LIOSARCOMAS are bimorphic intraaxial tumors consisting of malignant glial and sarcomatous components. They account for 2 to 8% of all malignant gliomas, and their most common location has been reported to be the temporal lobe in 37.5 to 44% of the patients. However, skull base involvement is highly unexpected, and we know of only one case that has been reported in the literature.

In the present report, we describe a case of gliosarcoma with significant infratemporal fossa extension.

Case Report

History and Examination. This 55-year-old man presented with a 1-month history of severe progressive headache. Neurological examination was unremarkable except for bilateral papilledema. Magnetic resonance imaging revealed a 6-cm right temporal mass with extension into the infratemporal fossa. The patient underwent a right frontotemporal craniotomy together with drilling of the sphenoid ridge and middle fossa floor. The tumor consisted of intraaxial, intracranial as well as extradural, and extracranial components with extension to the posterolateral wall of the sphenoid sinus. It had a relatively well-circumscribed dissection plane. Gross-total resection was achieved, and the middle fossa floor was reconstructed using a rotated temporalis muscle flap. The postoperative course was uneventful except for hypesthesia in the distribution of the maxillary division of the right trigeminal nerve. The histopathological diagnosis was consistent with gliosarcoma. Radiotherapy and chemotherapy consisting of temozolomide were administered subsequently, and the patient was recurrence free 12 months after his initial diagnosis.

In the presence of a mass lesion with both intraaxial and extracranial involvement, gliosarcoma should be considered among the differential diagnoses. Aggressive resection should be attempted, including the use of skull base surgical techniques to ensure an optimal outcome. The effect of skull base involvement to the overall treatment and outcome of patients with gliosarcomas would be difficult to determine given the rare occurrence of these lesions in such locations.

KEY WORDS • gliosarcoma • infratemporal fossa • skull base

Gliosarcomas are bimorphic intraaxial tumors consisting of malignant glial and sarcomatous components. They account for 2 to 8% of all malignant gliomas, and their most common location has been reported to be the temporal lobe in 37.5 to 44% of the patients. However, skull base involvement is highly unexpected, and we know of only one case that has been reported in the literature. In the present report, we describe a case of gliosarcoma with significant infratemporal fossa extension.

Case Report

History and Examination. This 55-year-old man presented with a 1-month history of severe progressive headache. Neurological examination was unremarkable except for bilateral papilledema. Magnetic resonance imaging revealed a heterogeneously enhancing 6-cm right temporal mass abutting the anteroinferior cavernous sinus, with extension into the infratemporal fossa and lateral sphenoid sinus (Fig. 1). The lesion’s overall appearance suggested a possible extraxial origin, with differential diagnoses including aggressive meningioma, sarcoma, and metastatic carcinoma.

Treatment. The patient underwent a right frontotemporal craniotomy with drilling of the sphenoid ridge and middle fossa floor. The tumor was easily visualized along the middle fossa floor. After dural opening, a partial lateral temporal lobectomy was performed for adequate intraaxial tumor exposure. The tumor had yellow and reddish-brown areas with an irregular consistency but was firm in general and showed a relatively well-circumscribed dissection plane. Frozen-section analysis suggested a spindle-cell malignancy. Extracapsular dissection was performed after internal debulking of the tumor. The entire intradural portion of the lesion was removed, and the involved dura mater was resected with a wide margin. Subsequently, extradural expo-
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sure of the cavernous sinus was performed. The foramen ovale, which was dramatically enlarged by the tumor, and the foramen rotundum were drilled and further enlarged. The maxillary division of the trigeminal nerve (V2) was sacrificed because it was invaded by tumor. The lesion was followed into the infratemporal fossa where it involved the lateral pterygoid muscle and then extended into the postero-lateral sphenoid sinus. Gross-total removal was achieved. The exposed sphenoid sinus and drilled middle fossa floor were repaired by rotating the temporalis muscle as a vascularized flap. The temporal dural defect was then repaired using DuraGen (Integra LifeSciences, Plainsboro, NJ).

Postoperative Course and Immunoreactivity Studies. The patient’s postoperative course was uneventful except for hypesthesia in the right V2 distribution. A lumbar subarachnoid drain was kept in place for 4 days. Postoperative MR imaging confirmed gross-total resection of the lesion.

