Resection of Anaplastic Meningioma via Combined Craniofacial Approach

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INTRODUCTION

Meningiomas, which represent one of the largest subgroups of intracranial neoplasms, are usually slowly growing tumors, arising from the arachnoid, and account for 13 to 36% of all intracranial tumors [1,2]. Meningiomas are classified by the World Health Organization (WHO) into three grades; Grade 1 meningiomas are benign lesions and are found in 90% of all cases whereas Grade 2 meningiomas—also called atypical meningiomas—occur in 5 to 7% of all cases with a higher rate of recurrence compared to grade 1 lesions [3–6]. Grade 3 meningiomas (Papillary, Rhabdoid, and Anaplastic) are diagnosed in only 1–3% and are very aggressively growing lesions with the highest recurrence rates after surgical removal of 50% to 75% [3,4,6–9]. Surgical resection is considered the best treatment for most patients with meningioma [10]. Multiple surgical resections may be required in patients with recurrent meningioma. When repeated operations are required, the chance of cure is significantly reduced. Furthermore, multiple operations also carry an increased risk of intracranial infection and other postoperative complications. In rare instances, a histological malignant transformation may occur and make the prognosis even worse [11–13].

We report a case of frontal base meningioma who underwent two previous craniotomies at another institution with grade 1 meningioma. The patient underwent surgery at our institution with histopathological results of Anaplastic Meningioma. There was a malignant transformation during multiple recurrences.

CASE PRESENTATION

A 41-year-old female presented with proptosis of both orbits with loss of visual ability in her left eye for 3 years as shown in Figure 1. The patient also complained of difficulty breathing through her nose with frequent bleeding from her nostrils. The patient has had two previous craniotomies in the last six years,
with the most recent procedure involving the exenteration of her right eye due to panophthalmitis and proptosis caused by the retroorbital tumor. Her previous histopathology result showed Meningiotheliomatous meningioma. The patient was fully conscious and aware of presenting with proptosis of the orbits and no visual acuity in her remaining eye. A brain magnetic resonance imaging (MRI) revealed a 15 x 20 cm mass with homogenous enhancement on contrast admission at the frontal base area extending into the nasal cavity and all paranasal sinuses as well as the cavernous sinus as shown in Figure 2.

The patient was scheduled for a multistage operation as pre-op embolization could not be done due to unavailability at our institution during this period. After induction of anesthesia, the transcranial stage started with a bicoronal scalp incision, followed by a large bifrontal osteoplastic craniotomy to approach the intracranial part of the lesion. The resection of the orbital and ethmoidal roofs, both frontal sinus, and ethmoidal sinus resection was performed. Bleeding from the bony edges was controlled with bone wax. Bleeding was extensive throughout the operation with approximately 2 liters of blood loss. It was difficult to determine the borders of the tumor during surgery, and the lesion had produced extensive local destruction.

Our team scheduled the second stage of operation one week after using the craniofacial approach as seen in Figure 3 through a Weber-Ferguson incision with extension to the eyelids and elevated cheek flap. Resection of the maxilla including the dentoalveolar segment and exenteration of the globe was accomplished. Closure of the defect between the anterior cranial fossa and the facial structures was performed by utilization of a galeal frontalis myofascial flap. The frontal base was reconstructed using titanium mesh, fascia lata, and fat to reduce the risk of cerebrospinal fluid leakage. The estimated blood loss was another 2 liters, but the patient’s condition was stable throughout both operations.

Figure 1. The patient clinical presentation shows both proptosis of orbit

Figure 2. Magnetic resonance Imaging (MRI) scans axial, sagittal, and coronal sections of T1-weighted image with gadolinium contrast (T1-Gd). MRI scans show a large enhancing homogenous mass at the frontal base area extending to the nasal cavity.
There was a small residual tumor that was not taken out which was adherent to the right internal carotid artery (ICA), anterior cerebral artery (ACA) branches, and the cavernous sinuses. The patient was further treated in ICU and went home after 10 days without any new neurological deficit. Microscopically, the tissue section showed a hypercellular tumor with meningotheelial differentiation arranged in whorls and sheet-like growth. Tumor cells had oval to spindle-shaped nuclei that were highly pleomorphic. Mitotic figures were abundant (more than 20/10 HPF). There were areas of necrosis and hemorrhage. Based on the histologic features, the tumor was categorized as WHO grade III, Anaplastic meningioma (Figure 4).

During our one-month post-op MRI scan, Figure 5 shows a residual tumor on the right side with some significant edema still present on both frontal lobes. The nasal cavity and paranasal sinus did not show any residual tumor. The patient underwent radiotherapy 30 times with 76 Gy.

The postoperative period was uneventful, but cerebrospinal fluid (CSF) leakage was present 8 months after radiation therapy and the patient declined any further workup or additional intervention. The patient died one year after her last operation at our institution.

Figure 4. Microscopic features of the tumors. (A) Highly cellular tumor arranged mainly arranged in sheets (H&E stain, magnification 100x); (B) High power view showed tumor cells with marked nuclear pleomorphism and atypia. Note the brisk mitotic activity (H&E stain, magnification 400x).

