

Coats Disease with Gliotic Nodule Simulating Retinoblastoma

BY A'SHA BROWN, BS; SWATHI KALIKI, MD; AND CAROL L. SHIELDS, MD

Coats disease, first described by Coats of Scotland in 1908,¹ is an idiopathic congenital condition manifesting with retinal telangiectasia, intraretinal and/or subretinal exudation and fluid, and without appreciable vitreoretinal traction.² In a review of 150 cases of Coats disease by Shields et al,² the median age of presentation was 5 years, male predilection (76%) was confirmed, and the manifestations were unilateral (95%). The hallmark fundus features of Coats disease include irregular caliber vessels, peripheral retinal telangiectasia, aneurysmal dilatation (appearing like "lightbulbs"), and intraretinal/subretinal exudation.² Retinal telangiectasia is present in 100% of cases, involving midperipheral or peripheral fundus in 99% of cases

and restricted to macula in 1%.² Intraretinal exudation is evident in 99% of cases, and 81% of cases develop exudative retinal detachment.² Based on the extent of retinal telangiectasia, foveal/extrafoveal location of subretinal/intraretinal exudation, subtotal/total retinal detachment, and secondary ocular complications, Shields et al³ classified Coats disease into 5 stages (Table 1).

In this condition, the macula is commonly involved with accumulation of exudation remote from the main areas of telangiectasia.^{2,4} Long-standing macular exudation can result in macular fibrosis and the formation of subfoveal gliotic nodule, occasionally resembling retinoblastoma.^{5,6} Herein, we present a case of Coats disease with subfoveal gliotic nodule simulating retinoblastoma.

TABLE 1. CLASSIFICATION OF COATS DISEASE

Staging	Feature	Treatment
Stage 1	Retinal telangiectasia only	Observation or laser photocoagulation
Stage 2 Stage 2A Stage 2B	Retinal telangiectasia with exudation Exudation in the extrafoveal region Exudation in the foveal region	Cryotherapy or laser photocoagulation
Stage 3 Stage 3A1 Stage 3A2 Stage 3B	Presence of exudative retinal detachment Subtotal extrafoveal exudative retinal detachment Subtotal foveal exudative retinal detachment Total exudative retinal detachment	Intravitreal triamcinolone with cryotherapy or laser photocoagulation
Stage 4	Total retinal detachment with glaucoma	Enucleation or intravitreal triamcinolone with cryotherapy or laser photocoagulation
Stage 5	Total retinal detachment with cataract and/or phthisis bulbi	Enucleation or observation

Adapted from: Shields JA, Shields CL, Honavar SG, Demirci H, Cater J. Classification and management of Coats disease: the 2000 Proctor Lecture. *Am J Ophthalmol.* 2001;131:572-583.

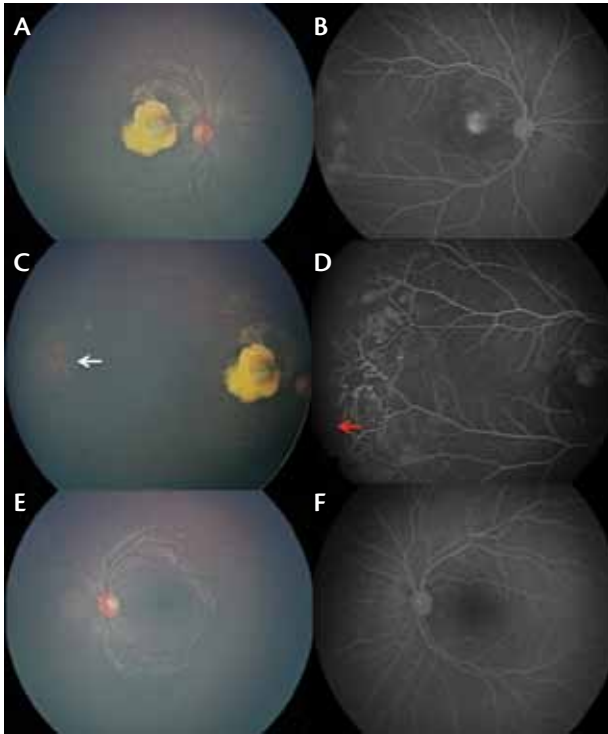


Figure 1. Coats disease with subfoveal gliotic nodule in a 5-year-old black male. In the macular regions there was a circumscribed white nodule surrounded by yellow exudation. The nodule measured 2 mm in diameter (A). Fluorescein angiography shows the gliotic nodule with hyperfluorescence and the exudation with mild blockage hypofluorescence (B). Peripheral retinal telangiectasia is seen temporally (white arrow) and superiorly (C). Fluorescein angiography confirms peripheral telangiectasia and non-perfusion (red arrow; D). The left fundus was normal clinically (E) and by fluorescein angiography (F).

