

# Cerebellar Liponeurocytoma: A Case Report

## ABSTRACT

Cerebellar liponeurocytoma is a rare cerebellar tumor, with only about 20 cases reported under many different names. Although the few cases described in the literature support the relatively benign nature of this lesion, the optimum treatment strategy and long-term behavior still have to be defined. A 39-year old man presented with a 6-month history of headache and gait disturbance. Magnetic resonance imaging (MRI) studies disclosed a midline cerebellar mass. Gross total resection of the tumor was accomplished through a suboccipital craniotomy. The histopathological diagnosis was cerebellar liponeurocytoma. The postoperative course was uneventful and the patient was discharged six days after the surgery. No radiotherapy was given. Cerebellar liponeurocytoma is a rare, benign neuroepithelial tumor that occurs exclusively in the cerebellum of adults. The morphological appearance of this neoplasm can be confused with that of oligodendroglioma, neurocytoma, ependymoma, medulloblastoma, solid hemangioblastoma and metastatic carcinomas. This tumor should be added to the differential diagnosis of mass lesions of the posterior fossa.

The small number of patients with reported cerebellar liponeurocytomas limits our understanding of the tumor's natural history. Most of the information available from case reports indicates that this tumor has benign biological behavior. The available follow-up data suggest a favorable prognosis, but this has yet to be confirmed in a larger series of cases.

## INTRODUCTION

Cerebellar liponeurocytoma is a rare tumor of the posterior fossa that has been reported fewer than 20 times in the literature. This tumor has been known by many names, including lipomatous medulloblastoma (5,7), lipidized medulloblastoma (8), medulloctoma (10), and lipomatous glineurocytoma (3). Cerebellar liponeurocytomas were recognized in the revised WHO classification of tumors of the central nervous system, 2000, as a distinct pathological entity (14). This uncommon tumor was first described by Bechtel (4) in 1978 .It has been suggested in the available literature that these tumors have a favorable prognosis (5,10,15).

Here, we report a case of cerebellar liponeurocytoma with clinical, histological, and radiological studies.

## CASE REPORT

The patient was a 39-year-old man who presented with gait disturbance and occipital headache. On physical examination, only nystagmus and papilledema was observed.

Neuroimaging findings: MRI studies disclosed a 6x5x2 cm midline cerebellar mass. The mass filled the fourth ventricle and displaced the brainstem forward. It extended caudally through the cerebellomedullary cistern and foramen magnum to the level of C2. (Figure 1 A,B,C).

Operation and postoperative course: Surgery was carried out with the patient in the sitting position. A median suboccipital craniotomy and

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Received : 22.02.2006

Accepted: 10.05.2006

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C1 laminectomy were performed. A dural incision was made. A gray-reddish tumor was exposed between the cerebellar tonsils. The tumor extended to the level of C2. It was partially attached to the surrounding tissue but well circumscribed. The tumor was easily cleavable and gross total removal was achieved. The postoperative course was uneventful and the patient was discharged 6 days after the surgery. The histopathological diagnosis was cerebellar liponeurocytoma. No radiotherapy was given. Neurological examination revealed normal findings while MRI confirmed gross total removal and no residual tumor was seen 45 days after operation. (Figure 2 A, B)

Pathological findings: The surgical specimen was a gray-reddish colored tissue with a smooth surface, measuring 7x5x2 cm. Histological examination revealed a neoplasm composed of neurocytes, uniform small cells with round to oval nuclei and scant oligo-like cytoplasm, besides prominent lipidization foci containing cells resembling adipocytes. Vascular septa were significant throughout the tumor. Necrosis and microvascular proliferation were absent. Mitoses were scant or absent (Figure 3A). Immunohistochemically, neurocytes stained diffusely and strongly with neurone-specific enolase (NSE) (Figure 3 B) and synaptophysin, whereas glial fibrillary acidic protein (GFAP) staining was weak and localized focally around the vessels showing neuropil-like substance. MIB-1 labeling index was measured below 1%. Tumor tissue did not express neurofibrillary protein (NFP).

