CASE REPORT

Transcranial Supraorbital Approach for Tumor Removal of Spheno-Orbital Meningioma with Favorable Clinical Outcomes

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ABSTRACT

Background: To describe and evaluate the surgery results regarding resectability and clinical outcomes of transcranial supraorbital approach for tumor removal of spheno-orbital meningioma
Case Illustration: A 58-year-old woman presented with proptosis of the right eye since 9 years prior. At initial examination, visual acuity was 6/15 with significant nasal visual field defect. There were prominent proptosis and inferior globe displacement of the right eye with no palpable mass. A fronto-parietal bone deformity was also observed. CT scan examination revealed hyperostosis of sphenoid, frontal, and temporal bone with extensive intraorbital mass with contrast enhancement suggestive of meningioma. Incisional biopsy was the performed and confirmed the histopathological diagnosis of meningothelial meningioma (WHO grade 1). Transcranial supraorbital in conjunction with lateral orbitotomy was performed in this patient as the definitive treatment for tumor removal of spheno-orbital meningioma. Craniotomy and opening of orbital roof were carried out by neurosurgeon to expose intraorbital region. Total tumor removal was than completed.
Conclusion: Transcranial supraorbital approach is an effective surgery for spheno-orbital meningioma removal as it offers excellent exposure. A radical resection through transcranial approach can be achieved with low morbidity, providing a significantly improved clinical outcome in long term period. In this case, the spheno-orbital meningioma was grossly resected totally with excellent visual outcome and acceptable cosmetic appearance.

Keywords: spheno-orbital meningioma, transcranial approach, tumor resection

Spheno-orbital meningiomas are complex tumors involving the sphenoid wing, orbit, and cavernous sinus. This makes their complete resection difficult or impossible.[1,2] However, meningioma resection has long been and is still being considered the primary and definitive treatment for meningioma.

The appropriate approach for removal of the tumor is determined by its location within the orbit. In general, anterior, medial, and lateral orbitotomies are ideal for tumors of the anterior orbit, lacrimal gland fossa, and anterior intraconal space. Because of

the narrowing of the posterior orbit and many structures that crowd the orbital apex, a transorbital approach to apical lesions may be risky and can result in vision loss.[3,4]

For tumors of the orbital apex and optic nerve tumors extending posterior to the optic nerve canal, a panoramic orbitotomy or transcranial orbitotomy has been the recommended approach. The safe removal of these lesions demands adequate surgical exposure. The transcranial approach to the orbit also has been recommended for processes that involve both the orbit and cranial cavity, such as optic nerve gliomas, orbital meningiomas, encephaloceles, and select mucoceles, hemangiomas, aneurismal bone cysts, and ossifying fibromas. Main objectives of the transcranial approach are the preservation of function and good cosmesis.[4]

In a study by Attia et al[5], it was shown that a combined cranioorbital approach appeared to be an effective one-stage surgery for resecting spheno-orbital meningioma. Transcranial approach may provide the best exposure and postoperative results.[2,4-6] The purpose of this case report is to describe and evaluate the operative results regarding resectability and fuctional outcome of transcranial supraorbital approach for spheno-orbital meningioma.

CASE ILLUSTRATION

A 58-year-old woman first came to Kirana Cipto Mangunkusumo Hospital with chief complaint of bulging of the right eye (RE) since 9 years prior. Initally, RE started to protrude slowly, but since 1 year ago, the proptosis progressed rapidly. At first, a small mass were noticed over the right upper eyelid which also enlarged slowly accompanied with minimal pain and redness. Blurry vision or double vision were denied. Long term use of oral contraceptives was also denied. Early in the course, Orbital CT scan was performed and revealed intraorbital mass. High dose oral steroid was given, but unfortunately the patient was lost to follow-up at that time.

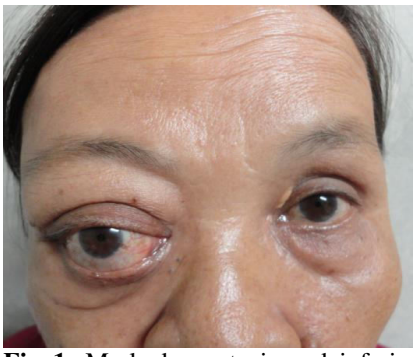

Fig 1. Marked proptosis and inferior globe displacement of the right eye at initial examination

Initial ophthalmological examination showed comparable best corrected visual acuity (VA), 6/6 and 6/7,5 of the RE and LE, respectively. Lefy eye was unremarkable. The movements of RE were limited to all directions. As shown in Fig 1, RE was proptotic with no palpable mass on the eyelid. There was also noticeable deformity over the fronto-parietal area. The anterior and posterior segment of the RE was unremarkable. Humphrey perimetry examination showed diminished nasal visual field, as seen in Fig 2.