Figure 5. Postoperative Magnetic resonance Imaging (MRI) scans of the patient. Axial, Sagital, and coronal section of T1-weighted image with gadolinium contrast (T1-Gd).
DISCUSSION

The majority of meningiomas are benign. However, Grade 3 meningiomas represent 1–3% of all meningiomas which are rare but bear a high recurrence rate and unfavorable prognosis [11,15].

The location of these tumors can be in any part of the brain. Frequent clinical symptoms must be considered given the cerebral mass effect and elevated intracranial pressure, such as headaches, nausea, and vomiting [1,6,16]. Meningiomas are usually located in the skull vault and the skull base; to be specific, the parasagittal area is the most frequent, followed by the falx, the cavernous sinus, the tuberculum sellae, the lamina cribrosa, the foramen magnum, and the torcular zones [17,18]. Parasagittal meningiomas makeup 17% to 20% of all the subtypes and most often involve the frontal lobe [10]. They can be asymptomatic for a long time and grow to a considerable size. The patient in our case had a large frontal base tumor extending inferior and lateral on the frontal base causing difficulty in breathing and proptosis of both orbits.

Various imaging techniques are important for the diagnosis of meningiomas. MRI is the standard modality for the radiologic diagnosis and surveillance of meningioma. Contrast-enhanced computed tomography (CT) may be used in patients who cannot undergo MRI. MRI typically reveals a dural-based, homogeneously enhancing, and well-circumscribed lesion. Benign meningioma will characteristically have a thickened, contrast-enhancing dural tail, and tumors are isointense to gray matter on non-contrast sequences. Meningiomas are extra-axial lesions, and the presence of a CSF cleft adjacent to the tumor can sometimes be seen. Digital Subtraction Angiography (DSA) is another helpful tool that can be used to not only exclude the possibility of an aneurysm or other cerebrovascular diseases but also to embolize the feeding vascular structures of the tumor [1,7,21]. Heavy blood loss was unavoidable due to the unavailability of DSA at our center during patient admission; fortunately, the patient was stable throughout the operations.

Pathologic diagnosis is obligatory to establish the histologic grading of meningiomas that has prognostic and therapeutic implications. The pathognomonic histologic feature of a meningioma is spherical formations of meningothelial cells, called whorls, which eventually mineralize into psammoma bodies. Grade III anaplastic meningiomas often resemble high-grade sarcomas and will show most or all the features of grade II atypical tumors, but the only required finding is more than 20 mitoses per ten consecutive high-power fields. This means that meningiomas with 4–19 mitoses, a very broad range, all still fall within the grade II spectrum. Rhabdoid and papillary morphologic variants are automatically classified as grade III tumors. However, even the lower grade meningiomas may demonstrate a histological progression to malignancy, the mechanism underlying malignant transformation remains unclear [16].

Tumorigenesis must be the result of exogenous or endogenous factors acting alone or together. Exogenous factors include trauma, viral infection, and prior brain irradiation. Endogenous stimulation can occur through the action of hormones or growth factors. The procedure of malignant transformation may take 2 to 16 years according to previous research [2,13,20]. For our patient, it took 6 years to accomplish malignant transformation after her first operation.

For tumors that are growing or causing symptoms, maximal safe surgical resection remains the standard of care for the therapeutic management of meningioma. However, the ability to achieve complete resection may be limited by several factors, including tumor location: involvement of nearby dural venous sinuses, arteries, cranial nerves, and brain invasion into eloquent tissue [21]. Tumors at the base of the skull (sphenoid wing, olfactory groove, tuberculum sella, cerebellopontine angle, or petroclival region) require more advanced surgical techniques and approaches to access the tumor without extensive brain retraction and injury safely. Some authors recommend a two-stage operation for intraosseous skull base meningiomas that have spread across the midline to involve the paranasal sinuses or facial structures. A combined transcranial and transfacial approach is needed for adequate exposure of the intracranial and extracranial portions of the tumor to provide the best hope of a surgical cure [17,22]. A large incision needs to be made for this kind of approach which would be the downside that needs to be faced with the risk of a higher volume of blood loss to achieve maximal tumor resection as what happens in our case. We highly recommend pre-embolization by using DSA to minimize the feeding artery if possible.

Radiotherapy was suggested to be adjuvant treatment after surgery on the first recurrent meningioma as it is reported to give better overall survival and local recurrence, although it does not affect 5-year progression-free survival. Patients with the first recurrence who undergo surgery alone would twice be likely to have a second recurrence compared with surgery complemented with radiotherapy [23].

The estimated 10-year overall survival for meningioma is 57.1 and 77.7% for patients at a younger age at diagnosis (20–44 years). The natural history of grade II and grade III tumors is much more aggressive, with rates of recurrence at 5 years approximating 50% for grade II tumors and 90% for grade III tumors. These recurrences translate into meningioma-specific mortality in these patients, with 10-year overall survival rates of 53% for grade II patients and 0% for grade III patients,
despite aggressive therapeutic efforts [11]. In our case, the patient survived another year since her last operation and almost 7 years since her first diagnosis and treatment.

CONCLUSIONS

Anaplastic meningioma portends a poor prognosis. An attempt at gross total resection should be implored in every case where feasible. Modifications in craniofacial resection techniques are based on the extent and nature of the underlying pathology of these types of extensive skull base meningiomas.

DECLARATIONS

Competing of Interest
The author(s) declare no competing interest in this study

Ethics approval and consent to participate
NA

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REFERENCES