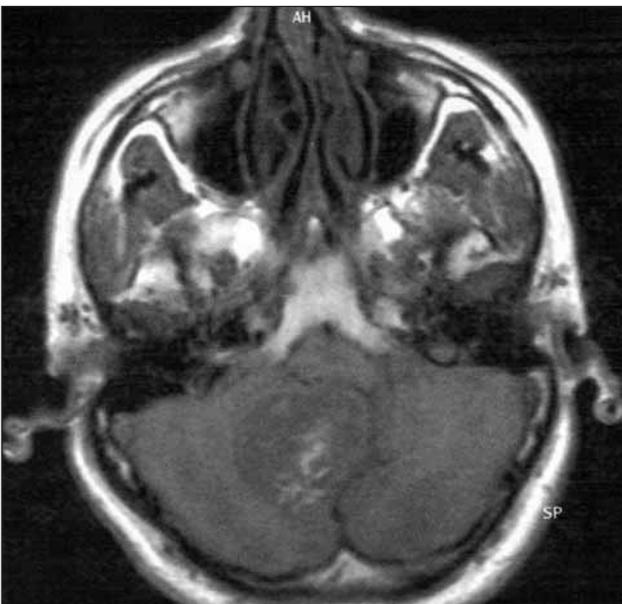


Figure 1A: Axial and sagittal.

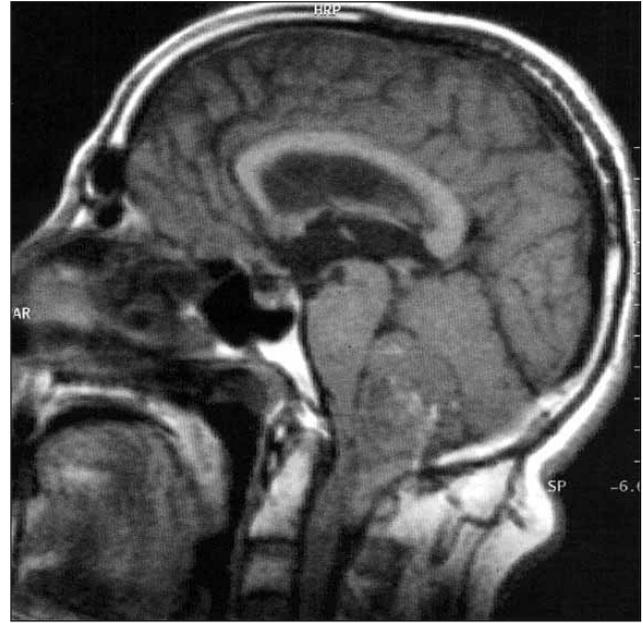


Figure 1B: T1-weighted MR images showing a heterogeneous mass with hyperintense areas due to adipose content. The mass is compressing the brainstem and extending to the level of C2. Axial.

