

CASE #9

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This child was first admitted to hospital at 7 months of age because of increasing head size and inability to sit up or roll over. Pregnancy had been complicated by spotting and mother had taken "nerve pills" throughout gestation. Infant was born at 38 weeks gestation. She turned her head to the left for the first time at 2 months but did not turn head to right. Head circumference was 48 cm. and demonstrated asymmetrical frontal bossing, greater on the left side and a larger right posterior fossa. The anterior fontanelle was widely patent and full and a skull defect was palpable in the right occipital bone; the foramen magnum was enlarged. The infant had bilateral papilledema and a wry neck with head turned to left. Various neurological signs and studies indicated the presence of a posterior fossa tumor which was located in the right cerebellar hemisphere but could only be incompletely removed. Post-operatively, she developed focal seizures, tachycardia, an elevated temperature and various other complications all of which slowly resolved. She was transferred to a home for disabled children and died at 3-1/2 years of age.

NECROPSY FINDINGS: Limited to nervous system cerebral hemispheres were diffusely atrophic. Large residual tumor mass right cerebellar hemisphere with extension into brain stem and rostral cervical spinal cord.

MATERIAL SUBMITTED: One (1) slide of original biopsy, one (1) slide from cerebellum (necropsy) both stained with H & E; two (2) Kodachromes of cerebellum, brain stem and cord and one (1) electron micrograph from biopsy.

POINTS FOR DISCUSSION:

What is the diagnosis?