quently accompanied by ALS features. The neurodegenerative diseases showing parkinsonism, dementia, or both, including PD, AD, progressive supranuclear palsy, striatonigral degeneration, dementia with Lewy bodies, and corticobasal degeneration, were excluded on the basis of the clinical diagnostic criteria and neuro-radiologic examinations. Ocular fundus of the patients was examined by one of the authors (K.I.), an ophthalmologist.

Results. PR was found in one of the five patients. The patient was a 63-year-old woman with Kii PDC and a dense family history of ALS/PDC. She was born and grew up in Hohara and developed parkinsonian gait disturbance at age 60. At age 63, she had ophthalmologic examinations that showed moderate parkinsonism and motor neuron signs, consisting of gait disturbance, rigidity of the upper extremities, and muscle wasting of the hand and lower extremities. She showed mild memory disturbance and slowing of psychomotor speed.

She did not have visual complaints. Corrected visual acuity was 25/20 on the right and 20/20 on the left. Pupils were equal in size and reacted promptly to light and accommodation. Ocular motility testing showed saccadic pursuit movement. Slit-lamp biomicroscopy of the anterior segment showed mild cataracts in both eyes. Dilated fundus examination revealed PR consisting of linear and crisscrossed defects in the retinal pigment epithelium in the lateral and lower areas of the fundus of the right eye (figure). The retina of the left eye was normal. Any causative substances including fly larva were not detected.

Discussion. PROG is characterized by meandering, crisscrossed, hypopigmented tracks of retinal pigment epithelium that mimics OIP.⁵ Patients with PROG or OIP show preservation of relatively good visual function with minimal or no signs or symptoms of inflammation. While OIP is caused by subretinal invasion by fly larva, the cause of PROG remains unclear. Thirteen eyes from seven ALS/PDC patients with PROG were pathologically investigated.⁶ Other disorders that present with PROG-like retinopathy include diffuse unilateral subacute neuroretinitis, angiod streaks, Siegrist streaks, myopic lacquer cracks, and histoplasmosis streaks, but PROG can be distinguished from them by its peculiar retinal findings.

In 1993, extensive investigation of the epidemiology of PR in the Guamanian Chamorro population revealed that PROG was found in 85 of 531 (16%) of the neurologically normal group and 38 of 72 (53%) of ALS/PDC patients. In addition, PROG was found exclusively in the Chamorro population and never in other ethnic groups on Guam.⁷ These findings suggest PROG is unique to the Chamorro population including individuals with ALS/PDC.

We found PR in one of the five patients with Kii ALS/PDC resembling PROG. A patient with OIP or PROG-like PR is novel. The existence of a very rare OIP-like PR in ALS/PDC patients from two different isolated ethnic groups suggests that OIP-like PR is not a coincidental finding but may be one of the clinical manifestations of ALS/PDC closely related to the etiology.

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Ophthalmoplegia in Powassan encephalitis

Simmons Lessell, MD; and T. Edward Collins, DO

Powassan encephalitis, a rare arthropod-borne infection, is being increasingly recognized in New England, four cases having recently been reported.¹ We describe an unusual ophthalmoplegia in one of these patients.

In September 2000, a 53-year-old woman residing in Maine developed nausea, vomiting, diarrhea, dizziness, diplopia, and incoordination followed within days by arm weakness, urinary retention, fever, delirium, and almost total ophthalmoplegia. Her pupils were 2 mm and weakly reactive. There was no history of a rash or tick bite.

A head CT scan was normal. MRI showed a tiny focus of signal hyperintensity in the white matter of each temporal lobe. Her

Figure. Fundus photograph of a patient with Kii parkinsonism-dementia complex, showing linear and crisscrossed tracks of depigmentation of the retinal pigment epithelium in the lateral periphery of the fundus of the right eye.

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erythrocyte sedimentation rate was normal, and her white blood cell count was 11,000/mm³. The CSF contained 148 white cells/mm³ (46% neutrophils, 40% lymphocytes, 14% monocytes) and 6 red cells. Protein and glucose levels were normal. Blood and CSF cultures were sterile. Serologic tests for *Herpes simplex*, *Listeria*, Lyme borreliosis, West Nile virus, eastern equine encephalitis virus, lymphocytic choriomeningitis virus, leptospirosis, and GQ1B antibody were negative. She was treated with ceftriaxone, ampicillin, and acyclovir.

Thirteen days after onset, she had anxiety, depression, impairment of recent memory, difficulty maintaining attention, bifacial weakness, ophthalmoplegia, dysarthria, and bilateral arm and neck weakness. Deep tendon reflexes were hypoactive in the arms but normal in the legs, with flexor plantar responses. Sensation was normal. MRI showed multiple areas of T2 hyperintensity in

the cerebellum and throughout the hemispheres, but none in the brainstem. Some enhanced with gadolinium.

Powassan infection was diagnosed from serum drawn 19 days after onset that was positive for Powassan-specific IgM and a neutralizing antibody titer of 1:640. PCR testing is not a requisite for diagnosis.

Over the succeeding months, all nonocular signs improved. Seven months after onset, she had normal visual acuity, color vision, visual fields, lids, pupils, anterior segments, vitreous, and fundi. The right eye was slightly exotropic. She appeared unable to execute any eye movements on command, look to an eccentric target, or follow a target. However, with prolonged effort, after a 10- to 20-second delay, both eyes would drift conjugately 30° up or down and 20° left or right. The eyes would return slowly to the primary position. Doll's eye maneuvers elicited nearly full, but slow and delayed, conjugate excursions. Three months later, she could promptly initiate up-gaze, but the movements remained slow. No improvement occurred thereafter.

Powassan encephalitis was first recognized in 1958 in a fatal case in Powassan, Ontario, Canada. At autopsy, lesions were present throughout the neuraxis. Several patients reported from Siberia had cerebellovestibular lesions, but ophthalmoplegia has not been noted.

Our patient's preserved doll's eye movements indicate a supranuclear basis for her ophthalmoplegia. Loss of her saccades could be from lesions involving the median and paramedian pontine reticular formation and median basis pontis.² Inability to maintain eccentric gaze suggests that there probably was involvement of the nucleus prepositus hypoglossi and medial vestibular nucleus.^{3,4} Impaired smooth pursuit could be from involvement of the dorsolateral pontine nucleus.⁵ Impairment of vertical movements could reflect involvement of the rostral interstitial nucleus

of the medical longitudinal fasciculus and the interstitial nucleus of Cajal⁶ or the nucleus reticularis tegmenti pontis (which would also explain the horizontal deficits).⁷

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Corrections

DC-EEG recording: A paradigm shift in seizure localization?

In the editorial “DC-EEG recording: A paradigm shift in seizure localization?” (*Neurology* 2003;60:1062–1063) by Lagerlund and Gross, the correct e-mail address for Dr. Robert Gross is: robert_gross@urmc.rochester.edu. The publisher apologizes for this error.

Variability of total phenytoin serum concentrations within elderly nursing home residents

In the article “Variability of total phenytoin serum concentrations within elderly nursing home residents” (*Neurology* 60;4:555–559) by Birnbaum et al., middle initials were incorrectly omitted from the names of two authors. The authors' names should read as follows: A.K. Birnbaum and T.E. Lackner. The publisher apologizes for this error.

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