Canadian Association of Neuropathologists

L'Association Canadienne des Neuropathologistes

CASE No 8

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This 8 year-old boy presented with a 2 week history of feeling unwell that included persistent nausea and fatigue. There was intermittent diplopia and a single episode of vomiting. His balance was impaired and he had left-sided coordination issues. Past medical and family histories were unremarkable.

On exam the boy displayed a diffuse reduction in tone and 3+ deep tendon reflexes. He had subtle left sided dysmetria and reduction in agility. Systemic exam was normal.
Initial CT scanning revealed a very large posterior fossa tumor. Subsequent MR imaging highlighted an enhancing left tentorial lesion that was several cm in dimension. There was significant mass effect, with distortion of the cerebellum and brainstem, and prominent peritumoral edema. At surgery, tumor was appreciated on both surfaces of the tentorium. A gross total resection was thought to be achieved, but there was some residual tentorial thickening noted on post-operative imaging. CSF was negative at diagnosis.

Immunohistochemical workup revealed strong staining for the following: vimentin, NSE, desmin (in part, dot-like), EMA and CD99, the latter two of which displayed a membranous pattern. Focal positivity was noted for AE1/AE3 and synaptophysin. Staining for non-phosphorylated neurofilament protein was equivocal. Tumor cells were negative for the following: CD34, CD31, LCA, HMB 45, S-100, myogenin, myoD1, SMA, GFAP, phosphorylated neurofilament protein (p-NFP) and WT1. INI-1/BAF47 staining revealed retention of nuclear positivity within tumor cells. The Ki67 index was high. S-100, GFAP, synaptophysin and p-NFP suggested a component of brain invasion.

Material submitted: 1 H&E section of the tumor

Questions:
1. Differential diagnosis?
2. Further workup?