CASE

A 5-year-old black male was noted at a school vision screening to have decreased vision in the right eye (OD). The patient was referred to Ocular Oncology Service, Wills Eye Institute, for further examination. On examination, visual acuity was counting fingers at 3 feet OD and 20/20 in the left eye (OS). The left eye was otherwise unremarkable. The intraocular pressure was normal in both eyes.

Fundus examination OD revealed a distinct gliotic nodule measuring 2.0 mm in the foveal region, with surrounding retinal exudation (Figure 1). Peripheral fundus examination disclosed peripheral intraretinal exudation and retinal telangiectasia involving 5 clock hours temporally and superiorly, accompanied by light bulb aneurysms. There was no clinical evidence of retinal dragging, retinal detachment, or hemorrhage. The findings of

Important differentiating characteristics of Coats disease vs retinoblastoma include age at detection, fundus details, arrangement of retinal vessels, USG findings, and FA features.

peripheral retinal telangiectasia and nonperfusion OD were confirmed on fluorescein angiography (FA). B-scan ultrasonography (USG) OD confirmed a solid echodense mass in the fovea with absence of calcification or retinal detachment and measuring 2.5-mm thickness. The clinical and imaging studies were consistent with Coats disease stage 3B. Cryotherapy was performed in the areas of retinal telangiectasia and the retinal detachment slowly resolved, leaving flat retina with subretinal fibrosis and subfoveolar gliosis.

DISCUSSION

Coats disease is the most common simulator of retinoblastoma.⁷ In a review of 2775 patients referred for retinoblastoma by Shields et al, 22% of cases had simulating lesions (pseudoretinoblastoma).⁷ Simulating lesions differed based on age at presentation, and the leading pseudoretinoblastomas in children younger than 1 year were persistent fetal vasculature (PFV; 49%), Coats disease (20%), or vitreous hemorrhage (7%); in those 2 to 5 years old were Coats disease (61%), toxocariasis (8%), or PFV (7%); and in those older than 5 years were Coats (57%), toxocariasis (8%), or familial exudative vitreoretinopathy (6%).⁷ Thus, Coats disease is an important simulating lesion of retinoblastoma, and the development of foveolar gliotic nodule in Coats disease further resembles the nodular tumors of retinoblastoma.

Important differentiating characteristics of Coats disease vs retinoblastoma include age at detection, fundus details, arrangement of retinal vessels, USG findings, and FA features.^{8,9} Patients with Coats disease are typically older (mean age, 5 years) than retinoblastoma cases (mean age, 1.5 years). The anterior segment is clear in Coats disease unless cholesterosis occurs with free-floating yellow crystalline cholesterol deposits. By contrast, retinoblastoma can display anterior segment tumor seeds appearing white and fluffy.¹⁰ Posterior segment features are critical in the differentiation of Coats disease and retinoblastoma. In Coats disease, the vitreous is clear or relatively murky from exudation compared to retinoblastoma, in which vitreous seeding can be detected. The retinal features are most dis-

tinctly different in these 2 conditions. In Coats disease, irregularly dilated blood vessels with visible microaneurysms and macroaneurysms (light bulbs) surrounded by exudation are found compared. In retinoblastoma, regularly dilated feeding blood vessels are uniform and disappear into the tumor without surrounding exudation. The vascular findings are confirmed on FA. Coats disease manifests distinctly irregular vessels on FA, and there is additional peripheral nonperfusion in most cases, not found with retinoblastoma. B-scan USG of Coats disease confirms retinal detachment with slightly echogenic, freely shifting subretinal fluid and no tumor or calcification, contrasted to retinoblastoma, which depicts an echodense calcified solid mass often with retinal detachment.^{8,9} Despite these numerous differentiating features, considerable overlap can occur.

Coats disease can lead to blindness. Most often this is from macular exudation, persistent subretinal fluid, or secondary glaucoma. Other less recognized features of Coats disease include retinal macrocyst and subfoveal gliotic nodule. The gliotic nodule can be mistaken for retinoblastoma and, when chronic, can assume a small feeder vessel and develop fibrosis or even ossification, further confusing the differentiation.¹¹ Unlike retinoblastoma, the gliotic nodule in Coats disease displays related exudation and telangiectasia. Foveal retinoblastoma generally shows no evidence of exudation.

