Central neurocytomas are relatively rare brain tumors that are usually attributed to midline septal or ventricular structures. A predisposition for extension into the lateral ventricle has been noted. Histologically these lesions resemble intraventricular oligodendrogliomas. Symptomatic tumors usually require resection. Progressive growth of untreated tumors may lead to focal neurological signs or hydrocephalus. Neurocytomas are histologically benign, although some tumors have cytological features that may herald more aggressive behavior.16,18,29 For tumors in critical locations that remain after resection or biopsy procedures, subsequent management options have included observation, radiation therapy, and now stereotactic radiosurgery. We report results in four patients who underwent radiosurgery for persistent neurocytomas.

Case Reports

Case 1

This 16-year-old young woman underwent two resections (one transfrontal and one transcallosal) for a large lateral and third ventricular tumor. The majority of the neurocytoma was removed during the first two surgeries. Residual tumor was seen in the left lateral ventricle wall (Fig. 1). After surgery the patient continued to exhibit mild neurocognitive deficits. She was referred to us for stereotactic radiosurgery to treat the residual tumor. Radiosurgery was performed using the Leksell gamma knife (Elekta Instruments, Atlanta, GA). A combination of five 14-mm isocenters was used to deliver a dose to the tumor margin of 14 Gy and a maximum dose of 28 Gy. The residual tumor volume measured 7.9 cm³. Subsequent imaging studies demonstrated complete regression of the contrast-enhancing tumor mass (Fig. 1). Now, 53 months after radiosurgery, she has had no new clinical symptoms and continues to undergo periodic imaging.

Case 2

This patient was a 46-year-old woman who presented with headaches. A mass in the left lateral ventricle was identified. Examination of a biopsy sample revealed a neurocytoma. She was referred to us for stereotactic radiosurgery as primary management of the tumor. Radiosurgery was performed with a combination of two 14-mm and two 8-mm isocenters. A dose to the margin of 15 Gy and a maximum dose of 30 Gy were administered. The initial tumor volume measured 4.2 cm³ (Fig. 2). Follow-up imaging studies obtained 40 months later revealed significant decrease in tumor size. She remains well 50 months later with no new neurological problems and continues to undergo periodic imaging.

Abbreviation used in this paper: MR = magnetic resonance.
Case 3

This patient was a 32-year-old woman who presented with hydrocephalus in association with an anterior third ventricular tumor that appeared to extend into the hypothalamus. An endoscopic biopsy procedure and septostomy were performed that confirmed the diagnosis of neurocytoma. She was referred to us for stereotactic radiosurgery as primary management of the tumor. A combination of one 8-mm and one 4-mm isocenter was used to deliver a dose to the tumor margin of 20 Gy and a maximum dose of 40 Gy. The small tumor volume measured 0.325 cm³. Now, 42 months after radiosurgery, imaging studies have demonstrated a reduction in tumor size. No neurological, visual, or endocrinological problems have developed, and she has returned to work.

Case 4

This 10-year-old boy had undergone resection of a hemorrhagic thalamic and third ventricle neurocytoma at the age of 6 years. He recovered to a normal neurological condition but suffered a tumor recurrence 4 years later. He underwent stereotactic radiosurgery for a 1.3-cm³ tumor in which three 8-mm and two 4-mm isocenters were used. A dose to the tumor margin of 16 Gy and a maximum dose of 32 Gy was administered. At 38 months he remains well, and serial imaging studies have demonstrated marked tumor regression.

Discussion

Central neurocytomas are uncommon neoplasms that were originally described in 1982.9 They predominantly occur in the lateral ventricles near the foramen of Monro29 and arise from the septum pellucidum or the walls of the lateral ventricles. Left ventricular tumors are more common than the right ventricular tumors, and they occur less commonly in the third ventricle. Fourth ventricular extension has also been described.29 Patients typically present with symptoms of obstructive hydrocephalus after fewer than 8 months. Extraventricular neurocytomas have also been reported.14,16,25 These tumors are of a neuronal nature, which is well demonstrated by immunohistochemical staining with synaptophysin.9,11,26,27 Ultrastructural studies reveal synaptic structures. In rare cases, mature ganglion cells are present.26 These tumors are characterized by clear cells within a vascular meshwork and are reminiscent of oligodendrogliomas or ependymomas but easily distinguished from these glial origin tumors by the lack of immunoreactivity for glial fibrillary acidic protein. In the majority of tumors the proliferation index is low.7 Extraventricular neurocytomas span a histological spectrum from purely neuronal to mixed neuronal–glial tumors.8 Neurocytomas are typically a disease of young adulthood, occurring in the second and third decades of life. The lesions are slow growing and are primarily treated by excision. These tumors are generally associated with a good prognosis but occasionally are more aggressive in nature.5,12,17,18,22,24–26,29 Patients with anaplastic features at the time of initial resection have undergone additional radiotherapy in certain cases.5,12 Most neurocytomas can be cured by complete excision, although recurrences have been reported.25 The natural history of these lesions can be quite variable. Subtotal resections are not always followed by immediate recurrence, and for that reason some surgeons simply follow patients without further treatment after partial removal. In an attempt to predict tumor behavior by using histological techniques, some investigators have used MIB-1 labeling indices or signs of histological atypia.7,28

