Clinical Outcome and Quality of Life After Treatment of Patients with Central Neurocytoma

Chae-Yong Kim, MD, PhD[a], Dong Gyu Kim, MD, PhD[b,*], Jin-Deok Joo, MD[a], Yong Hwy Kim, MD[a]

KEYWORDS

• Central neurocytoma • Functional outcome • Quality of life • Radiation therapy • Radiosurgery • Surgery

KEY POINTS

- Complete resection of a central neurocytoma (CN) is associated with longer progression-free survival; whether complete resection correlates significantly with overall survival remains controversial.
- The long-term outcome of radiosurgery seems to surpass that of conventional radiotherapy.
- A tailored multimodal treatment could be an appropriate next step in the treatment of CN.

INTRODUCTION

Since the first report of central neurocytoma (CN) in 1982 by Hassoun and colleagues, the diagnosis of this new entity has been gradually increasing with the accumulation of information regarding its clinical, radiologic, and histopathologic features.[1,2] Approximately 500 cases have been reported to date, and the overall incidence of CN is 0.1% to 0.5% among all primary brain tumors.[3–6] The mainstay of initial treatment for CN has been total operative resection whenever possible. Several previous reports have demonstrated that complete extirpation of CN is correlated with better local tumor control and longer overall survival compared with incomplete resection.[7–9] However, CNs cannot be resected completely in more than one half of all patients, and a number of recurrences, even after complete resection, have been reported.[1,7,8] Furthermore, because CN has a deep-seated intraventricular location and a close proximity to critical structures, such as the fornix and thalamus, surgical morbidity and mortality are not negligible.[1]

Recently, radiosurgery has been found to be an important primary or secondary treatment for many intracranial tumors. Similarly, for the treatment of CN, radiosurgery seems to be a highly attractive treatment modality because of the radiosensitive nature of the tumor and anatomic factors, such as well-demarcated borders and an intraventricular location.[1,10] Most radiosurgical studies for CN have reported favorable outcomes; however, these results were based on case series that included a limited number of patients and short follow-up periods. A thorough investigation of the long-term outcomes of CN after various treatments is required to establish the optimal management strategies for these rare tumors.[1,3] This article reviews the long-term results of various treatments and management strategies with

[a] Department of Neurosurgery, Seoul National University Bundang Hospital, Seoul National University College of Medicine, 101 Daehangno, Jongno-gu, Seoul 110-744, Korea; [b] Department of Neurosurgery, Seoul National University Hospital, Seoul National University College of Medicine, 101 Daehangno, Jongno-gu, Seoul 110-744, Korea
* Corresponding author.
E-mail address: gknife@plaza.snu.ac.kr

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reference to functional outcome and quality of life of patients with CNs.

CLINICAL OUTCOME IN ACCORDANCE WITH TREATMENT MODALITIES

Surgical Resection

The main objectives of surgery are to perform the maximum resection with minimal neurologic impairment, to establish and/or restore cerebrospinal fluid pathways, and to provide tissue for the definitive pathologic diagnosis of CN.[2,11] Most CNs are intraventricular and do not invade surrounding parenchyma, allowing for the potential of gross total removal (GTR) using microsurgical techniques. However, concurrently, CNs are situated close to critical structures, such as the fornix and thalamus; thus, aggressive and excessive resection necessarily poses a serious risk of neurologic deterioration under certain circumstances.[1,2,4] Resectability typically depends on tumor size, location, extent of lesion, adherence to surrounding critical structures, vascularity, and the surgeon's experience.[4,11]

In a meta-analysis of 310 patients with CN by Rades and Fehlauer,[12] local control at 3 and 5 years for GTR was 95% and 85%, compared with 55% and 46% for subtotal resection (STR), respectively. Five-year survival rates were 99% and 86% for GTR and STR, respectively. For the treatment of atypical CN, Rades and colleagues[13] showed that the 3- and 5-year local control rates for GTR (n = 15) were 73% and 57% compared with 60% and 31% for STR (>25% resection), respectively, and survival rates after 5 years were 93% and 57% after GTR and STR (>25% resection), respectively. There is no doubt that the maximal surgical resection with minimal neurologic deficit plays the most important role in the management of CN. Although most CNs are benign, recurrence or progression after STR and GTR have been widely reported.[12,14–16] The complete resection of a CN has been significantly associated with longer progression-free survival in several studies; however, whether complete resection correlates significantly with overall survival remains controversial.[2,3,6,9,10,16–18]

As for postoperative complications, surgical morbidity and mortality are not negligible. Qian and colleagues[22] reported that in a surgical series of 92 patients with CN, the GTR rate was 70.7% but postoperatively, 62% had hydrocephalus, 26.1% had decreased memory, and 29.3% had a Karnofsky Performance Scale (KPS) score of 70 or less. Lubrano and colleagues[19] reported a surgical series of 92 cases of CN, emphasizing the maximal surgical resection with regard to the tendency toward transience of postoperative neurologic deficits (**Table 1**). The complication rate differs in each report, which could result from disagreements regarding what constitutes a complication. However, it is clear that a relatively high rate of postoperative complications, such as memory impairment and a decline in the KPS score, are significantly related to the quality of life of the treated patients. Currently, a careful reappraisal is being conducted concerning the advancement of adjuvant therapies, such as radiation therapy (RT) and stereotactic radiosurgery, to determine whether the maximal surgical resection, which risks postoperative neurologic deficits and deterioration of the quality of life of patients, is necessary.[1,4,12,20]

Conventional Radiotherapy

Conventional RT has been applied frequently for the management of residual or recurrent CN. However, side effects of radiation have been documented, including long-term radiation-induced toxicity and a secondary malignancy similar to a radiation-induced anaplastic astrocytoma after treatment of a CN.[10,23] Currently, most studies agree that RT is not necessary after GTR owing to the indolent clinical course of the disease and a potential for radiation-induced toxicity.[11,12,20] Rades and colleagues found that RT after STR improved local control significantly from 39% to 94% and survival from 82% to 100% at 10 years; this difference in survival was not significant ($P = .16$).[11,20] Meanwhile, apart from benign CNs, atypical and extraventricular neurocytomas, which have typical features but a poorer prognosis, have generated interest in the use of radiation therapies.[11] For atypical neurocytomas treated with STR, RT has been shown to improve local control from 5% to 65% at 5 years and survival from 46% to 69% at 5 years, and several studies have reported that conventional RT seemed to play a role, to a certain degree, in the treatment of extraventricular neurocytoma.[18,24,25] However, irradiation has not improved the KPS scores of patients after STR, and some authors have not opposed the use of RT after STR.[11,18] After all, radiotherapy as a primary treatment option for CN has not been thoroughly evaluated, and the necessity of adjuvant RT remains controversial. It is obvious that long-term radiation-induced toxicities resulting in cognitive dysfunction and memory impairment significantly affect the quality of life of the patients. Unless RT is proven to be advantageous for improving the KPS score of patients, a careful decision should be made regarding the use of RT as a primary or secondary treatment for CN.