The subfoveal gliotic nodule of Coats disease represents a secondary reaction to chronic retinal injury from the retinal vascular leakage. This feature is not specific to Coats disease and can be found with other chronically leaking retinal conditions.^{6,12} In a study of 150 patients with Coats disease by Shields et al,² retinal exudation involved the entire fundus in 75% of the cases, commonly affecting areas remote from the vascular abnormalities, particularly the macular region. In a subsequent study of 47 patients with Coats disease by Jumper et al,¹² subfoveal gliotic nodule was noted at presentation or developed during the course of follow-up in 11 patients (23%). There was FA evidence of intraretinal vascular anastomosis within the area of fibrosis in 7 of the 11 eyes (64%). Based on their results, Jumper et al¹² speculated that macular fibrosis in Coats disease could be the result of neovascularization in response to the accumulation of lipid exudation.

The primary goal of treatment in Coats disease is to preserve vision and salvage the eye. Management depends on disease staging, as listed in Table 1. The presence of subfoveal gliotic nodule is associated with poor visual prognosis despite successful treatment.^{2,12}

In summary, we present a child with Coats disease and subfoveal gliotic nodule simulating retinoblastoma.

The diagnosis of Coats disease was established based on clinical and FA findings of peripheral retinal telangiectasia with aneurysmal lightbulbs. Careful clinical examination, FA, and USG features aid in the differentiation of Coats disease from retinoblastoma. ■

Support provided by the Carlos G. Bianciotto Retinoblastoma Research Fund c/o the Eye Tumor Research Foundation, Philadelphia, PA (CLS, JAS), The Lucille Wiedman Fund for Pediatric Eye Cancer, Philadelphia, PA (CLS, JAS), Lift for a Cure, Morrisdale, PA (CLS), and Eye Tumor Research Foundation, Philadelphia, PA (CLS, JAS). The funders had no role in the design and conduct of the study, in the collection, analysis and interpretation of the data, or in the preparation, review or approval of the manuscript. Carol L. Shields, MD, has had full access to all the data in the study and takes responsibility for the integrity of the data and the accuracy of the data analysis.

The authors state that they have no financial relationships to disclose.

A'sha Brown, BS, is a fourth-year medical student at Drexel University College of Medicine in Philadelphia.

Swathi Kaliki, MD, is a former fellow with the Ocular Oncology Service, Wills Eye Institute, Thomas Jefferson University in Philadelphia. Dr. Kaliki is currently a Consultant at the Ocular Oncology Service at the LV Prasad Eye Institute in Hyderabad, India.

Carol L. Shields, MD, is the Co-Director of the Ocular Oncology Service, Wills Eye Institute, Thomas Jefferson University. She is a Retina Today Editorial Board member. Dr. Shields may be reached at +1 215 928 3105; fax: +1 215 928 1140; or at carol.shields@shieldsoncology.com.



1. Coats G. Forms of retinal diseases with massive exudation. *Roy Lond Ophthalmol Hosp Rep.* 1908;17:440-525.
2. 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;131:561-571.
3. Shields JA, Shields CL, Honavar SG, Demirci H, Cater J. Classification and management of Coats disease: the 2000 Proctor Lecture. *Am J Ophthalmol.* 2001;131:572-583.
4. Lee ST, Friedman SM, Rubin ML. Cystoid macular edema secondary to juxtafoveal telangiectasis in Coats' disease. *Ophthalmic Surg.* 1991;22:218-221.
5. Vezzola D, Mapelli C, Canton V, Viola F, Rattiglia R. Macular fibrosis in Coats disease. *Retina.* 2011;31:2136-2137.
6. Khurana RN, Samuel MA, Murphree AL, Loo RH, Tawansy KA. Subfoveal nodule in Coats' disease. *Clin Experiment Ophthalmol.* 2005;33:301-302.
7. Shields CL, Schoenberg E, Kocher K, Shukla SY, Kaliki S, Shields JA. Lesions simulating retinoblastoma (pseudoretinoblastoma) in 604 cases. Results based on age at presentation. *Ophthalmology.* 2013;120(2):311-316.
8. Shields JA, Shields CL. Differentiation of Coats' disease and retinoblastoma. *J Pediatr Ophthalmol Strabismus.* 2001;38:262-266.
9. Kaliki S, Shields CL. Differential diagnosis of retinoblastoma. In Ramasubramanian A, Shields CL, eds. *Retinoblastoma.* New Delhi, India: Jaypee Brothers Medical Publishers; 2012:46-60.
10. Patel AK, Murphy M, Shields CL. Picture of the month. Anterior chamber cholesterolosis in Coats disease. *Arch Pediatr Adolesc Med.* 2011;165:1131-1132.
11. Chang MM, McLean IW, Merritt JC. Coats' disease: a study of 62 histologically confirmed cases. *J Pediatr Ophthalmol Strabismus.* 1984;21:163-168.
12. Jumper JM, Pomerleau D, McDonald HR, Johnson RN, Fu AD, Cunningham ET. Macular fibrosis in Coats disease. *Retina.* 2010;30:59-514.