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CASE 2003-7

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Clinical History: The patient, an 11-year old girl, was born and raised in Tampa FL. She was in her usual state of health (with the possible exception of a recent onset of subtle behavioral problems), when one night, she developed repetitive episodes of stiffness of all extremities lasting for about 10 seconds and recurring 40 times during the first night. This evolved into generalized tonic-clonic seizures that transiently responded to depakote; while on this antiepileptic medication, one week later, she developed "head drops", 20-40 per day that necessitated an increase in depakote dosage and addition of Dilantin. This combined drug treatment worked for about one week, when head drops recurred at increasing frequency. Three months later another medication (keppra) was added. The seizures eventually stopped, but one month later, the patient became progressively more lethargic. She was transferred from the local hospital to MCH for evaluation of "progressive encephalopathy."

On admission, she was unresponsive, had a fever of 99.1 and supple neck; there was horizontal nystagmus, 2+ deep tendon reflexes and bilaterally positive Babinski. Bacterial and viral CSF cultures were negative; the CSF contained no cells, but there was a minimal increase in protein and presence of oligoclonal bands. Past medical history was unremarkable; the patient was vaccinated according to the widely practiced schedule in USA.

Dual-echo FLAIR-MRI study demonstrated bilateral, diffusely hyperintense signal most pronounced in the hemispherical white matter; there was no contrast enhancement. MR-spectroscopy demonstrated reduction in N-acetylaspartate, normal choline and elevated lactate & myoinositol.

A diagnostic right parietal brain biopsy was performed.

Pathohistologically, there were minimal multifocal lymphocytic-plasmacellular subarachnoidal infiltrates. Both cerebral cortex and subcortical white matter contained perivascular lymphocytic infiltrates and rather pronounced ongoing neuronal, myelin and axonal degeneration. Bacterial and viral brain tissue cultures were negative.

A treatment was initiated and the patient was discharged. She expired about one year after the onset of her illness. Autopsy was not performed.

Material submitted: H&E stained brain biopsy tissue sections.

Points for discussion: Diagnostic tests ?