

Case 2

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Clinical Abstract:

The patient was an 11 month old female who developed nose and gingival bleeding of one week's duration. The child was well until age 8.5 months when she fell out of bed. Aside from a scalp bruise, there were no problems until 3 weeks later, when she developed hypersomnia and vomiting. Also at about this time there began a gradual decline in the child's previously normal developmental abilities. She was felt to have had a concussion, and an anemia was discovered for which iron and vitamins were prescribed. The mother had had some type of anemia as a child, from which she had fully recovered. Past medical history was otherwise unremarkable.

On admission at age 10 months physical examination revealed massive splenomegaly and multiple cranial nerve palsies. Pertinent laboratory findings included WBC 1700 with 2% segs., 3% bands, 89% lymphocytes, 4% monocytes, 1% metamyelocytes, 1% myelocytes and 8,000 platelets. Hgb 6.2 gm., Hct 20%, reticulocyte count 12.2.

She developed a right hemiplegia. Bone marrow examination was consistent with severe peripheral destruction of blood elements. EEG showed diffuse marked slowing. Because of her hypersplenism, splenectomy was performed. Liver biopsy showed massive fatty change. She developed choreo-athetoid movements and cortical blindness. Lumbar puncture was considered to be traumatic. Chemotherapy was begun 4 days prior to death. She further deteriorated and died.

Necropsy Findings: The general autopsy showed generalized lymphadenopathy and hepatomegaly. There was Candidiasis of the pharynx, larynx, trachea and esophagus.

The dura, basilar vessels, spinal cord, and external examination of the brain were unremarkable. The white matter of the left frontal lobe and both occipital lobes was diffusely granular with a homogeneous gelatinous translucency and depressed cut surface. The cerebral cortex and U-fibers appeared spared. The cerebellar white matter and cortex were similarly involved. Sections of the brainstem were unremarkable.

MATERIAL SUBMITTED: One slide of pons, stained with H & E. One slide of cerebrum, unstained.

Points for Discussion:

Diagnosis