Many times, patients with incomplete resections undergo radiotherapy, as do some patients with recurrent tumors.5,13,19 Kim, et al.13 compared results in seven patients who underwent postoperative radiotherapy with eight who did not. Two of the latter group developed recurrences at 8 and 21 months, whereas in patients who underwent

Fig. 1. Case 1. Axial MR images with contrast enhancement demonstrating a residual neurocytoma (left) in a 16-year-old woman and 18 months later (center) demonstrating significant tumor regression. The radiosurgical dose plan is also shown (right). Five 14-mm isocenters were used; the 50% isodose line was targeted to the tumor margin.
radiotherapy the disease remained controlled. Similarly, Schild, et al., found no recurrences in their series of patients who underwent radiation therapy, whereas tumors recurred in half the patients if left untreated following surgery. Thus, radiotherapy has been shown to be effective for neurocytoma, with improved 5-year local control rates in patients whose tumors were irradiated after subtotal resection. The most common recurrence is local, but craniospinal dissemination has also been reported. Chemotherapy has also been used at recurrence.

Radiosurgery may offer therapeutic advantages over fractionated radiotherapy for patients with neurocytoma. First, most tumors are fairly well circumscribed and can be confidently irradiated with a precise radiosurgical dose plan. The tumor border is better defined within the ventricle than within parenchyma, although different MR sequences and use of contrast enhancement are useful. Such definition of the tumor border is not possible in patients with more infiltrative glial neoplasms, in which radiosurgery is rarely performed as the sole treatment. By avoiding larger-field brain irradiation, we hoped to spare regional structures such as the fornix, thalamic and basal ganglia nuclei, and deep frontal lobe from late radiation effects. Additionally, the biological effect of a single fraction of radiation is much higher than when given in divided doses. This should be associated with a more potent cytotoxic effect and may be the reason for the dramatic tumor regression seen in this series. Nevertheless, Maruyama, et al., hypothesized that neurocytomas may not be radiosensitive, because these authors did not find increased fluorodeoxyglucose uptake on positron emission tomography within hours after 24-Gy maximum dose radiosurgery in one patient. This was in contrast with a series of patients with brain metastases in whom acute changes were noted on positron emission tomography scans. This finding, however, is difficult to interpret because of their limited experience with this tumor and the bulk of data that argues for this tumor being responsive to radiation therapy. Cobery, et al., and Anderson, et al., each reported four patients who underwent gamma knife surgery with similar results. A marked reduction in tumor size was found in all four patients in each study. Pollock and Stafford added a case to the literature, again showing tumor regression with 3-year follow up. Extended follow up of patients from all these series will be necessary to identify the long-term results of radiosurgery for this tumor.

Little is known about the radiation tolerance of the fornices, corpus callosum, and other septal structures. Some data exist regarding the tolerance of the optic nerves and chiasm, and most medical centers try to limit the radiation dose to those structures to less than 8 or 9 Gy. Overall, most parenchymal tumors receive doses to the margin in the 10 to 20-Gy range, depending on volume, tumor type, past irradiation history, other available treatment options, and perceived risks. In this series the dose received by the fornix was kept below 15 Gy. In the third patient a higher tumor dose was selected because the small tumor volume allowed for a steep dose fall-off into the surrounding brain.

Although most patients may fare well following resection of a neurocytoma, radiosurgery may prove to be an effective treatment for residual or recurrent tumors. Obtaining a histological diagnosis before treatment remains important. For tumors suspected to be neurocytomas on imaging studies, but where brain location may prohibit resection, stereotactic biopsy procedures followed by radiosurgery may be a worthwhile strategy.

References


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