

Abstract

The biological origin of cerebellar liponeurocytomas is unknown, and hereditary forms of this disease have not been described. Here, the authors present clinical and histopathological findings of a young patient with a cerebellar liponeurocytoma who had multiple immediate family members who harbored similar intracranial tumors. A 37-year-old otherwise healthy woman presented with a history of progressive headaches. Lipomatous medulloblastoma had been diagnosed previously in her mother and maternal grandfather, and her maternal uncle had a supratentorial liponeurocytoma. MRI revealed a large, poorly enhancing, lipomatous mass emanating from the superior vermis that produced marked compression of posterior fossa structures. An uncomplicated supracerebellar infratentorial approach was used to resect the lesion. Genetic and histopathological analyses of the lesion revealed neuronal, glial, and lipomatous differentiation and confirmed the diagnosis of cerebellar liponeurocytoma. A comparison of the tumors resected from the patient and, 22 years previously, her mother revealed similar features. Cerebellar liponeurocytoma is a poorly understood entity. This report provides novel evidence of an inheritable predisposition for tumor development. Accurate diagnosis and reporting of clinical outcomes and associated genetic and histopathological changes are necessary for guiding prognosis and developing recommendations for patient care.

KEYWORDS: GFAP = glial fibrillary acidic protein; IDH1 = isocitrate dehydrogenase 1; MAP-2 = microtubule-associated protein 2; MIB-1 = monoclonal antihuman Ki 67; brain tumor; familial; lipomatous medulloblastoma; neurolipocytoma; oncology

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