Cerebellar liponeurocytoma

Case report and review of the literature

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Cerebellar liponeurocytoma is a rare tumor of the posterior fossa that has many morphological similarities to medulloblastoma and neurocytoma. Recently the World Health Organization working group for classification of central nervous system neoplasms adopted the term “cerebellar liponeurocytoma” to provide a unified nomenclature for a tumor variously labeled in the literature as lipomatous medulloblastoma, lipidized medulloblastoma, medullocytoma, neurolipocytoma, lipomatous glio-neurocytoma, and lipidized mature neuroectodermal tumor of the cerebellum. The rarity of this tumor and paucity of pertinent information regarding its biological potential and natural history have resulted in the application of various treatment modalities. It is suggested in the available literature that these lesions have a much more favorable prognosis than typical medulloblastomas, and that adjuvant therapy for liponeurocytoma need not be as extensive as that administered for medulloblastomas.

KEY WORDS • cerebellum • cerebellar liponeurocytoma • lipidized medulloblastoma • lipomatous medulloblastoma • tumor • adjuvant therapy

Case Report

History and Physical Examination. This 66-year-old man presented with a 2-year history of frontal headache, along with a 1-month history of nausea and emesis. He reported ataxia manifested by falling to the left and had experienced increasing sleepiness for approximately 2 weeks. On physical examination he showed no evidence of papilledema or nystagmus. His strength was normal and there was no pronator drift. His gait was unsteady and slow; however, he demonstrated no tendency to fall to either side. Results of Romberg testing were negative. He had mild left-sided dysmetria and decreased ability for rapid alternating movement on the left. An MR image demonstrated a 6 × 4.5 × 4-cm mass in the left cerebellar hemisphere, with central subacute to chronic hemorrhage and heterogeneous moderate enhancement throughout. There were two proteinaceous cysts in the medial aspect of the mass, and there was minimal to no surrounding edema (Fig. 1).

Operation and Postoperative Course. The patient underwent left paramedian suboccipital craniotomy for resection of his tumor. On opening of the dura, abnormal tissue was immediately apparent. The tumor was resected using biopsies forceps and ultrasonic aspiration. The initial diagnosis on review of frozen sections was primary small cell neoplasm, most likely medulloblastoma. The tumor was believed to be consistent with cerebellar liponeurocytoma on fixed sections. The patient tolerated the procedure well and was discharged home on postoperative Day 2. His mild left-sided dysmetria was unchanged from his preoperative examination. Postoperative MR imaging demonstrated no evidence of residual contrast enhancement in the cerebellum. An MR image of the lumbar spine revealed no evidence of drop metastatic lesions. The patient subsequently...
received conventionally fractionated (1.8 Gy/fraction) craniospinal irradiation to 36 Gy, followed by a boost to the posterior fossa to a total of 54 Gy.

Pathological Findings. The tissue appeared amorphous on gross inspection, and was red and variably semitranslucent. Light microscopy examination demonstrated features of neuronal and glial differentiation in various areas (Fig. 2). There was very prominent lipocytic differentiation in the tumor cells. A significant portion of the tumor appeared to be primitive, highly cellular, and entirely characteristic of medulloblastoma in morphological features and growth pattern (Fig. 2 upper). There were few mitoses.

Electron microscopy examination demonstrated small, closely packed cells with variations in nucleocytoplasmic features according to the degree of cellular maturation. Cytoplasmic microtubules were common to both primitive and mature forms. In areas typical of primitive neuroectodermal tumors, nuclear features included invagination and lobulation of the nuclear membrane, finely granular chromatin, and small nucleoli. Uneven margination of heterochromatin was present beneath the nuclear membrane. Organelles were scant and consisted almost entirely of polyribosomes and mitochondria. In most of the tumor examined ultrastructurally, the cells displayed a higher degree of maturation. Instead of adjacent nuclei indenting the neighboring cells, individual cells possessed more cytoplasm and demonstrated a greater variety of organelles and nuclei with no or few invaginations. Focal cell membrane thickening was noted but no well-formed junction-like membrane densities were seen. Nucleoli were more conspicuous. The most obvious feature of the tumor was the numerous small and large, cleared nonmembrane-bound vacuolar structures compatible with intracellular lipid. Intracytoplasmic coalescence of small lipid droplets into large macrodroplets was prominently seen (Fig. 3). Dense core membrane-bound vesicles were noted in the cytosol, and endoplasmic reticulum stacking and unusual concentric whorls were present (Fig. 3, inset). Microfilament-packed processes compatible with astrocytic cell processes were present but scarce. The ultrastructural appearance was similar to previously published descriptions of this tumor.1,5,8,12

The proliferation index assessed using MIB-1 immunohistochemical studies was 3%, which is within the previously reported range of less than 1 to 6% for liponeurocytoma.10

Discussion

A neuroectodermal tumor with well-differentiated adipose tissue occurring in the cerebellum was first reported by Bechtel, et al.2 To date, there have been fewer than

![Fig. 1. Appearance of a cerebellar liponeurocytoma on axial T1-weighted MR images without (upper left) and with (upper right) addition of contrast agent, a T2-weighted MR image (lower left), and a computerized tomography scan (lower right).](image1)

![Fig. 2. Photomicrographs showing appearance of cerebellar liponeurocytoma in tissue sections. H & E, original magnifications ×82 (upper), ×413 (lower).](image2)
20 reported cases filling the description of cerebellar liponeurocytoma. The available information indicates that, despite their similar microscopic appearance, cerebellar liponeurocytomas differ significantly from medulloblastomas. Approximately 75 to 80% of the time, medulloblastomas occur in children. A retrospective review of 354 cases of medulloblastoma in children failed to disclose any tumors with a significant lipid component. Medulloblastomas presenting in adulthood tend to affect young adults (mean age 28 years) and are rare in persons above the age of 50. In contrast, cerebellar liponeurocytomas tend to occur in older patients. The reported cases have occurred in patients ranging from 36 to 77 years of age, with a mean age of approximately 53 years (Table 1).

