Intraretinal Calcification and Osseous Metaplasia in Coats Disease

We report a case of Coats disease in a 9-year-old boy who had a profound visual deficit and increasing pain in his right eye. Funduscopic examination revealed a complex, exudative retinal detachment with a subretinal mass and peripheral retinal telangiectasis. B-scan ultrasonography revealed a heterogeneously echogenic subretinal mass with several highly reflective foci consistent with calcification. Enucleation was performed and confirmed the diagnosis of Coats disease. Histopathological examination revealed the heretofore unreported finding of intraretinal calcification.

In 1908, George Coats described a clinical entity characterized by telangiectasis, aneurysmal retinal vessels, and intraretinal and/or subretinal exudates in young males. In 1912, Leber reported a similar condition with telangiectasis and multiple retinal aneurysms but without the massive subretinal exudation found in Coats’ series. In 1956, Reese observed that patients with multiple miliary aneurysms progressed to massive subretinal exudation and proposed that Leber’s and Coats’ cases represented a continuum of a single pathologic process.

A recent review has further classified the criteria for Coats disease as idiopathic retinal telangiectasis, intraretinal and/or subretinal exudation, and frequent exudative retinal detachment without retinal or vitreous traction. Coats disease may be manifested at any age but is most often diagnosed between the ages of 5 and 11 years. It is typically unilateral, and approximately three fourths of the affected patients are male. The majority of patients have decreased vision, strabismus, or leukocoria.

Retinoblastoma is the most common primary intraocular malignancy in childhood and may be difficult to distinguish from Coats disease based on history and ophthalmologic examination. The presence of intraretinal calcification within a retinal mass strongly supports the diagnosis of retinoblastoma.

Herein, we present the clinico-pathologic correlation of a patient with Coats disease consisting of a complex, exudative retinal detachment and subretinal mass with retinal telangiectasis. Although the history and clinical examination supported the diagnosis of Coats disease, retinoblastoma could not be definitively excluded because of the detection of apparent foci of intraretinal calcification with ultrasonography.

Report of a Case. A 9-year-old boy arrived at the Bascom Palmer Eye Institute complaining of severe ocular pain in his right eye for 2 weeks and progressive visual loss during several months. His past medical history and family history were unremarkable. There was no history of ocular trauma.

On examination, the patient’s best corrected visual acuity was 2/200 in the right eye and 20/20 in the left eye. A relative afferent pupillary defect was present in the right eye. Anterior segment examination was unremarkable.

Dilated funduscopic examination of the right eye revealed a complex, exudative retinal detachment associated with multiple telangiectasias in the temporal retina. Extensive subretinal fluid and apparent foci of lipid deposition were present (Figure 1). No holes, tears, breaks, epiretinal membrane, or pro-

Figure 1. RetCam (Massie Research Laboratories Inc, Dublin, Calif) images of the right eye reveal exudative retinal detachment with extensive subretinal fluid and foci of calcification in the posterior pole (A) and, in the temporal periphery, telangiectatic vessels visible on the surface of a complex, exudative retinal detachment (B).
Proliferative vitreoretinopathy formation was identified. Examination of the left eye revealed no evidence of vascular anomaly, retinal mass, or tumor. Ultrasonography revealed a heterogeneously echogenic mass as well as highly reflective signals consistent with intraretinal calcification in the temporal periphery (Figure 2).

The clinical evaluation in this case was most consistent with long-standing Coats disease. However, retinoblastoma could not be definitively excluded given the detection of intraretinal calcification by ultrasonography. After a thorough discussion of the treatment options, the family elected to proceed with enucleation.

The enucleation specimen was sent to the Florida Lions Ocular Pathology Laboratory for evaluation. Gross pathologic examination revealed a yellow-white subretinal mass in the temporal retina with large, tortuous vessels on its surface (Figure 3). Total retinal detachment was present, extending from the ora serrata to the posterior pole with a large accumulation of subretinal fluid (Figure 3).

Microscopic examination revealed the classic histopathological components of Coats disease. A focus of retinal telangiectasis was identified along with cholesterol cleft formation and lipid-laden foamy macrophages (Figure 4). Moreover, features of long-standing Coats disease were revealed, including cystic retinal degeneration, a marked amount of subretinal fibrosis, and nodular proliferation of the retinal pigment epithelium. A focus of osseous metaplasia was present within the fibroretinal subretinal mass. Overlying the subretinal mass were several areas of intraretinal calcification (Figure 5). There was no evidence of retinoblastoma or tumor.

Comment. The histopathological findings in this case are consistent with long-standing Coats disease. It has previously been reported that chronic Coats disease may progress to total retinal detachment and the development of a subretinal fibro-osseous nodule. Moreover, calcification...
tion and osseous metaplasia may occur within a subretinal mass in Coats disease. To our knowledge, this is the first report of intraretinal calcification in Coats disease. Intraretinal calcification classically occurs in retinoblastoma; however, it may also be detected in retinocytoma, tuberous sclerosis, and epiretinal membranes.

This case reinforces the importance of a complete ophthalmic history, examination, and ancillary tests to distinguish Coats disease from retinoblastoma because intraretinal calcification may be present in Coats disease. Nearly one third of patients with Coats disease are referred to oculoncology specialists for presumptive retinoblastoma. Distinguishing Coats disease from retinoblastoma may be difficult given the overlap of symptoms in a pediatric population. Moreover, ophthalmological examination alone is often insufficient to establish a diagnosis. Ultrasonographic and computed tomographic scans are frequently used to further evaluate the posterior pole for masses, retinal detachment, and calcification.

Early diagnosis and treatment are critical in Coats disease. Patients with retinal telangiectasis and retinal exudates generally have good visual outcomes with laser photocoagulation. In cases of Coats disease with retinal detachment, the visual outcomes are generally poor. However, drainage of subretinal fluid, reattachment of the retina, and cryotherapy of retinal telangiectasis may halt the progression to neovascular glaucoma.

Enucleation is performed in approximately 16% of patients with Coats disease. Enucleation is typically indicated in blind, painful eyes secondary to neovascular glaucoma. Enucleation was performed in this case primarily because of profound visual deficit and pain with little potential for visual rehabilitation with surgical retinal reattachment. Moreover, the presence of intraretinal calcification limited our ability to definitively exclude retinoblastoma.

In conclusion, we present a case of Coats disease with extensive retinal pigment epithelium hypertrophy, osseous metaplasia, and intraretinal calcification. This is the first report of intraretinal calcification in Coats disease, and it emphasizes the challenge of distinguishing advanced Coats disease from retinoblastoma.

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Orbital Ganglioneuroma in a Patient With Chronic Progressive Proptosis

Ganglioneuromas are rare benign neoplasms of neuroblastic origin that have been reported in multiple anatomic locations. Orbital involvement is extremely rare, with only a single reported case of direct extension to the orbit from an adjacent parasinus. This is the first reported case of a ganglioneuroma arising in the orbit.

Report of a Case. A 12-year-old African American male had progressive proptosis of the right eye during a 1-year period (Figure 1). The child had a history of stage IV neuroblastoma 10 years earlier that had originated in the adrenal gland and was treated with 7 courses of cyclophosphamide and teniposide after induction with Adriamycin and cyclophosphamide. Neuroblastoma with metastases was confirmed by biopsy of the adrenal gland prior to induction of chemotherapy. No additional surgery was performed because of the advanced stage of the disease, and the...