edge, this is the first reported case of de novo epithelial-myoepithelial carcinoma of the lacrimal gland without any other tumor background. This malignant tumor, though rare, should be included in the differential diagnosis of lacrimal gland tumor.

Malignant tumor may mimic a benign lesion by deceptively appearing as a painless and slowly growing mass. In our case, sudden disproportionate rapid growth suggested its malignant nature, and repeated radiologic imaging was implemented accordingly. Theoretically, computed tomographic scanning with contrast could help to discriminate epithelial from lymphoid or inflammatory lesions of the lacrimal gland. However, the reliability of this modality in differentiating a malignant from a benign epithelial lesion is known to be poor. Radiologic features such as hyperostosis and tumor calcification, which were generally considered to be features of malignancy, might not be apparent in all cases, as in our patient. Pathological diagnosis by open biopsy is therefore the last resort in confirming underlying disease.

Recurrence and metastasis rates of epithelial-myoepithelial carcinoma from salivary gland have been reported to be from 35% to 50% and from 8.1% to 25%, respectively. In this case, the presence of tumor at the surgical margin suggests that radiotherapy or orbital exenteration might not be apparent in all cases, as in our patient. Pathological diagnosis by open biopsy is therefore the last resort in confirming underlying disease. Recurrence and metastasis rates of epithelial-myoepithelial carcinoma from salivary gland have been reported to be from 35% to 50% and from 8.1% to 25%, respectively. In this case, the presence of tumor at the surgical margin suggests that radiotherapy or orbital exenteration may be necessary. However, the ophthalmic complications or morbidity of the treatment should be discussed with the patient, especially in considering the patient’s expectations, older age at initial examination, and, in our patient, the sole functional eye. Our patient accepted the limited procedure of tumor excision only. During 24 months of follow-up, there was no evidence of either local recurrence or metastasis, and useful vision was successfully preserved. This rare tumor may behave as a low-grade malignant neoplasm, in contrast to other epithelial malignant tumors of the lacrimal gland, such as adenoid cystic carcinoma, which usually has a much poorer prognosis. However, lifelong follow-up is required, as recurrence or metastasis may occur years after surgery.

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Progression of Familial Exudative Vitreoretinopathy After Laser Treatment

Familial exudative vitreoretinopathy (FEVR) is an inherited vitreoretinal dystrophy with a variable clinical course. Early disease with no retinal detachment has been shown to respond well to primary laser treatment. The following case manifested symmetrically but responded asymmetrically to appropriate and aggressive laser treatment.

Report of a Case. A 17-month-old girl was referred to the Vitreoretinal Surgical Service of the Bascom Palmer Eye Institute, Miami, Fla, for a strong family history of FEVR. The patient was delivered at full term, her medical history was unremarkable, and her developmental milestones were intact. She had no previous ocular history, but her mother noticed that the patient was bringing objects close to her face, squinting, and bumping into things. External examination revealed primary alternating esotropia. Dilated retinal examination of both eyes showed symmetric anterior ischemic retinopathy and secondary ridge neovascularization consistent with FEVR. Focal traction in this region was noted. The patient was treated with large spot–size laser ablation by indirect ophthalmoscopy to the anterior ischemic retina. One month following treatment, the patient had good regression of the neovascularization (Figure 1). Despite this regression, extramacular vitreoretinal traction alterations persisted in both eyes. Four months later, there was minimal progression of traction in the left eye with a localized area of extramacular schisis formation. The right eye showed progressive tractional alterations through the macula with development of a falciform retinal fold (Figure 2).

Comment. Familial exudative vitreoretinopathy is an inherited vitreoretinal dystrophy characterized by premature arrest of vascularization of the peripheral retina. There is a broad spectrum of disease involvement from mild avascular reti-
nal changes that are slowly progressive to rapidly progressive tractional and/or exudative changes with total retinal detachment early in life. There has never been a prospective clinical trial for the management of active FEVR in infancy. Avascular retinal periphery with extraretinal vascularization alone has been shown to respond to laser treatment in retrospective reviews. There is a broad spectrum of treatment response, and FEVR does not always respond like retinopathy of prematurity when the avascular retina is treated with laser. Advanced disease with retinal detachment usually requires scleral buckling and/or vitreous surgery.

