Coats Disease with Gliotic Nodule Simulating Retinoblastoma

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Coats disease, first described by Coats of Scotland in 1908, is an idiopathic congenital condition manifesting with retinal telangiectasia, intraretinal and/or subretinal exudation and fluid, and without appreciable vitreoretinal traction. In a review of 150 cases of Coats disease by Shields et al, 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. Retinal telangiectasia is present in 100% of cases, involving midperipheral or peripheral fundus in 99% of cases and restricted to macula in 1%. Intraretinal exudation is evident in 99% of cases, and 81% of cases develop exudative retinal detachment. Based on the extent of retinal telangiectasia, foveal/extrafoveal location of subretinal/intraretinal exudation, subtotal/total retinal detachment, and secondary ocular complications, Shields et al 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. Long-standing macular exudation can result in macular fibrosis and the formation of subfoveal gliotic nodule, occasionally resembling retinoblastoma. Herein, we present a case of Coats disease with subfoveal gliotic nodule simulating retinoblastoma.

<table>
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<th>Staging</th>
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<tr>
<td>Stage 1</td>
<td>Retinal telangiectasia only</td>
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<td>Stage 2</td>
<td>Retinal telangiectasia with exudation</td>
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<td>Stage 2A</td>
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<td>Stage 2B</td>
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<td>Stage 3</td>
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<td>Stage 3A1</td>
<td>Subtotal extrafoveal exudative retinal detachment</td>
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<td>Stage 3A2</td>
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<td>Stage 3B</td>
<td>Total exudative retinal detachment</td>
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<td>Stage 4</td>
<td>Total retinal detachment with glaucoma</td>
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<td>Stage 5</td>
<td>Total retinal detachment with cataract and/or phthisis bulbi</td>
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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 diameter 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 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 subfoveal gliosis.

**DISCUSSION**

Coats disease is the most common simulator of retinoblastoma. In a review of 2775 patients referred for retinoblastoma by Shields et al, 22% of cases had simulating lesions (pseudoretinoblastoma). Simulating lesions differed based on age at presentation, and the leading pseudoretinoblastomas in children younger that 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%). 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.

**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).
tinctly different in these 2 conditions. In Coats disease, irregularly dilated blood vessels with visible microaneu-
ysms 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.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.

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