

Submitted by: Angeline R. Mastri, M.D. and Joo Ho Sung, M.D.
Departments of Pathology and Neurology, University of Minnesota
Medical School, Minneapolis, MN

Clinical Abstract:

This 4 month old white female infant was admitted to the hospital because of cyanosis, respiratory distress and possible seizures. She required intubation, ventilatory support and antibiotics for pneumonia. She was also noted to have arching activity, hypertonicity and eye deviation thought to be seizures. Neurological examination showed no focal signs. Strength was normal but tone was increased and deep tendon reflexes were brisk. She had deep furrows on her palms and soles of the feet. EEG showed multifocal spikes and suppression of the background activity. CT scan showed enlarged ventricles bilaterally with some enhancement in the periventricular regions suggesting ventriculitis or calcifications. CSF was normal except for slightly elevated protein. During her hospitalization she was noted to have marked choreoretinitis consistent with a congenital viral infection. TORCH and viral titers were normal. Open lung biopsy showed interstitial pneumonitis of unknown etiology. When her respiratory condition improved she was transferred to a local hospital but she returned one week later because of worsening respiratory status. She again required ventilatory support and had uncontrollable seizures despite adequate drug levels of anticonvulsants (Tegretol, Depokene, Dilantin and phenobarbital). CT scan showed atrophy of the brain and dilated ventricles. The infant died at 4 months of age of cardiopulmonary failure.

Autopsy Findings: Bronchopulmonary dysplasia, interstitial pneumonia, acute bronchopneumonia, pulmonary air block syndrome, growth retardation.

Neuropathology: The temporo-occipital regions showed localized polymicrogyria. Olfactory bulbs and tracts were unusually large. In sections of the cerebrum, there was moderate dilatation of the frontal horns but severe dilatation of the atria and occipital and temporal horns. The cerebral tissue in most areas was extremely firm. The thalami were fused and the third ventricle small. The striae of the thalami were unusually large. The corpus callosum and white matter were reduced in volume. The inferior olives in the medulla were enlarged and the pyramids were small. The dorsal vermis and anterior lobes of the cerebellum were firm and shrunken.

Material Submitted: 1) olfactory bulb (H & E) and
2) medulla and spinal cord (H & E)

Points for discussion: 1) Diagnosis
2) Pathogenesis