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Case Report

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

Sang-Bong Chung, M.D.,[1] Yeon-Lim Suh, M.D.,[2] Jung-Il Lee, M.D.[1]

Departments of Neurosurgery,[1] Pathology,[2] Samsung Medical Center, Sungkyunkwan University School of Medicine, Seoul, Korea

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.

INTRODUCTION

First described by Bechtel et al.[3], cerebellar liponeurocytomas are posterior fossa tumors composed of densely packed neuronal cells admixed with foci of well-differentiated adipocyte-like cells. Similar tumors have been referred to as lipomatous medulloblastoma, lipidized medulloblastoma, medullocytoma, neurolipocytoma, lipomatous glioneurocytoma, or lipidized mature neuroectodermal tumor of the cerebellum[1,7-9,14,16]. However, cerebellar liponeurocytoma is genetically distinct from medulloblastoma and central neurocytoma[10], and is now recognized as a separate entity by the World Health Organization (WHO)[13], although only about 40 cases have been reported since it was first described in 1978. This rare tumor is of WHO grade I-II, and generally exhibits an indolent behavior. Here, we report a case of cerebellar liponeurocytoma with an unusually aggressive histopathology.

CASE REPORT

A 49-year-old previously healthy man, presented with a four-month history of headache, vertigo, and a progressive swaying gait. General physical examination was unremarkable, but neurological examination revealed dysmetria of the right upper extremity and decreased rapid alternating movement of the right hand. His gait was unsteady and he had a diminished ability to tandem walk. Magnetic resonance imaging (MRI) 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 (Fig. 1A). The mass lesion seemed to have cystic component isointense to cerebrospinal fluid on T1- and T2-weighted images (Fig. 1A, B). Postcontrast images revealed strong enhancement of the solid portion and the cyst wall (Fig. 1C, D). The ventricular system did not seem dilated.

The patient underwent sub-occipital craniectomy and tumor removal. The tumor was purple colored and had a friable consistency. Intra-operative frozen section was reported as being consistent with ependymoma. Near total resection only was achieved because of some infiltration on both peduncles by the tumor.

Pathologic examination revealed a highly cellular neoplasm composed of monomorphic small round neuronal cells mixed with lipomatous cells and characterized by a single cytoplasmic

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• Address for reprints : Jung-Il Lee, M.D.
Department of Neurosurgery, Samsung Medical Center, Sungkyunkwan University School of Medicine, 50 Irwon-dong, Gangnam-gu, Seoul 135-710, Korea
Tel : +82-2-3410-3494, Fax : +82-2-3410-0048, E-mail : jilee@skku.edu

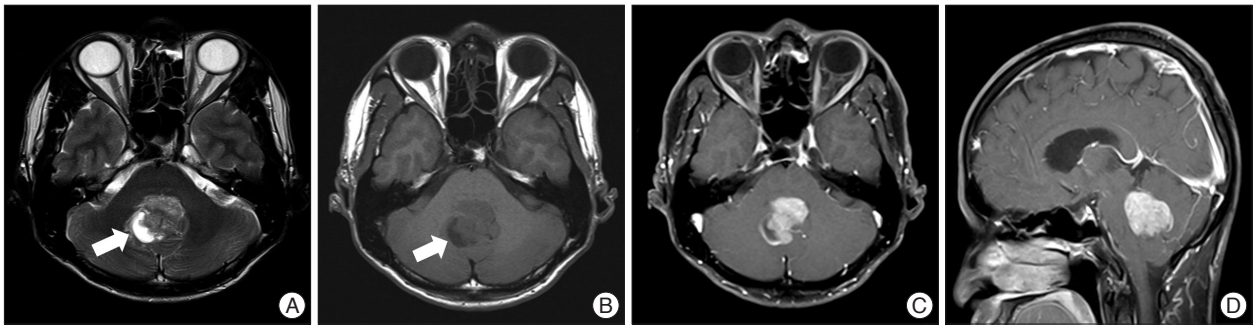

Fig. 1. Magnetic resonance (MR) images of the lesion. T2-weighted (A) and T1-weighted (B) MR images showing a 3×3.5 cm sized relatively well-demarcated round mass in the fourth ventricle. The lesion had a cystic component isointense to cerebrospinal fluid (arrows in A and B). Postcontrast axial (C) and sagittal (D) MR images showing strong enhancement.

vacuole displacing the nucleus to the periphery (Fig. 2A). The majority of cells demonstrated neuronal differentiation characterized by widespread immunopositivity for synaptophysin (Fig. 2B). Tumor cells were largely negative for glial fibrillary acidic protein (Fig. 2C). 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.