Previous authors have observed that patients whose onset of symptoms is before 3 years of age are at increased risk of poor visual outcome. It appears that older patients have a better prognosis because they are likely to have asymmetric disease with only 1 eye severely affected. However, loss of good visual acuity may occur even well into adulthood.

Our patient was diagnosed at 17 months of age with symmetric anterior ischemic retinopathy and secondary ridge neovascularization with limited traction. There was no evidence of retinal detachment or falciform fold at initial examination. Both eyes were treated with near confluent laser to the avascular retina. While both eyes showed regression of arborizing vessels, tractional changes progressed in 1 eye with the development of a falciform retinal fold. Pendergast and Trese described 7 patients who had similar disease involvement on initial examination as our patient. Only 1 of these patients progressed to retinal detachment after laser treatment, requiring surgical intervention. Surgical intervention was considered in our patient; however, falciform detachments are difficult to eradicate and visual outcomes are often poor. Our experience emphasizes the possibility for disease progression despite appropriate laser therapy and initial vascular involution.

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Figure 1. Familial exudative vitreoretinopathy 1 month after laser therapy. There has been complete resolution of peripheral neovascularization with mild peripheral tractional changes in both eyes. A, Right eye. B, Left eye.

Figure 2. Familial exudative vitreoretinopathy 4 months after laser therapy. There is progressive tractional alteration with falciform fold formation in the right eye (A) but minimal change in the left eye (B).
Acute retinal necrosis (ARN) occurs more often in immunocompetent individuals and is defined clinically by discrete areas of peripheral retinal necrosis with rapid confluence, vascular sheathing, and prominent inflammation in the vitreous and anterior chamber. Pathologically, full-thickness necrosis of the retina is seen in the setting of an obliterative arteritis. Fluorescein angiography and histopathologic studies have shown that the arteritis is not confined to the retinal vessels but is seen in virtually all tissues of the eye including the iris, ciliary body, choroid, and optic nerve. This vasculitic process extending beyond the ocular vessels has not been well documented. We report a case of ARN and subclinical, diffuse cerebral vasculitis that was discovered after 3.0-T magnetic resonance imaging and magnetic resonance angiography were performed.

Report of a Case. A 46-year-old man had a 2-week history of decreasing vision in the right eye. He was initially diagnosed with anterior uveitis and treated with topical prednisolone and cycloplegic drops. His vision worsened to hand motions. Fundus examination at that time showed vitritis, vascular sheathing with hemorrhages, and patchy retinal opacities that at times coalesced. The diagnosis of ARN was made. The patient was generally healthy but had suffered recently from some nonspecific, diffuse headaches and problems with forgetfulness. However, he reported no focal neurologic symptoms or signs. He had no recent skin pain or lesions.

Intravenous acyclovir was given for 10 days. Optic nerve involvement was suspected, and the patient was transferred to our hospital where high-resolution magnetic resonance imaging and magnetic resonance angiography of the orbit and brain were performed using a 3.0-T scanner. There was no optic nerve sheath distension. No abnormalities of the visual nuclei or radiations were seen. However, signal abnormalities in the right basal ganglia and right thalamus were consistent with subacute strokes of different ages (Figure 1). The angiography revealed multiple focal abnormalities in flow enhancement of the vessels throughout the brain including those supplying the areas of the stroke (Figure 2). These observations were consistent with a diffuse vasculitis. Finally, the ophthalmic arteries demonstrated irregular flow enhancement that was more severe on the right side consistent with a vasculitic process.

Cerebrospinal fluid studies revealed a pleocytosis of white cells and a positive IgG antibody titer to varicella-zoster virus. Serologic study results were unremarkable. The patient did not develop any focal neurologic signs or symptoms during the course of his follow-up.

Comment. On review of the literature, we found only 1 report of a...