

CASE 2007-6

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Clinical and neurological history: The patient was a 5-year-old girl with developmental delay, seizures and recurrent strokes. She was born at full-term, birth wt 5 lbs. At 5 days, she developed generalized convulsions, which resolved after correction of hypocalcaemia. Neuroimaging and EEG were unrevealing. Chromosome analysis was normal. She walked at 18 months of age, spoke around 12 months of age, but did not use short phrases until age 3. By age 3, her right arm was smaller than the left and she was strongly left-handed. Intermittent episodes of transient weakness involved the right side, but there were no generalized convulsions. MRI revealed bilateral infarcts in the temporal, parietal and occipital lobes, some of which appeared recent. At age 5, her height and weight were at the 23rd percentile, with head circumference <5th percentile. She had low-set ears with joined lobules, mild facial hypoplasia, a single palmar crease on the right, and a transverse plantar crease on the left sole. Cranial nerve function was intact. Her right arm was hypoplastic, with flexor posturing at the right shoulder and elbow and decreased proximal right arm tone. Plantar responses were flexor and there were no cerebellar signs. Arteriogram showed multifocal vascular stenoses, with left anterior cerebral artery filling by collaterals. A neurosurgical procedure under EEG monitoring was performed. Immediately post-operatively, the patient had three generalized tonic-clonic seizures. The following morning, MRI showed significant infarcts in the bilateral ACA, bilateral MCA, and left PCA territories. Her neurologic status progressively worsened and the family chose to withdraw life support.

Neuropathology findings: The brain weighed 1170 g. The external diameter of the supraclinoid right ICA was 1.5-2 mm and the lumen was <1mm. The external diameter of the supraclinoid left ICA was <1mm with no grossly apparent lumen. The basilar artery was small and was severely narrowed at its midpoint. The vertebral arteries were normal. The cerebral cortex was markedly attenuated and consisted of a thin, firm yellow-white band. The underlying white matter was also firm. Many other cortical areas were soft and dusky gray (bilateral ACA territory, bilateral inferior frontal lobes, both frontal lobes immediately superior to the Sylvian fissure, left posterior superior temporal gyrus, temporal tips, left insular cortex, and left parieto-occipital lobe). There was diffuse vascular congestion.

Material submitted: H&E stained section of a narrowed artery segment.

Point for discussion: Diagnosis;
Pathogenesis.