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.

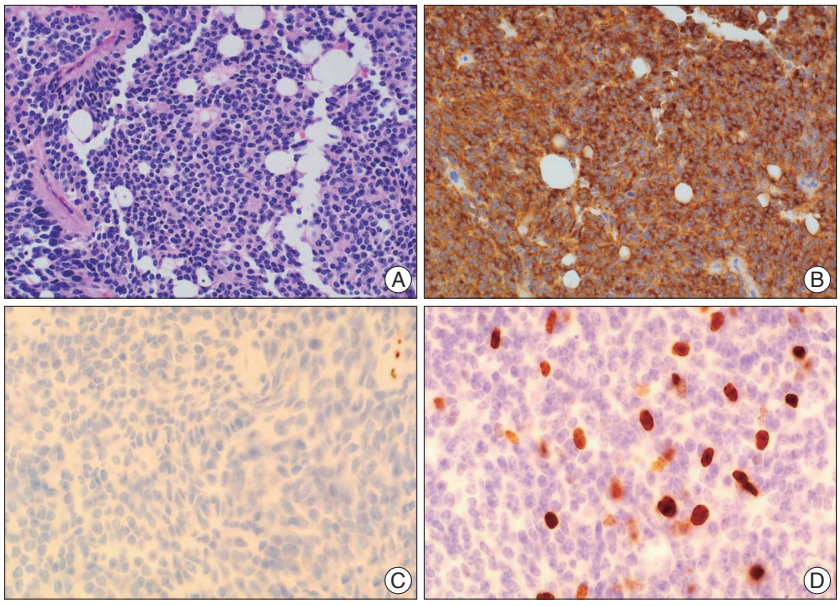

Fig. 2. Pathologic examination reveals a highly cellular neoplasm composed of small round cells mixed with lipomatous cells characterized by a single cytoplasmic vacuole, displacing the nucleus to the periphery (A, H&E, ×200). The majority of cells demonstrates neuronal differentiation characterized by immunopositivity for synaptophysin (B, immunostain, ×200) and immunonegativity for glial fibrillary acidic protein stains (C). The Ki-67 proliferation index was 13.68% in small cell areas (D).

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[2]. Cerebellar liponeurocytoma commonly presents as a hypointense mass with scattered 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[2,11,12]. In the described case, the lesion was iso- to hypointense on T1-weighted images and the cyst wall was enhanced after contrast administration, which are findings suggestive of ependymoma. Hyperintense area suggesting fat content was not observed during the T1-weighted sequence. Moreover, cyst formation is often seen in ependymoma and is rare in liponeurocytoma[2]. Cerebellar liponeurocytoma might have been difficult to suspect based on neuroimaging findings in the present case. MRI with a fat-suppression sequence would have been helpful for differential diagnosis, especially if no CT scan is conducted, as in our patient.

Cerebellar liponeurocytoma is characterized by densely packed

Table 1. Cases of cerebellar liponeurocytoma with high proliferation index reported with treatment and follow-up

Authors (year)	Age/Sex	Diagnosis	Proliferation index	Treatment	Follow-up
Cacciola et al.[5] (2002)	61/M	Cerebellar liponeurocytoma	15%	GTR	No recurrence at 1 years 6 months
Owler et al.[15] (2005)	34/M	Cerebellar liponeurocytoma	20%	GTR+RT	No recurrence at 8 months
Buccoliero et al.[4] (2005)	64/M	Cerebellar liponeurocytoma	15%	GTR+RT	No recurrence at 5 months
Châtillon et al.[6] (2009)	42/F	Cerebellar liponeurocytoma	10%	GTR+RT	No recurrence at 3 years
Present study (2012)	49/M	Cerebellar liponeurocytoma	13.6%	NTR+RT	No recurrence at 12 months

GTR : grow total removal, RT : radiation therapy, NTR : near total removal

neuronal cells with rare Homer-Wright rosettes and pseudorosettes admixed with foci of well-differentiated adipocyte-like cells[1,4,5,10], 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[4-6,15]. 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.[5] 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.[4] 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[6]. Châtillon et al.[6] 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.

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