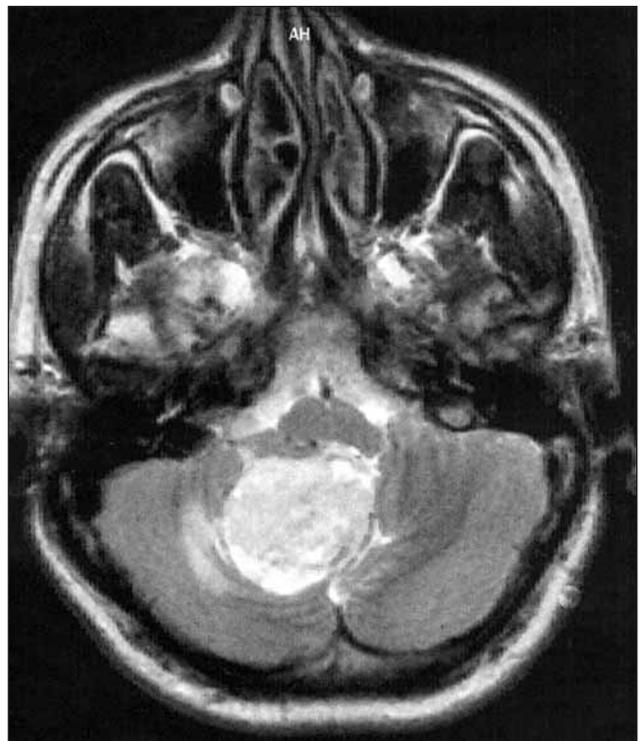
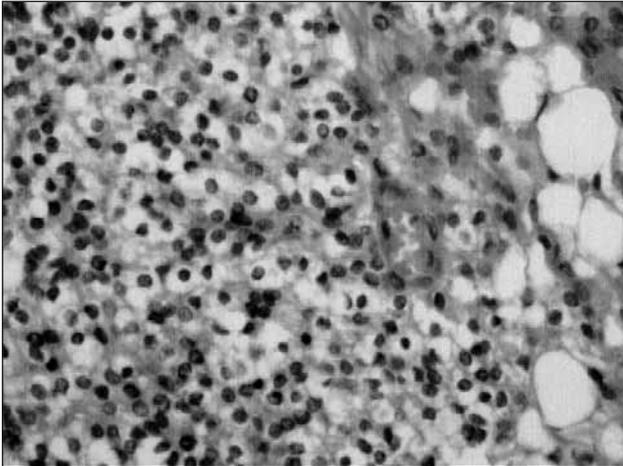
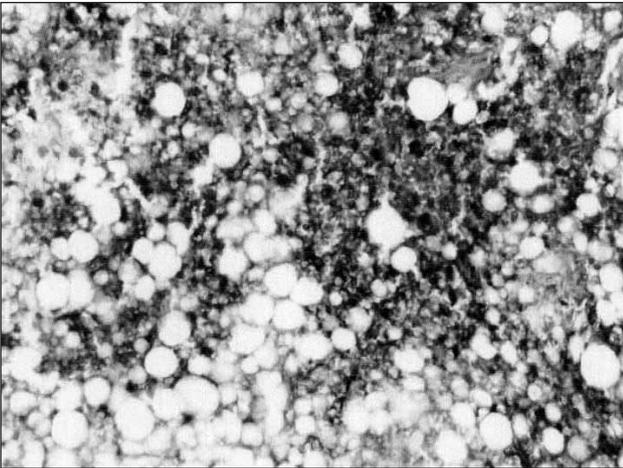


Figure 1C: T2-weighted image depicting the slightly hyperintense signal of the tumor compared with cerebellum.



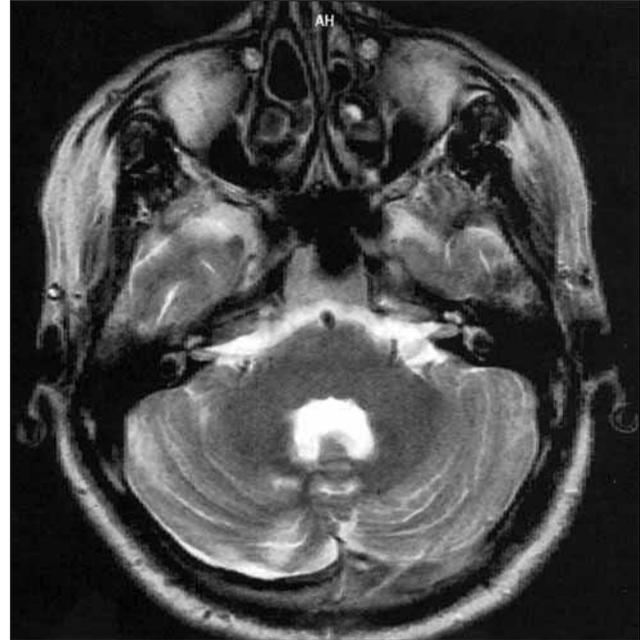
**Figure 2A:** Light microscopic findings of cerebellar liponeurocytoma in tissue section H&E, x400.



