The effect of intravitreal bevacizumab in the treatment of Coats disease in children. *Retina.* 2010;30(4):617-622.
8. Shields JA, Shields CL, Honavar SG, Demirci H, Cater J. Classification and management of Coats disease: the 2000 Proctor Lecture. *Am J Ophthalmol.* 2001;131(5):572-583.