Cerebellar Liponeurocytoma with an Unusually Aggressive Histopathology: Case Report and Review of the Literature

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We report a rare case of cerebellar liponeurocytoma with an unusually aggressive histopathology. A 49-year-old man presented with a four-month history of headache, vertigo, and progressive swaying gait. Magnetic resonance imaging showed a 3×3.5 cm sized relatively well-demarcated round mass lesion in the fourth ventricle, characterized by high signal intensity on T2-weighted images. Postcontrast images revealed strong enhancement of the solid portion and the cyst wall. The patient underwent suboccipital craniectomy and tumor removal. The pathologic diagnosis was cerebellar liponeurocytoma. Adjuvant radiotherapy was offered due to concerns related to the high proliferative index (Ki-67, 13.68%) of the tumor. At the last routine postoperative follow-up visit (12 months), the patient complained of no specific symptom and there was no evidence of tumor recurrence. However, long-term follow-up and the analysis of similar cases are necessary because of the low number of reports and the short follow-up of cases.

Key Words: Liponeurocytoma · Ki-67 index · Radiotherapy.
foci of hyperintense signals that displays moderate contrast enhancement on T1-weighted images. On T2-weighted MRI, the tumor is slightly hyperintense at its cortex. Areas of fat density on computed tomography (CT) scans and of T1 hyperintensity on MR images help to distinguish this rare neoplasm from the more common ependymoma and medulloblastoma. The Ki-67 labeling index (the percentage of Ki-67 positive nuclei in 500 tumor cells from a microscopic field with the highest labeled nucleus density) was as high as 13.68% (Fig. 2D). Focal necrosis was evident.

The patient’s postoperative course was uneventful and the vertigo and unsteady gait resolved before discharge. Adjuvant radiotherapy was offered due to concerns related to the high proliferative index of the tumor, and subsequently, he underwent conventional radiotherapy (5400 cGy/10 fractions to whole brain). At the last routine postoperative follow-up visit (12 months), the patient complained of no specific symptom and there was no evidence of tumor recurrence.

**DISCUSSION**

Cerebellar liponeurocytoma is a rare, newly identified neoplasm found in adults, and is reputed to be benign. The most challenging aspect of the differential diagnosis based on radiological findings is to distinguish this disease entity from adult medulloblastoma and ependymoma. In particular, ependymomas are usually hypo- to iso-intense compared with brain parenchyma on T1-weighted magnetic resonance (MR) images, although hemorrhage, necrosis, calcification, or tumor vascularity may account for observations of intratumoral heterogeneity with foci of increased signal intensity. Cerebellar liponeurocytoma commonly presents as a hypointense mass with scattered vacuole displacing the nucleus to the periphery. The majority of cells demonstrated neuronal differentiation characterized by widespread immunopositivity for synaptophysin and glial fibrillary acidic protein. These pathologic features were consistent with liponeurocytoma. The Ki-67 labeling index (the percentage of Ki-67 positive nuclei in 500 tumor cells from a microscopic field with the highest labeled nucleus density) was as high as 13.68% (Fig. 2D). Focal necrosis was evident.

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neuronal cells with rare Homer-Wright rosettes and pseudorosettes admixed with foci of well-differentiated adipocyte-like cells, and for some time has been considered an adult lipomatous variant of medulloblastoma with much more benign clinical character than the common non-lipomatous medulloblastoma. The proliferative index of the tumor is usually low as determined by Ki-67 indices. However, several cases of elevated mitotic activity have been reported. Here, we report a case of cerebellar liponeurocytoma with an unusually high proliferative index showing histopathological evidence of a more aggressive form of cerebellar liponeurocytoma (focal necrosis and a high proliferation index). No consensus has been reached regarding the treatment of cerebellar liponeurocytoma, especially concerning the necessity for chemo- or radiotherapy a component of the postoperative treatment regimen. Cacciola et al. reviewed the findings of all reported cases of cerebellar liponeurocytoma and concluded that complete macroscopic resection with long-term follow-up would be the most appropriate treatment. Buccoliero et al. reported a recurrent case and reviewed the related literature. It was concluded that liponeurocytoma is an uncertain malignant potential lesion when mitoses are present and MIB-1 positive cells constitute more than 10% of total neoplastic cells. In general, most cerebellar liponeurocytomas have low proliferative activity and a favorable clinical outcome. However, this should be interpreted with caution because of rarity of this tumor and lack of systemic follow-up; recurrence rates reach 50% in reported case of gross total resection without radiotherapy. Châtillon et al. suggested that radiotherapy should be considered at least following incomplete resection and after complete resection of tumors with an elevated proliferation index (>6%). Table 1 summarizes treatments and follow-up of the reported cases of cerebellar liponeurocytoma with high proliferation index, which was searched from MEDLINE database. After reviewing the literature and thoroughly discussing the subject with each other, we recommended adjuvant radiotherapy because of the incomplete resection due to adhesion to both peduncles and the high Ki-67 index (13.68%) despite the primary nature of this case. We also assumed that tumors with a higher proliferation index behave more aggressively and tend to recur earlier than those with a lower proliferation index. Fortunately, in the described case there was no evidence of recurrence at last follow-up. However, tumor recurrence and the potential morbidity associated with adjuvant radiotherapy must be kept in mind.

**CONCLUSION**

Cerebellar liponeurocytoma is a newly recognized posterior fossa tumor with characteristic histological features. The proliferation index in this tumor is usually low and previous reports support a benign natural history. However, our case, which masqueraded as ependymoma radiologically, is noteworthy for a high proliferation index, although its clinical course has not yet been fully determined. The low number of reported cases and the short follow-up of cases make the prognosis of cerebellar liponeurocytoma difficult. Accordingly, long-term follow-up and the analysis of similar cases are necessary.

**References**