The biological behavior of these tumors is also quite different when compared with medulloblastoma; they tend to have a much lower rate of mitotic activity. Growth fractions have ranged from less than 1 to 6%, with a mean value of 2.5%. Growth fractions for medulloblastomas range from 15 to greater than 50% in the most active areas. There have been no reported cases of drop metastasis with cerebellar liponeurocytomas, whereas six (75%) of eight adults with medulloblastomas who did not receive spinal cord radiation developed metastatic lesions, according to the review published by Hubbard, et al. Table 1 demonstrates the relatively good prognosis for patients with cerebellar liponeurocytoma whose cases have been reported in the literature to date. Among the eight patients who did not receive adjuvant therapy, two died within 1 year of the operation (one immediately from a postoperative complication and one from an unrelated cause). Among the six surviving beyond 1 year, three (50%) developed tumor recurrence limited to the posterior fossa between 5 years and 12 years postoperatively. Imprecise

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**TABLE 1**

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Age (yrs), Sex, Tumor Location</th>
<th>Adjuvant Therapy</th>
<th>Follow-Up Findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>Davis, et al., 1993</td>
<td>49, F vermis; 53, M vermis</td>
<td>none</td>
<td>NED, &gt;5 yrs recurr 5 yrs postop; at 10 yrs, NED after 2nd resection</td>
</tr>
<tr>
<td>Ellison, et al., 1993</td>
<td>36, F lt hemis</td>
<td>none</td>
<td>NED, 2 yrs recur 10 yrs postop; recur 1 yr after 2nd resection</td>
</tr>
<tr>
<td>57, F vermis; 37, M lt hemis</td>
<td>59 Gy PF</td>
<td>none</td>
<td>NED, &gt;3.5 yrs</td>
</tr>
<tr>
<td>Budka &amp; Chimelli, 1994</td>
<td>53, M lt CPA</td>
<td>none</td>
<td>died of unrelated cause 1 yr postop</td>
</tr>
<tr>
<td>Giangaspero, et al., 1996†</td>
<td>48, F rt hemis; 59, F lt CPA</td>
<td>50 Gy, 55 Gy &amp; 24 Gy</td>
<td>NED, &gt;5 yrs</td>
</tr>
<tr>
<td>Walter, et al., 1994†</td>
<td>77, F rt hemis</td>
<td>none</td>
<td>died of unrelated cause 1 yr postop</td>
</tr>
<tr>
<td>Giangaspero, et al., 2000†</td>
<td>66, M rt CPA</td>
<td>54 Gy</td>
<td>NED, 5 yrs</td>
</tr>
<tr>
<td>Schild, et al., 1997</td>
<td>44, M rt hemis; 50, F rt hemis</td>
<td>none</td>
<td>died 18 hrs postop NED, 7 yrs</td>
</tr>
<tr>
<td>present study</td>
<td>66, M lt hemis</td>
<td>54 Gy &amp; 36 Gy</td>
<td>NED, 6 mos</td>
</tr>
</tbody>
</table>

* CPA = cerebellopontine angle; hemis = hemisphere; NED = no evidence of disease; PF = posterior fossa; recur = recurrence.
† Personal communication with author.

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**FIG. 3.** Electron micrograph demonstrating primitive neuroectodermal and neurocytomatous tumor cell morphology. Note the lipid macro- and microdroplet accumulation. Original magnification × 4000. Inset: Electron micrograph showing a peculiar concentric endoplasmic reticulum whorl. Original magnification × 13,500.
knowledge of the natural history of this tumor has resulted in the use of many forms of postoperative adjuvant therapy. In those patients receiving adjuvant radiotherapy, there were no recurrences (although the follow-up period in these patients was shorter than for those not receiving adjuvant therapy). All reported recurrences were confined to the posterior fossa. No distant metastases or, as mentioned previously, drop metastases have been reported. The prognosis appears to be very similar to that of central neurocytoma. In a review of central neurocytomas by Schild, et al.,11 the authors found a 5-year survival rate of 81% (88% for those who received radiotherapy compared with 71% for those who did not). In their series of 32 cases, there were no distant metastases, only local recurrences. Three patients who received salvage radiotherapy after recurrence were alive 1 to 6 years posttreatment.

Conclusions

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 is less aggressive than typical medulloblastomas and is similar in prognosis to central neurocytomas. The low mitotic rate, absence of drop metastases, and long disease-free survival in several patients who received no adjuvant therapy point to a less aggressive course. There have been late recurrences in 50% of patients who survived surgical resection and who did not receive adjuvant therapy. Because of the high rate of recurrence in patients who have not received postoperative radiotherapy and the absence of recurrence outside of the posterior fossa, radiotherapy that is limited to the posterior fossa appears to be a reasonable adjuvant treatment.

References


Manuscript received March 5, 2001. Accepted in final form June 15, 2001.

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