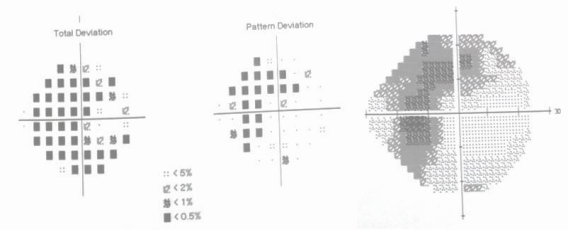

Fig 2. Humphrey examination showed marked defect over the nasal visual field

The patient was assessed with non-axial proptosis of the RE due to retrobulbar tumor with differential diagnoses of meningioma and fibrous dysplasia. Contrast and noncontrast CT scan were performed immediately and revealed hyperostosis of sphenoid, frontal, and temporal bone with extensive right intra-orbital mass suggestive of meningioma, as shown in Fig 3. Two months after initial visit, the patient underwent incisional biopsy.

Histopathological examination revealed a mixed type meningioma consisting meningio-theliomatous, fibrous, and transitional type (WHO grade 1). Craniotomy and lateral orbito-tomy followed by tumor removal were performed jointly with Neurosurgery Department. The

surgery took place 8 months after initial visit due to extensive queue of the surgery.

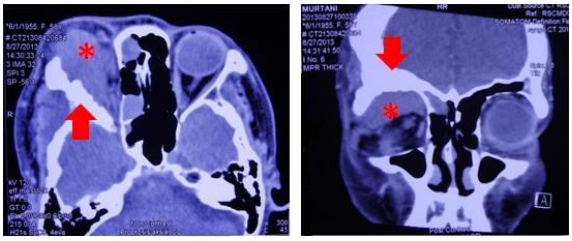

Fig 3. Orbital CT scan showing sphenoid hyperostosis (arrow) and massive intraorbital mass (asterix)

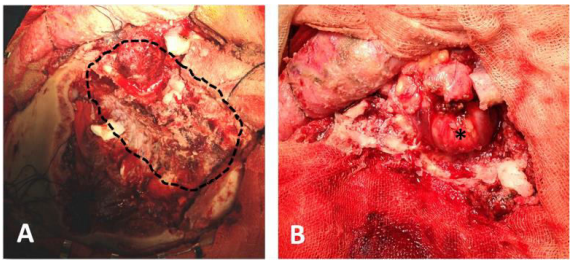

Fig 4. A) Craniotomy and opening of orbital roof was first carried out, followed by tumor removal from surrounding normal tissue (dashed line); and then **B)** intraorbital tumor removal was also performed around the globe (asterix)

The summary of the surgery was shown in Fig 4. The surgery was first performed by neurosurgeon through right semicoronal incision along temporal line towards right orbital rim and extended left semicoronal incision 3 cm contralateral from the midline. The border of healthy bone tissue was identified from the tumor-infiltrated bone tissue. Craniotomy was performed over the healthy bone tissue on the right frontal area. Duramater stripping was then performed and the infiltrated bone tissue was removed. Next step was craniotomy of orbital rim and roof to expose retrobulbar area. There was grayish-red mass within the retrobulbar space filling the superior, medial, and lateral orbit, and the surgery was continued by orbital surgeon. The mass was separated from surrounding healthy and tissue, and then removed as clean as possible. Neurosurgeon took over the surgery once again to put back the orbital rim, performed duraplasty and closed to wound with flap.

Histopathological examination showed concurrent result with initial biopsy which showed mixed-type meningioma predominantly meningiotheliomatous, transitional, angiomatous, fibrous, metaplasia, and psamomma (WHO grade 1). Two weeks after surgery, uncorrected VA of RE was 6/12. There were inability of movement of the right eye and ptosis. The anterior and posterior segments were unremarkable.

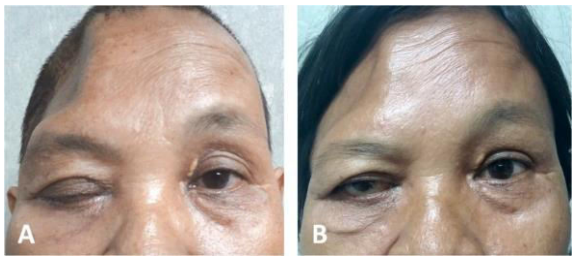

Fig 5. A) One month and **B)** 11 months after surgery with apparent fronto-parietal deformity and improvement of proptosis and globe displacement.

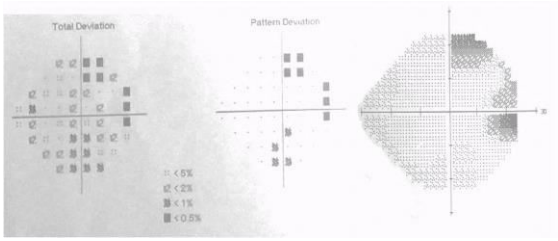

Fig 6. Humphrey examination 11 months after the surgery showed improvement of previous nasal visual field defect before tumor removal.

