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Anaplastic ganglioglioma arising from a Lhermitte-Duclos-like lesion. Case report.

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The authors report the case of a 7-year-old boy with a history of developmental delay who presented with aggressive behavior. A magnetic resonance (MR) image showed a mass lesion originating from the cerebellar vermis with an atypical folial pattern and contrast enhancement. Histologically, the subtotally resected specimen consisted mostly of neuropil with nodular foci of ganglion cells. Lhermitte-Duclos disease (LDD) was diagnosed in the patient. A retrospective review of the tissue sections showed a nidus of associated astrocytic proliferation, suggesting a diagnosis of ganglioglioma. Five years later, the patient experienced an altered mental state and a facial droop. An MR image revealed a cerebellar mass with cystic areas and an enhancing nodule. The resected tissue specimen consisted primarily of a mixed proliferation of glial and ganglion cells consistent with a ganglioglioma. Two years later, a third craniectomy was performed in the patient for worsening headache and ataxia. Histologically, the tumor showed progressive anaplasia and was most accurately classified as an anaplastic ganglioglioma. Immunohistochemically, most of the tumor cells were immunoreactive for anti-phospho-mammalian target of rapamycin (mTOR) and phospho-S6 ribosomal protein antibodies. In contrast, the subpopulation of neoplastic ganglion cells in the tissue, particularly from the first surgery, did not express phosphatase and tensin homolog deleted from chromosome 10 (PTEN). This immunohistochemical pattern suggests that the large dysplastic ganglion cells (the gangliocytomatous component) forming the greater part of the lesion were associated with activation of the phosphatidylinositol 3-kinase-PTEN/Akt/mTOR signaling pathway, a feature previously reported in LDD. This case represents the first report of an anaplastic ganglioglioma arising in an LDD-like lesion.

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