

## Case 1991-2

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### CLINICAL HISTORY:

The patient is a 32 year old male with no significant past medical history. When he was 28 years old, he developed seizures and optic disc swelling without loss of visual acuity. CT showed diffuse enhancement predominantly in the white matter of the left frontal and right parietal areas. MRI showed multiple areas of increased T2 signals in the same distribution. An LP was done. The opening pressure was not recorded. No cells were present. The protein was 35mg, glucose 69mg, and IgG was 6.17mg/dl (normal up to 3.7mg/dl). Isoelectric focusing showed multiple bands. Visual evoked potentials and EEG were normal. Over the ensuing years his seizures were difficult to control and he developed progressive cognitive decline, followed by reflex asymmetries. On the present admission, he had moderate dementia characterized mainly by frontal lobe type behavior, accompanied by marked reflex asymmetries and bilateral ankle clonus. At this time, there was no papilledema or optic disc pallor. CT showed large ring enhancing lesions that on MRI appeared to have progressed to a mild degree over the course of his illness. EEG's on several occasions remained normal. Antibody testing for HIV and HTLV-1 was negative, and very long chain fatty acids and arylsulfatase A were in the normal range. A left frontal craniotomy revealed a diffusely firm cortical surface. Two 3 x 3 x 2.5cm biopsies were taken, which included cerebral cortex and underlying white matter.

MATERIAL SUBMITTED: one H & E slide, one unstained slide, one gross photograph

- POINTS FOR DISCUSSION: 1) Diagnosis
- 2) Pathogenesis