tic neuritis,3 globe necrosis, and vitreous hemorrhages and necrosis, all observed in our patient, are features of the Viperidae family.5 Hemorrhagic components of viperine venom (including hyaluronidase, phospholipase A, and venom metalloproteinase) may cause severe vasospasm, endothelial damage, and increased vascular permeability. Vasospasm and/or DIC may result in vascular occlusion. Fibrin thrombi in the capillaries, perivascular hemorrhages, and necrosis, all observed in our patient, are features of DIC.

The likely cause of visual loss could be (1) ophthalmic artery occlusion with subsequent dislodgement of fibrin emboli into the end arterioles at the posterior pole; or (2) retinal necrosis and macular infarction secondary to an aborted DIC process associated with toxic optic neuropathy (venom or ASV serum toxicity).

Ocular complications following a snake bite range from keratomalacia to vitreous hemorrhage,3 uveitis, optic neuritis,3 globe necrosis, and visual loss due to cortical infarction.5 We are unaware of any previous report in the literature of macular infarction following a viperine snake bite. Visual prognosis is poor despite medical treatment.

Comment. Snake venom is a complex heterogeneous composition of substances that predominantly affects the synapse (neurotoxic) or coagulation pathway (hemotoxic). Large doses can result in disseminated intravascular coagulopathy (DIC) and ischemic damage to vital organs. Toxic vasculitis has reportedly been caused by certain species of the Viperidae family.3 Hemorrhagic components of viperine venom may induce severe vasospasm, endothelial damage, and increased vascular permeability. Vasospasm and/or DIC may result in vascular occlusion. Fibrin thrombi in the capillaries, perivascular hemorrhages, and necrosis, all observed in our patient, are features of DIC.

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Slowly Progressive Cancer-Associated Retinopathy

Cancer-associated retinopathy (CAR) is caused by antiretinal antibodies cross-reacting with pathogens expressed by carcinoma cells. Cancer-associated retinopathy with antirecoverin antibody generally shows rapidly progressive visual deterioration.1,3 Autoimmune retinopathy (AIR) is characterized by antiretinal antibody–positive serum in the absence of systemic carcinoma but with slowly progressive visual deterioration,1,3,4 although the pathogenic mechanism is uncertain. We describe a patient with CAR with antirecoverin antibody who had slowly progressive visual deterioration resembling AIR.

Report of a Case. In 2004, we examined a 62-year-old woman with a 10-year history of progressive visual loss and night blindness. In 1994, Goldmann perimetry and electroretinography (ERG) at another hospital showed retinitis pigmentosa–like findings in the right eye and a normal appearance in the left. In 2000, ERG response showed further deterioration in the right eye and a normal response in the left. In 2002, the left eye also exhibited a visual field defect and ERG abnormality. The patient had no history of carcinoma or ocular trauma.

When examined in 2004, our patient’s visual acuities were 20/30 OD and 20/20 OS. Slitlamp examination demonstrated mild iridocyclitis and nuclear cataract bilaterally. Funduscopy examination results demonstrated optic disc pallor, retinal artery whitening, and retinal pigment epithelial atrophy with partial pigmentation in the midperipheral area across 360° (Figure 1A). Fluorescein angiography demonstrated a window defect corresponding to the retinal degeneration (Figure 1B) and late leakage from retinal capillary vessels. Goldmann perimetry demonstrated central and temporal islands in the right eye and a ring scotoma in the left. Results of single-bright-flash ERG were unrecordable bilaterally. Western blot analysis detected no serum autoantibodies. One year later, a visual field defect developed in the left eye. Western blot analysis detected serum autoantibody against a 23-kDa protein (recoverin) (Figure 2A). Although the slowly progressive vi-
visual dysfunction suggested AIR, systemic screening detected a 2-mm bronchioloalveolar carcinoma without metastasis, and the patient was treated with lobectomy. Subsequently, CAR was diagnosed. A few carcinoma cells exhibited cytoplasmic immunoreactivity for recoverin (Figure 2C). One month after the lobectomy, the patient's visual acuity decreased and late-phase fluorescein angiography demonstrated cystoid macular edema in the left eye and marked leakage from retinal capillary vessels bilaterally. Oral administration of prednisolone, 40 mg/d, was initiated and then tapered across 5 months. At the last follow-up examination, the patient's visual acuity remained unchanged (20/30 OD and 20/20 OS), with no further progression of the visual field defect.

