The Management of Central Neurocytoma
Radiosurgery

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KEYWORDS

• Central neurocytoma • Radiosurgery • Stereotactic radiosurgery • Gamma Knife • Linear accelerator

KEY POINTS

- Central neurocytomas are rare, usually benign, tumors that typically present with hydrocephalus and usually require surgical resection.
- Surgical resection frequently is incomplete, and even when complete, tumor recurrence is not uncommon.
- Stereotactic radiosurgery is a highly selective and conformal technique that avoids the inconvenience and many of the toxicities of conventional radiation therapy.
- Stereotactic radiosurgery appears to be a safe and effective primary or adjuvant treatment for all but widely disseminated neurocytoma.
- Most patients will experience notable tumor regression after SRS, but careful long-term surveillance is required to detect complications and recurrence.

INTRODUCTION: NATURE OF THE PROBLEM

Central neurocytoma was first described as a distinct pathologic entity in 1982 by Hassoun and colleagues[1] in a report of 2 cases. It is a rare lesion that comprises only 0.1% of adult primary central nervous system tumors.[2] Patients are typically young adults and often come to clinical attention due to signs or symptoms related to obstructive hydrocephalus.[3,4] On MRI studies, central neurocytomas appear as well-circumscribed, heterogeneously enhancing, intraventricular, "bubbly" masses that are often attached to the septum pellucidum or another area of ependymal surface (**Fig. 1**).[5]

These tumors are histologically and molecularly glioneuronal tumors that are classified as World Health Organization grade II. Although 75% or more of central neurocytomas are histologically benign, they can be designated as atypical on the basis of a proliferation index of greater than or equal to 2% (via Ki-67 or MIB-1 labeling) or histologic features including areas of necrosis, vascular proliferation, and frequent mitoses.[6–8] A high proliferative index via MIB-1 labeling is associated with a more aggressive biological behavior that can include episodes of intraventricular hemorrhage and cerebral spinal fluid (CSF) dissemination.[9,10] **Table 1** indicates the common pathologic and clinical features of central neurocytomas contrasted with less-frequent variants.

The first-line treatment for a young adult with an intraventricular mass, especially in the setting of

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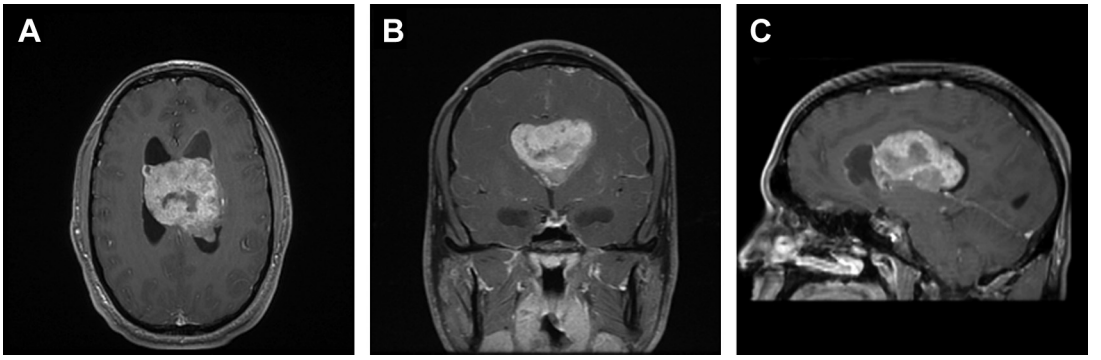

Fig. 1. Select axial (*A*), coronal (*B*), and sagittal (*C*) contrast-enhanced MRI images demonstrating the typical appearance of a central neurocytoma showing a well-circumscribed, heterogeneously enhancing, "bubbly" appearing intraventricular lesion. Note the associated hydrocephalus with very enlarged temporal horns on the coronal image.

symptomatic obstructive hydrocephalus, is surgical resection. This allows for the establishment of a tissue diagnosis and management of hydrocephalus. A meaningful proportion of patients ultimately require ventriculoperitoneal shunt placement (ie, 22.8% in a recent report by Qian and colleagues).[11] However, surgery is not without complications, the most frequent being cognitive and memory dysfunction related to forniceal injury, and hemiparesis.[9,12–15] Gross total resection is the goal of microsurgery, but this is only variably achieved (60%–100%) due to the tumor's proclivity to be highly vascular and adherence to local critical structures.[3,4,11,16,17] Tumor recurrence is relatively common (~20%), even after what is felt to be complete resection.[18] Residual or recurrent tumor can lead to intracerebral hemorrhage, neurologic deficits, and hydrocephalus, and typical neurocytomas can transform into atypical lesions and disseminate throughout the CSF.[6,19–22]

In the face of residual, recurrent, and atypical neurocytomas, conventional fractionated radiation therapy (RT) became the initial adjuvant therapy of choice. Conventional RT has proven to increase the rate of local control after incomplete resections, establishing neurocytomas as fairly radiosensitive[18]. However, the long-term or delayed consequences of conventional RT are increasingly well understood and include leukoencephalopathy, cognitive dysfunction, and secondary malignancies.[23–26] Some clinicians have recommended observation of a neurocytoma with a potentially indolent clinical course to avoid the toxicity of conventional RT.[27,28]

STEREOTACTIC RADIOSURGERY AS A THERAPEUTIC OPTION

Stereotactic radiosurgery (SRS) has evolved as a minimally invasive adjuvant or primary treatment

Table 1
Common and less frequent features of central neurocytoma

Features	Common	Variants
Location	Lateral and third ventricles near the foramen of Monro, midline	Extraventricular *(cerebral neurocytoma)* Cerebellar liponeurocytoma
Presentation	Due to symptoms of obstructive hydrocephalus	Incidental
Grade	Benign (75%)	Atypical (25%)
Proliferative index	MIB-1 <2%	MIB-1 ≥2%
Behavior	Localized	Hemorrhagic Cerebrospinal fluid dissemination Malignant transformation
Molecular	Synaptophysin positive	Synaptophysin negative, neuronal specific enolase positive Focal glial fibrillary acidic protein positive
Prognosis	Good	Poor