The Management of Incidental Central Neurocytoma

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KEYWORDS

• Central neurocytoma • Incidental • Management • Radiosurgery • Surgery

KEY POINTS

- Although central neurocytoma is rare, the number of incidentally found tumor is increasing because of improvement in neuroimaging for early detection of the lesion in recent years.
- Although surgical resection is a well-accepted primary treatment of central neurocytoma, upfront stereotactic radiosurgery (SRS) is also a good alternative treatment of asymptomatic or incidentally found tumor.
- The tumor control rates of SRS for central neurocytoma are about 90% at 5-year and 80% at 10-year follow-up according to several long-term retrospective studies.
- The optimal radiosurgery dose for central neurocytoma seems comparable with the dose used in other benign brain tumors; however, the tumor volume reduction rate after SRS is significantly higher in central neurocytoma, compared with those in other intraventricular tumors.
- Based on the characteristic imaging findings for diagnosis in central neurocytoma before treatment, and the characteristic tumor response after treatment, SRS can serve as a primary treatment of certain asymptomatic, incidentally found central neurocytomas.

INTRODUCTION

Central neurocytomas are neuronal tumors that usually arise from the septum pallucidum or the wall of lateral ventricles. Most of these tumors are benign and classified as grade II tumors under the Word Health Organization's classification for tumors in nervous system, although some atypical tumors have also been reported.[1] Because central neurocytomas are intraventricularly located and usually behave in an indolent clinical course initially, their symptoms and signs mostly come from late tumor growth and blockage of cerebrospinal fluid pathways. Some of the central neurocytomas are even found incidentally, either by routine imaging examinations during physical check-up or in neuroimaging studies for other reasons.

The treatment of choice for a large, symptomatic central neurocytoma is surgical resection. The outcomes of microsurgical removal depend on the extent of the resection and the histologic grading.[1–3] Radiation therapy, including fractionated conventional radiotherapy (FCRT) and stereotactic radiosurgery (SRS), is usually used for residual or recurrent tumors. Tumor control rates after SRS in cases of subtotal tumor removal for central neurocytoma can be 90% in 5-year and larger than 80% in 10-year follow-up.[4,5]

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Because the patient population is rare, the optimal management of an asymptomatic or incidentally found central neurocytoma has not been well established. This article reports the current treatment options with clinical evidence.

RADIOLOGIC FINDINGS AND DIFFERENTIAL DIAGNOSIS OF CENTRAL NEUROCYTOMA

In the neuroimaging study, central neurocytomas usually present as well-demarcated, lobulated masses in the lateral ventricle.[6] On computed tomography (CT) scan, they typically appear to be isodense to hyperdense with various degrees of heterogeneity. Hypodense areas are usually related to cystic degeneration, whereas hyperdense area may be related to calcification.[7,8] Like other intraventricular tumors, central neurocystomas are typically attached to the ventricular wall, mostly superior and lateral walls of ventricles.[7] Mild to moderate contrast enhancement is generally seen.[3] On MRI, central neurocytomas are typically heterogeneous isointense or slightly hypointense on T1-weighted MRI, and isointense to hyperintense on T2-weighted images. The enhancement after gadolinium-DTPA injection varies from mild to strong.[6–9] The existence of calcification, hemorrhage, and intratumoral cyst leads the variability of MRI presentation. Nevertheless, the overall imaging features of a central neurocytoma are quite characteristic. On MRI, this tumor usually presents as an intraventricular, lobular, and "bubbly" mass lesion.[5]

The diagnosis of a central neurocytoma should be meticulously differentiated with several other intraventricular tumors, such as subependymal giant cell astrocytoma (SGCA), oligodendroglioma, ependymoma, papilloma, and meningioma.[6,10,11] The focus of the differential diagnosis can be narrowed considerably by knowing the patient's age, the exact location and extention of the tumor in or around ventricles, presence of the cyst component, and the density on precontrast CT scan.[6,10,11] On CT scan, central neurocytomas are either isodense or hyperdense, whereas subependymomas and SGCAs are usually hypodense. After the contrast injection, central neurocytomas may show mild to moderate enhancement, whereas subependymomas appear with either no or scare enhancement.[11] Calcification with a punctate appearance may be observed in central neurocytomas or SGCAs. For intraventricular meningiomas or papillomas, they typically demonstrate a strong, homogenous enhancement on CT and MRI.

Some special MRI examinations including magnetic resonance spectroscopy and diffusion-weighted MRI have emerged as additional tools for the diagnosis of central neurocystoma. In magnetic resonance spectroscopy, central neurocytomas present as high glycine, increased choline and alanine, and decreased *N*-acetylaspartate and creatine/phosphocreatine peaks.[12,13] On diffusion-weighted images, the tumor has a heterogeneous hyperintense appearance when compared with the contralateral parietal lobe white matter. The mean value of normalized apparent diffusion coefficient of central neurocytomas had been measured around 0.63 ± 0.05.[13]

MICROSURGICAL RESECTION FOR CENTRAL NEUROCYTOMA

Surgical resection is the treatment of choice for symptomatic central neurocytomas, and complete resection usually provides the best outcome of patients.[2,14–20] Although there is no report directly from asymptomatic or incidental cases, 3% (12 of 362) of patients with asymptomatic central neurocytomas were found in several large surgical series (**Table 1**).

The common surgical approach used for microsurgical resection of central neurocytoma is through transcallosal-transventricular or transcortical-transventricular routes.[7,14] Given that the extent of resection is the most important prognostic factor of the tumor control rate and survival, several reports suggest that the only way to cure the disease is to completely resect the tumor. In a meta-analysis of 438 patients with a central neurocytoma,[21] the tumor control rate at 10 years after gross total resection was 74%, and 35% in patients with subtotal resection. The patients' 10-year survival for total and subtotal resections were 99% and 82%, respectively. Some large surgical series (>40 cases) demonstrated a significantly better outcome in patients with gross total tumor resection.[2,21]

However, the capability of the gross-total resection ranged between 33% and 78% in the reported series despite the advancement of modern microsurgical techniques in the past 15 years. Multivariate logistic regression showed that patients of younger age and with the absence of signs of raised intracranial pressure would more likely make a complete resection. In addition, these two factors would have a protective effect against postoperative complications.[18]

Table 1 lists the possible complications related to surgical resection reported in the major series of central neurocytomas. These complications include persistent hydrocephalus, hemiparesis, postoperative hemorrhage or hematoma in the tumor bed, aphasia, memory impairment, new onset of seizure, infection, and rarely hypothalamic