Comment. Patients with AIR exhibit slowly progressive visual deterioration mimicking retinitis pigmentosa, cystoid macular edema, and retinal vascular edema on fluorescein angiography.1,2 Some patients have systemic benign tumors.3 The patient we describe was diagnosed as having CAR with antirecoverin antibody 11 years after the initial visual symptoms appeared despite clinical features resembling AIR.

Our group previously described a patient with CAR who had aberrant expression of recoverin in many carcinoma cells.2 In the current patient the causative tumor was quite small and only a few carcinoma cells expressed the recoverin antigen, suggesting that the slow clinical course correlated with the low number of recoverin-immunopositive tumor cells, unlike patients in previously reported cases.1,2

Based on the clinical course and immunohistochemical findings, we infer that lung adenocarcinoma or preneoplastic lesions, such as atypical adenomatous hyperplasia expressing recoverin, were present when the initial visual symptoms were noticed, suggesting that some patients with AIR have an occult preneoplastic or malignant tumor expressing recoverin. We conclude that regular screening for systemic cancer may be necessary to save the lives and vision of patients with presumed AIR.

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5. Greenberg AK, Yee H, Rom WN. Preneoplastic adductive movements simultaneously observed both a slight narrowing of the palpebral fissure and missing innervation of the left eye was limited on attempted adduction of the left eye (Figure 1B and C). Upgaze and downgaze were normal. No ptosis or enophthalmos were correlated with these findings. Pupillary function was normal, with no unusual changes in eye movements. Right-eye motility was unremarkable.

Comment. Congenital ocular misinnervation can occur in a variety of forms. It typically involves the sixth cranial nerve. Most common is the Duane, or congenital retraction, syndrome, which consists of hypoplasia of the sixth nerve (nucleus) and innervation of the lateral rectus muscle by a branch of the oculomotor nerve.1

Depending on the relative contribution of third nerve fibers to the medial and lateral rectus, the patient is first with either predominant limitation of abduction or adduction. Some of our patient’s clinical features can be explained by an extreme form of the Duane syndrome, where most, if not all, oculomotor nerve branch fibers originally directed to the medial rectus innervate the lateral rectus, thus leading to simultaneous abduction on attempted adduction (also referred to as synergistic divergence).2

Interestingly, although no left-eye adduction could be elicited on lateral gaze or convergence, we observed adductive movements during sucking. Missing innervation of the medial rectus by oculomotor nerve fibers was replaced by fibers, most likely originating from a motor branch of the trigeminal nerve. Thus, the lack of innervation of the lateral rectus (Figure 2A) appears to have triggered a sequence of aberrant nerve sprouting, resulting, initially, in a shift of fibers originally meant for the medial rectus toward the lateral rectus (Figure 2B). Second, and possibly as a consequence of lack of innervation of the medial rectus muscle, a shift of trigeminal nerve motor fibers to the medial rectus took place (Figure 2C), leading to a trigemino-oculomotor synkinesis between the lateral pterygoid or one of the suprahyoid muscles and the medial rectus. The slight narrowing of the palpebral fissure observed during sucking can be explained by synkinetic contraction of the medial muscle against a tight lateral rectus muscle in that specific situation, which led to discrete retraction of the globe.

The combination of the Duane and Marcus Gunn syndromes or other misinnervation syndromes involving the sixth, third, and fifth cranial nerves also recurs. Other cases with synergistic divergence and a trigemino-oculomotor synkinesis have been reported.