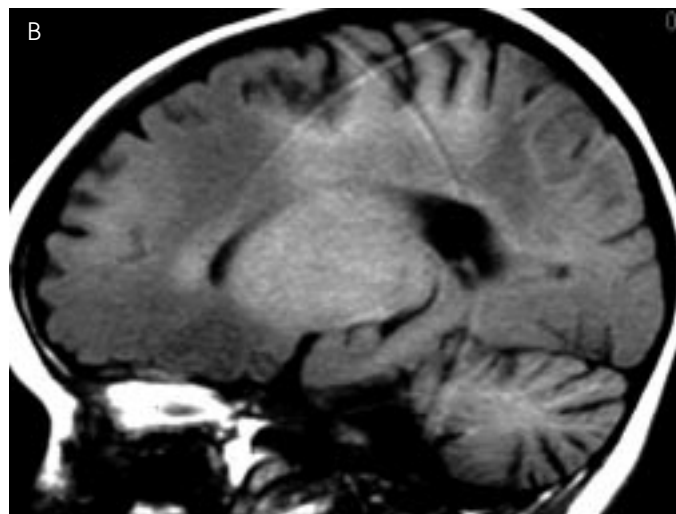

Case W7

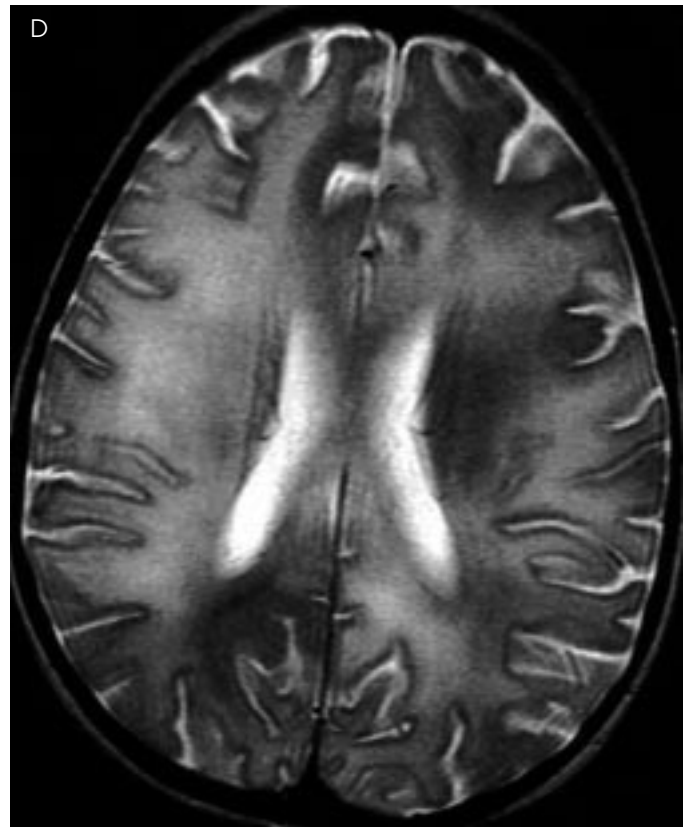
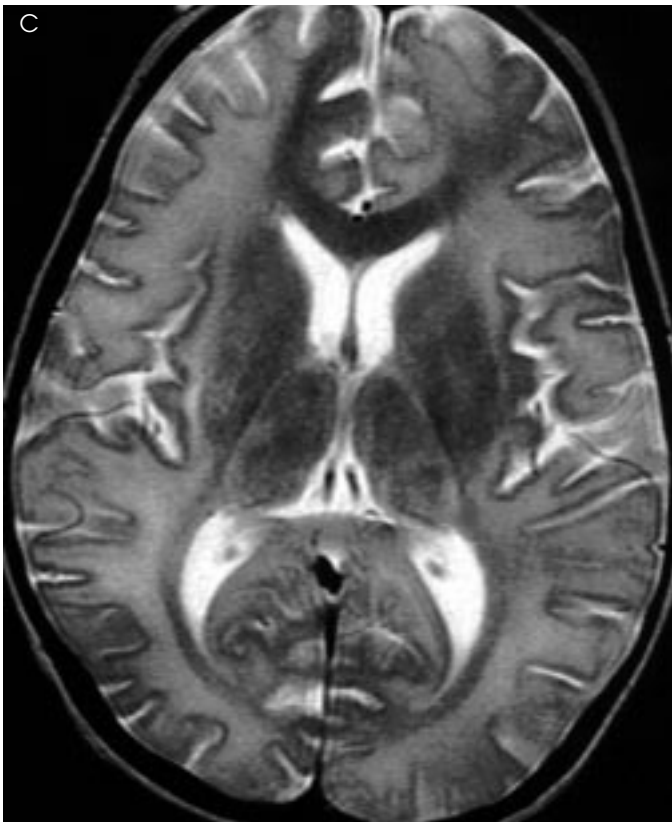
Clinical Presentation