**Figure 2B:** Diffuse NSE expression of the tumor tissue. (Immunochemistry for NSEx300).



**Figure 3A:** Post-operative MR images: Sagittal T1-weighted and axial.



**Figure 3B:** T2-weighted images showing no evidence of residual tumor.

### DISCUSSION

Since 2000, the Central Nervous System tumors classification (WHO) includes a subset called cerebellar liponeurocytoma (13). Cerebellar liponeurocytomas are neuroectodermal tumors consisting of both neuronal and glial elements. Immunohistochemistry for GFAP, synaptophysin and NSE are usually positive indicating the mixed glial and neuronal elements. Microscopically, the tumor consists of small, round to ovoid cells with eosinophilic, scanty cytoplasm. Interspersed regions of lipidized cells resemble mature adipocytes. Mitoses, areas of vascular proliferation and necrosis are all rare (14). Mitotic activity is usually absent and the growth fraction, as reflected by the MIB-1 labeling index, is in the range of 1%-3% (14). In our case, MIB-1 labeling index was measured below 1%. These tumors tend to occur in older patients ranging from 36 to 77 years of age, with a mean age of approximately 53 years (5, 7, 8-10).

Cerebellar liponeurocytoma has a relatively benign clinical course and a recurrence may appear after a long period of time (1, 3, 8-10, 12, 14). Reviews published in the literature show a 5-year survival rate of 81% (11) but Jenkinson (12) reported a patient who developed a recurrence 12 months after a subtotal resection. His case demonstrated an atypical clinical course of a highly aggressive and radiation-

resistant tumor, despite the consistent absence of aggressive histological features.

Radiological diagnosis is difficult due to the rarity of the tumor and a variable imaging appearance (2). The most challenging differential diagnosis of these tumors is to distinguish them from medulloblastomas, ependymomas, solid hemangioblastomas, astrocytomas and metastases (1). MRI appearance is variable and may be related to the distribution and proportion of lipidized tissue. On T1-weighted MRI the tumor is usually hyperintense but heterogeneous. Heterogeneity is due to the large island of lipid in the tumor (8). Enhancement with gadolinium is usually irregular and heterogeneous. On T2-weighted images, the tumor is heterogenous and slightly hyperintense. Peritumoral edema is not usually seen.

Surgery to establish the diagnosis and remove the lesion should be the initial therapeutic maneuver. The aim of the surgery is gross total resection of the tumor. In most of cases there is a reasonable border between the tumor and surrounding tissue (2, 6, 11) and gross total removal of the tumor is possible. In our case, there were no adherences or invasion between the tumor and the adjacent structures. The most common dilemma is whether to administer postoperative radiotherapy. According to Kleihues (14), the prognosis is favorable if the MIB-1 index is in the range of 1%-3%. and aggressive adjuvant therapy is not mandatory. There have been no reports of spinal drop metastases in the literature and it is therefore reasonable to avoid spinal radiation.

### CONCLUSIONS

The most recent 2000 WHO classification of Tumors of the Nervous System describes a new entity, cerebellar liponeurocytoma (13). This rare tumor manifests with cerebellar symptoms in adults and is characterized histopathologically by the presence of mature adipocytes in an otherwise typical primitive neuroectodermal neoplasm with low mitotic activity. Most of the information available from case reports indicates that this tumor has a favorable prognosis, but this is yet to be confirmed in a larger series of cases. If the tumor is removed grossly totally and the MIB-1 index is not high, adjuvant radiotherapy may not be necessary.

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