AANP Slide Session 1959 - Case 6

Submitted by: E.P. Richardson, Jr. M.D. Neuropathology Laboratory Massachusetts General Hospital Boston, Massachusetts

This male infant was thought to be normal at birth, which occurred after normal pregnancy and was uncomplicated. At one month, however, he was thought to be abnormally listless and slack. Essentially no progress was made in motor development. At 8 months, he was seen at the Children's Medical Center, Boston, Massachusetts where he was found to show no interest in his surroundings, did not play with toys, and did not use his hands. He followed a light with his eyes and smiled when spoken to. Tonic neck reflexes were active, the tendon reflexes were exaggerated, and the plantar reflexes bilaterally extensor.

He died with signs of

acute pneumonia at age 15 months.

The eldest sibling died at about 6 months of age with a progressive cerebral disorder, thought to have been brought on by chicken pox. Three sisters were well.

Autopsy: The cause of death was an interstitial pneumonia. The findings in the organs generally were otherwise unremarkable.

The skull measured 52 cm. in circumference (chest 44 cm.). The brain weighed 1540 gm. The most striking macroscopic abnormality was a peculiar softness and translucency of the white matter on section, most pronounced in the subcortical and convolutional portions. The ventricles were not enlarged.

Microscopically, the changes in other parts of the brain were exactly similar to those illustrated by the material submitted.