A 9-year-old Mexican boy did poorly in school for some months and became increasingly withdrawn. He came to medical attention when he developed “jerking” movements consistent with myoclonus.



Radiologic Findings

A non-contrast CT scan (Fig. A) shows generalized prominence of the ventricles and sulci for the patient's age. In addition, the white matter is abnormally hypodense. A sagittal T1-WI (Fig. B) shows patchy areas of abnormal hypointensity within the hemispheric white matter.



Two axial T2-WIs (Figs. C and D) demonstrate global parenchymal volume loss as well as diffuse, mostly symmetric T2 prolongation involving both the deep and peripheral white matter. Note the complete lack of mass effect associated with this process. On post-gadolinium images (not shown), there was no evidence for any pathologic enhancement.

Diagnosis

Subacute sclerosing panencephalitis (SSPE)

Differential Diagnosis

- Other viral encephalitides (usually have a more acute presentation)
- Acute disseminated encephalomyelitis (more acute presentation, typically multifocal rather than diffuse)
- Other demyelinating conditions (multiple sclerosis, toxin exposure, radiation, chemotherapy—clinical history essential)
- Dysmyelinating diseases (leukodystrophies: often earlier onset, may have a family history of the disorder)

Discussion

Background

SSPE is a typically fatal neurodegenerative disease that develops as a consequence of early childhood measles infection. It has been nearly eradicated in de-

Pearls

- SSPE may occur in HIV-positive children. In this setting, it often has an earlier onset and more rapid progression.
- Consider SSPE in an immigrant child with behavioral changes and multifocal white matter disease.

Pitfall

- There is a relatively poor correlation between the clinical stage of SSPE and the MR findings.

veloped nations by widespread measles vaccination, but it is still endemic in many developing nations. In the United States, it most commonly occurs among immigrant populations.

Etiology

SSPE is caused by measles virus which becomes altered and remains dormant intracellularly following the initial infection. The factors that trigger reactivation are poorly understood.

Clinical Findings

SSPE initially presents as a minor behavioral disturbance in a previously healthy child: irritability, impaired school performance, and social withdrawal are common. Most patients are between ages 4 to 14 years at the onset of symptoms; however, adult onset may occur, often with an atypical course.

There is an inevitable progression to myoclonus, dementia, coma and eventually death. The time course of progression is variable: many patients will die within a year of becoming symptomatic, but others have survived >10 years.

Pathology

Gross

- The brain is atrophic and the white matter may be abnormally firm

Microscopic

- Subacute inflammatory changes, extensive demyelination and gliosis, and neuronal and glial viral inclusions are identified

Imaging Findings

CT

- Atrophy, low attenuation white matter lesions

MR

- Progression of findings over time in a constant pattern:
 - Early: focal T2-prolonging lesions in the white matter. No mass effect or enhancement is seen.
 - Intermediate: progressive atrophy and gray matter involvement
 - Late: almost total loss of white matter, severe gray matter atrophy, and basal ganglia abnormalities (multifocal T2-prolonging lesions)

Treatment

- Experimental protocols include treatment with interferon alpha and oral isoprenosine
- Treatment is often begun late due to diagnostic difficulties: viral genomic analysis may allow earlier diagnosis

Prognosis

Generally poor, although there is significant variability in the course of the disease and some patients experience plateaus or even spontaneous improvement

Suggested Readings

Brismar J, Gascon GG, von Steyern KV, Bohlega S. Subacute sclerosing panencephalitis: evaluation with CT and MR. *AJNR* 17:761–772, 1996.

Callebaut DP, Cras P, Martin JJ. Prolonged and atypical course in some cases of subacute sclerosing panencephalitis. *Acta Neurologica Belgica* 97:39–44, 1997.

Tuncay R, Akman-Demir G, Gokyigit A, et al. MRI in subacute sclerosing panencephalitis. *Neuroradiology* 38:636–640, 1996.