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 ophthalmologic 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 ocul Oncology specialists for presumptive retinoblastoma. Distinguishing Coats disease from retinoblastoma may be difficult given the overlap of symptoms in a pediatric population. Moreover, ophthalmologist 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 photoagulation. 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 paranasus. 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

Figure 1. Clinical photograph prior to surgery showing proptosis of the right eye.

Figure 2. Axial view of an orbital T1-weighted magnetic resonance image revealing a superior orbital neoplasm.
patient responded to chemotherapy and remained in remission. His past medical history also included multiple cutaneous ganglioneuromas of the abdomen and gastrointestinal ganglioneuromas. He was profoundly mentally handicapped since infancy. Eye examination revealed no light perception in both eyes. He had 4 mm of proptosis of the right eye. His pupils were slowly reactive to light with no afferent pupillary defect. The cornea of the right eye evidenced diffuse punctate epithelial erosions and moderate conjunctival injection. Results of an anterior segment examination of the left eye were normal. Ductions were full in both eyes. Dilated funduscopic examination revealed optic nerve pallor with cup to disc ratio of 0.6 in both eyes. The macula, retina, and vitreous were normal in both eyes.

Magnetic resonance imaging revealed a lobulated neoplasm of the right superior orbit that molded around the globe (Figure 2 and Figure 3). The neoplasm was enhanced with contrast and appeared homogeneous in composition. There was no evidence of involvement of the bone. Orbital exploration via an anterior orbitotomy incision revealed a circumscribed lesion with a gray-tan surface without necrosis (Figure 4). Histopathologic examination of an incisional biopsy revealed neural tissue containing mature ganglion cells, and no neuroblastic elements were present. Because of progressive proptosis and corneal exposure, debulking was performed. Extensive histologic sampling revealed the neoplasm to be composed entirely of mature ganglioneuromatous tissue; again, no foci of neuroblastoma or sarcoma were detected (Figure 5). Immunohistochemical stains showed ganglion cells exhibiting strong cytoplasmic positivity for neuron-specific enolase and synaptophysin; only occasional ganglion cells demonstrated weak positivity for neurofilament and S-100 protein. The spindle cell component of the neoplasm demonstrated strong cytoplasmic positivity for S-100 protein, neurofilament, synaptophysin, and neuron-specific enolase. The immunohistochemical findings are consistent with ganglioneuroma.1

Comment. Neuroblastic neoplasms can be broadly subcategorized as neuroblastoma, ganglioneuroblastoma, or ganglioneuroma.2 Ganglioneuromas are benign neoplasms composed of neural elements, including mature ganglion cells. Grossly, ganglioneuromas appear circumscribed with a gray or tan cut surface having a whorled appearance on sectioning. In addition to its association with metastatic neuroblastoma, ganglioneuroma has been associated with neurofibromatosis type 13 and multiple endocrine neoplasia.4 Ganglioneuromas of the gastrointestinal tract are not uncommon in the neurofibromatosis. They also commonly arise from the sympathetic chain and can be found in the retroperitoneum or posterior mediastinum.5 A single orbital case has been documented that described local extension from the sinuses.6 Metastases do not occur; however, patients who have had metastatic neuroblastoma may develop ganglioneuromas that are thought to represent rests of neuroblastoma cells that have undergone differentiation to ganglioneuromas.7 These neoplasms are rare compared with other neurogenic lesions, such as neurofibroma and schwannoma. To our knowledge, ours represents the first case of ganglioneuroma arising in the orbit.

Ganglioneuromas may arise de novo or in patients who have had chemotherapy for metastatic neuroblastoma. Typically, they enlarge slowly; rapid growth should raise suspicion for a poorly differentiated neoplasm. Neoplasms of primitive neuroectodermal origin, such as neuroblastoma, contain pluripotent cells that have the capacity to differentiate into mature cellular elements, such as ganglion cells.8 When ganglion cell differentiation is present within an otherwise typical neu-
roblastoma, the term ganglioneuroblastoma is appropriate. In the present case, clinical uncertainty regarding cellular composition coupled with progressive proptosis and its associated corneal complication prompted debulking of the lesion. The history of metastatic neuroblastoma and the presence of multiple ganglioneuromas lend credence to the theory purporting the presence of rests of metastatic neuroblastoma that subsequently undergo maturation. Ganglioneuromas, as fully differentiated neoplasms, do not have the capability to metastasize, so extensive surgical resections or chemotherapy is not normally necessary, provided surgical sampling is sufficient to allow adequate histologic analysis and to assure no neuroblastic cellular elements are present. Excision may be considered when the pathologic diagnosis is uncertain or visual function is compromised by the neoplasm.

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Primary Epithelial-Myoepithelial Carcinoma of the Lacrimal Gland

Most lacrimal gland lesions are inflammatory or lymphoid neoplasms.1 Nonlymphoid neoplasms are less common, and most are primarily epithelial in origin.1,2 Among them, pleomorphic adenoma and adenoid cystic carcinoma are the most common benign and primary malignant tumors, respectively, accounting for 12% and 4% of all lacrimal gland lesions.2 Epithelial-myoepithelial carcinoma is an exceptional malignant epithelial tumor in view of its rarity and the relative lack of understanding of its clinical behavior. These rare tumors usually occur in the salivary gland, and, to our knowledge, only 2 cases in the lacrimal gland have been reported. One of these was a hybrid carcinoma and the other was an epithelial-myoepithelial carcinoma with pleomorphic adenoma background.1,3 We herein report a case of de novo epithelial-myoepithelial carcinoma of the lacrimal gland.

Report of a Case. An 80-year-old Chinese man had a painless, palpebral subcutaneous mass in his left upper outer eyelid for 9 months. On examination, a contrast-enhancing mass of 1 cm in diameter was confirmed to arise from the left lacrimal gland, as demonstrated on the computed tomographic scan. The left eye was the patient’s only functioning eye, with a visual acuity of 20/50; his right eye had been lost to trauma 15 years earlier. He was treated conservatively, as he refused any intervention for diagnosis. The mass gradually enlarged and displaced the left eye nasally and inferiorly. Adduction was reduced. There was also significant chemosis involving the upper bulbar conjunctiva. No lymph nodes could be palpated over the cervical and supraclavicular regions. Systemic reviews, including abdominal, respiratory, cardiovascular, and neurologic examinations, were unremarkable. The complete blood cell

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Figure 1. Computed tomographic scan of the orbit showing a mildly enhanced lacrimal soft-tissue mass arising from the superolateral aspect of the left orbit.

Figure 2. Low-power photomicrograph showing a well-circumscribed encapsulated tumor (hematoxylin-eosin, original magnification ×2).