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Cerebellar liponeurocytoma: An updated comprehensive review of clinicopathologic, immunohistochemical, and molecular features of an unusual but distinct tumor

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Abstract

Cerebellar liponeurocytoma (CL) is a rare WHO grade 2 tumor characterized by advanced neuronal differentiation and variable lipomatous features. Initially classified as a subtype of medulloblastoma, CL was later considered as a distinct entity owing to its peculiar morphological and molecular features and significant better outcome. Typically affecting adults, CL often presents with symptoms related to cerebellar dysfunction, including headaches, ataxia, and gait disturbances. On magnetic resonance imaging, this tumor presents as a well-defined, heterogeneous mass with lipomatous components, which may be less or more apparent depending on their extent. Histologically, CL is composed of neurocytic cells and lipidized tumor cells; the immunohistochemical positivity for synaptophysin and NeuN confirms the neuronal differentiation of neoplastic cells. In spite of its morphological similarity to medulloblastoma, CL lacks the genetic alterations commonly found in this tumor, but some cases display *TP53* mutations. Complete surgical resection is the gold standard treatment, whereas the benefit of adjuvant radiotherapy is controversial. CL generally harbors a favorable prognosis, with low recurrence rates in cases with incomplete resection or high proliferative index. The present paper comprehensively reviews the literature about CL, emphasizing the clinicopathologic and molecular features of this unusual but distinct neuropathological entity.