

**Canadian Association of Neuropathologists
L'Association Canadienne des Neuropathologistes**

CASE No 8

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AK was a previously healthy 10 month old infant when she initially presented with 12 hours of vomiting and generalized illness plus 2 weeks of irritability and mild drowsiness. AK was admitted to her local hospital with possible gastroenteritis, but she suddenly deteriorated and became unresponsive with a fixed right pupil. CT scanning revealed a 7x7x8 cm hemorrhagic tumor in the right cerebral hemisphere causing midline shift. AK underwent emergency surgery wherein a very vascular tumor was encountered. Because of significant blood loss only 25% of the tumor was resected. After stabilization 1 week later, the remaining tumor was gross totally resected (confirmed by MRI). The tumor resected during these surgeries was identical and remarkably uniform throughout histologically (slide 8a, 2008). Adjuvant treatment included intensive and aggressive Head Start III Regimen C chemotherapy (Carboplatin, Vincristine, temozolomide and GCSF support). Unfortunately, AK did not tolerate this treatment well, and due to irritability, a subdural-peritoneal shunt was placed; MRI suggested areas of residual tumor that were confirmed via biopsy. AK continued to do poorly and a subsequent MRI suggested progression of disease; treatment was subsequently directed towards palliation. Surprisingly, AK slowly improved and eventually returned home. 21 months after her initial surgery, she was thriving with residual deficits including left hemiparesis. The family was reluctant to return for follow up, but re-imaging was agreed to. 3 weeks prior to this scheduled visit AK began to deteriorate. CT scanning revealed a large mass at the site of previous surgery and hydrocephalus. MRI confirmed the CT findings; an 8.5 x 5.5 x 9.0 cm primarily solid enhancing tumor was identified. AK again underwent gross total resection (slide 8b, 2010). Again, the histologic appearance of the tumor was remarkably uniform throughout. Postoperative MRI revealed only a small region of abnormality that was suspicious of tumor. At last follow up, 3.5 months after her latest surgery, AK was doing well and heading back home after undergoing intensive rehabilitation.

Materials submitted: H&E stained representative slide from each of the initial (2008) and latest (2010) ~gross total resections.

Question: What is your overall diagnosis?