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# Skull base chondrosarcoma

Chondrosarcomas of the skull base are rare compared with other skull base tumors but are an important differential diagnosis as surgical resection and management are affected by the preoperative diagnosis.

#### Petroclival chondrosarcomas

Endonasal endoscopic approach for resection of petroclival chondrosarcomas between 2010 and 2014, 8 patients (4 men and 4 women) underwent endonasal endoscopic operations to resect petroclival chondrosarcomas at 2 institutions. The patients' mean age was 44.8 years (range 30-64 years). One of the patients had previously undergone radiation therapy and another a staged craniotomy. Using volumetric software, an independent neuroradiologist assessed the extent of the resections on MRI scans taken immediately after surgery and at the 3-month follow-up. Immediate complications and control of symptoms were also recorded. In addition, the authors reviewed the current literature on surgical treatment of chondrosarcoma.

The mean preoperative tumor diameter and volume were 3.4 cm and 9.8 cm(3), respectively. Six patients presented with cranial neuropathies. Endonasal endoscopic surgery achieved > 95% resection in 5 of the 8 patients and < 95% resection in the remaining 3 patients. One of the 6 neuropathies resolved, and the remaining 5 partially improved. One instance of postoperative Cerebrospinal fluid fistula required a reoperation for repair; no other complications associated with these operations were observed. All of the patients underwent adjuvant radiotherapy.

According to the experience of Moussazadeh et al. the endoscopic endonasal route is a safe and effective approach for the resection of appropriately selected petroclival chondrosarcomas <sup>1)</sup>.

## **Treatment**

Complete resection of skull base chondrosarcomas offers the potential for a durable, or even lifelong, cure and is best achieved at the first surgery.

When a skull base chondrosarcoma is located at the upper clivus and in the interpeduncular cistern and invading laterally toward the petrous apex and cavernous sinus, the traditional approaches, ie, endonasal endoscopic approach or middle fossa approaches, are not adequate for the exposure and resection. The transcavernous sinus approach has been utilized to remove tumor from the cavernous sinus and as a corridor to the interpeduncular cistern and upper clivus, originally described for the clipping of basilar apex aneurysms.

Essayed et al. presented a case of chondrosarcoma centered in the upper clivus and eroding the right posterior clinoid, analogous to the location of a giant basilar apex aneurysm. Detailed study of the tumor extension, bony invasion, and relationship with neuroanatomy dictated the most effective surgical approach.

Neuronavigation and intraoperative magnetic resonance imaging (MRI) facilitated the gross total resection of the tumor in the Advanced Multimodality Image-Guided Operating (AMIGO) suite. Achieving a gross total tumor removal of this World Health Organization (WHO) grade I

chondrosarcoma, adjuvant irradiation can be withheld and the patient monitored with serial imaging. The patient did well after the surgery <sup>2)</sup>.

Gamma Knife radiosurgery for skull base chondrosarcomas has a favorably low risk of radiation-induced adverse effects (RAEs) and could be a reasonable therapeutic option for SBC in multimodality management. A sufficient GKRS prescription dose is necessary for higher local control rates (LCRs). Histological grading and subtype evaluations are important for excluding exceptional SBCs. Patients with conventional SBCs have a long life expectancy and should be observed for life after treatment <sup>3)</sup>.

### **Outcome**

Skull base chondrosarcoma outcome.

### Case series

Data of patients who underwent GKRS for SBCs at Gamma Knife centers in Japan were retrospectively collected. Patients without a histopathological diagnosis and those who had intracranial metastases from extracranial chondrosarcomas were excluded. Histologically, grade III and some nonconventional variants were identified as aggressive types. The cumulative local control rates (LCRs) and disease-specific survival (DSS) rates were calculated using the Kaplan-Meier method. Factors potentially affecting the LCR were evaluated using the Cox proportional hazards model for bivariate and multivariate analyses. The incidence of radiation-induced adverse effects (RAEs) was calculated as crude rates, and factors associated with RAEs were examined using Fisher's exact test.

Fifty-one patients were enrolled, with a median age of 38 years. Thirty patients (59%) were treated with upfront GKRS for residual SBCs after resection (n = 27) or biopsy (n = 3), and 21 (41%) underwent GKRS as a salvage treatment for recurrence. The median tumor volume was 8 cm3. The overall LCRs were 87% at 3 years, 78% at 5 years, and 67% at 10 years after GKRS. A better LCR was associated with a higher prescription dose (p = 0.039) and no history of repeated recurrence before GKRS (p = 0.024). The LCRs among patients with the nonaggressive histological type and treatment with  $\geq$  16 Gy were 88% at 3 years, 83% at 5 years, and 83% at 10 years. The overall survival rates after GKRS were 96% at 5 years and 83% at 10 years. Although RAEs were observed in 3 patients (6%), no severe RAEs with Common Terminology Criteria for Adverse Events grade 3 or higher were identified. No significant factor was associated with RAEs.

GKRS for SBCs has a favorably low risk of RAEs and could be a reasonable therapeutic option for SBC in multimodality management. A sufficient GKRS prescription dose is necessary for higher LCRs. Histological grading and subtype evaluations are important for excluding exceptional SBCs. Patients with conventional SBCs have a long life expectancy and should be observed for life after treatment <sup>4)</sup>.

1)

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