The morphological features of the lesion were consistent with a diagnosis of gliosarcoma (World Health Organization Grade IV). Areas of the tumor resembled GBM (Fig. 2A and B) and were marked by prominent cellularity and nuclear pleomorphism, readily identifiable mitotic activity, vascular proliferative changes, and necrosis. These areas demonstrated positive immunoreactivity for S100 protein and GFAP antibodies. Reticulin was observed in the gliomatous regions of the lesion with significant vascular proliferative changes. Other areas of the tumor were marked by spindle-cell proliferation, which was reticulin-rich and negative for GFAP and S100 protein (Fig. 2C and D). Neither component of the tumor demonstrated immunoreactivity for antibodies to Melan A, cytokeratin, cell adhesion molecule 5.2, epithelial membrane antigen, or p53. Results of

Fig. 1. Preoperative axial (left) and coronal (right) Gd-enhanced T₁-weighted MR images demonstrating a right temporal mass involving the cavernous sinus and infratemporal fossa and extending into the sphenoid sinus.

Fig. 2. Photomicrographs revealing cellular components of the gliosarcoma. A: Gliomatous area of the tumor with features of a GBM. B: Gliomatous area with positive immunoreactivity for GFAP. C: Sarcomatous area showing spindle-cell proliferation. D: Sarcomatous area rich in reticulin. H & E (A and C), original magnification × 200 (A–D).
molecular studies (fluorescence in situ hybridization) demonstrated no evidence of epidermal growth factor receptor amplification or losses on chromosomes 1p and 19q. The patient was subsequently treated with fractionated whole-brain radiation, followed by 5 days of temozolomide every 28 days for six cycles. He was recurrence-free at the 12-month follow-up evaluation (Fig. 3).

**Discussion**

Gliosarcomas consist of glial and sarcomatous cell populations—the glial component being GBM and the sarcomatous element having the features of any sarcoma type but most often those of the malignant fibrous histiocytoma. It has been initially suggested that gliosarcomas should be regarded as GBM with proliferating endothelium having sarcomatous features. Vascular smooth-muscle and multipotent stem cells have also been proposed in the context of histogenesis. However, data from recent studies have shown common genetic features for both cell populations, and sarcomatous areas are believed to represent a phenotypic change of GBM cells instead of a separate malignancy.

On MR images, gliosarcomas appear as an extraaxial lesion despite the intraaxial origin of the tumor, as in the present case. The lesions can appear well-circumscribed with focal edema as well as a broad dural base and dural enhancement, mimicking meningiomas. Despite the tumor’s tendency to be superficial and adjacent to the dura, we know of only one other case of gliosarcoma with significant skull base extension: the tumor extended into the maxillary sinus through the middle fossa floor at the time of tumor recurrence. In the case in the present study, involvement of the infratemporal fossa was apparent at the time of initial presentation.

Intraoperatively, gliosarcomas (like meningiomas) can be firm, well-circumscribed, and adherent to the dura, or they (like astrocytomas) can exhibit irregular boundaries with the surrounding cerebral tissue, which can lead to limited resection. In the present case, intraoperatively observed features were more similar to those of aggressive meningiomas. From a surgical perspective, another interesting feature of this case was that the posterolateral wall of the sphenoid sinus was eroded, and aggressive removal of the tumor led to direct communication between the sinus and intracranial cavity. Temporalis muscle was rotated as a vascularized flap and laid over the middle fossa floor to prevent postoperative cerebrospinal fluid leakage. Although the mandibular division of the trigeminal nerve (V3) could be spared despite significant expansion of the foramen ovale, the V2 was infiltrated by the tumor and had to be sacrificed.

Results of clinical studies have shown that patients with gliosarcoma and GBM have similar outcomes and therefore an identical treatment has been recommended for both pathological entities. Patients harboring gliosarcoma with imaging and intraoperative features suggestive of meningioma, or with more sarcomatous components histologically, may have a prolonged survival. In a recent study, the median recurrence time in patients with predominantly gliomatous components was reported to be 53 weeks compared with 62 weeks in patients with predominantly sarcomatous features. The patient in the present case, having completed radiotherapy and chemotherapy, was recurrence-free at the 12-month follow-up evaluation.

**Conclusions**

In the presence of a mass lesion with both intracranial and extracranial involvement, gliosarcomas should be considered among the differential diagnoses. Aggressive tumor removal and adequate reconstruction can be achieved by using basic skull base techniques. We believe that the effect of skull base involvement on the treatment and outcome in patients with these tumors would be difficult to determine given the extremely rare occurrence of these lesions in such locations.

**References**


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Fig. 3. Axial (left) and coronal (right) Gd-enhanced T1-weighted MR images obtained at the 1-year follow-up evaluation with no evidence of recurrent tumor. The temporal muscle was rotated over the middle fossa floor to eliminate communication with the sphenoid sinus (arrows).
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