Eleven months after surgery, best corrected VA of the RE was 6/6. From ophthalmological examination, the condition was similar to the last follow up, as shown in Fig 5 comparing the condition 1 month and 11 months after the surgery. Inability of movement and ptosis of the RE was still apparent. Humphrey examination showed marked improvement of the nasal visual field defect compared to the perimetry prior to tumor removal surgery, as seen in Fig 6. Postsurgical reconstruction was further planned by neurosurgeon.

DISCUSSION

Spheno-orbital meningiomas represent up to 9% of all intracranial meningiomas.[7,8] Spheno-orbital meningiomas are secondary tumors of the orbit that originate from the dura of the sphenoid wing bone. They exhibit

interosseous tumor growth, leading to hyperostosis and a thin, carpet-like soft tissue growth at the dura.[2,9] It is mainly characterized by painless progressive proptosis and unilateral visual loss.[8]

Our patient was 58 years old female who first came with complaint of slow growing proptosis of the right eye since 9 years prior. The proptosis was not associate with pain, blurry vision, or double vision. Spheno-orbital meningioma occurred exclusively in females (94%). These tumors often become symptomatic because of their proximity to the orbit and visual system. Proptosis is the most common clinical sign of spheno-orbital meningioma. In a significant number of patients, proptosis may be the only clinical sign, as seen in our patient. Hyperostosis of the orbital walls and venous statis due to compression of the ophthalmic vein can all lead to proptosis.[9]

Radiological imaging is needed to deliminate the extension of the tumor within the orbit and the intracranial region. Radiological findings in orbital meningiomas may include hyperostosis, thickening of the optic nerve or the 'tram track sign' in optic nerve sheath meningioma, and calcification, which can be seen in CT scanning.[8] In our case, orbital CT scan showed hyperostosis of sphenoid, frontal, and temporal bone with extensive right intraorbital mass with contrast enhancement involving lateral and superior rectus muscle suggestive of meningioma.

The causes of hyperostosis in meningiomas are theorised to include vascular disturbances of the bone caused by the tumor, irritation of the bone without actual invasion, previous trauma, the production of bone by the tumor itself, and the stimulation of osteoblasts in normal bone by factors secreted by tumor cells. The most accepted cause of hyperostosis associated with spheno-orbital meningioma is direct tumor invasion, which could explain the disproportionate amount of hyperostosis relative to the dural size of the tumor.[9,10] This is why a complete resection of the hyperostotic bone must be done when possible, as most of the recurrences seem to be due to residual tumor.[8]

Among several possible disease entity as a differential diagnosis, fibrous dysplasia is one of the most common misdiagnosis preoperatively in cases of meningioma.[8] However, from orbital CT scan, it was more suggestive of meningioma. Incision biopsy is sometimes required to rule out other possible diagnosis in cases of suspected meningioma. Our patient underwent incision biopsy which showed a WHO grade I meningioma or a benign meningioma. Approximately, 78-81% of meningiomas are benign (non-cancerous). Less common meningiomas are grade II (atypical) and grade III (malignant or anaplastic meningioma) which count for 15-20% and 1-4% of all cases of meningioma, respectively.[11]

Meningioma resection has long been considered the primary and definitive treatment for meningioma. The location of the tumor remains the primary factor in the determination of the surgical approach for tumor excision. Other criteria include the size of the tumor, probable histopathology, signs of vision impairment, tumor invasion of surrounding tissues, and involvement of structures contiguous to the orbit. If the tumor extends into the superior orbital apex, medial to the optic nerve, or into the intracranial cavity, a frontal craniotomy may provide the best exposure and postoperative results. The craniotomy may or may not require removal of the actual superior orbiyal rim along with the roof of the orbit.[12] Different surgical approaches, such as pterional, fronto-temporal, transzygomatic, and transcranial-transmalar for the resection of spheno-orbital meningioma have been described.[13,14]

One of the techniques is removing the superior orbital rim and part of the anterior orbital roof throug a biocoronal skin incision. This allows excellent access to the orbit while minimizing brain retraction.[4]

In our case, the transcranial supraorbital and lateral orbitotomy approach was the chosen technique. Craniotomy of the

orbital rim and roof was performed to maximize exposure to the retrobulbar area, and gross total tumor resection was done. In recent years, it has been shown that most cranio-orbital tumors can be extirpated completely by transcranial surgery. However, according to the several studies, primary complication is impaired vision and drawback of eye movement.[15,16] This was also seen in our patient who developed inability of eye movement and ptosis after the surgery.

The visual prognosis of spheno-orbital meningioma patients was proportional to the degree of extension and chronicity of meningeal compression.[14,17] Visual field defects should not be ignored, and the presence of an intraorbital soft-tissue component was shown to be a risk factor for severe final visual field sequelae. Based on these observations, patients with spheno-orbital meningioma display a better visual prognosis if surgical intervention is carried out early before the appearance of adverse clinical signs while being performed by a surgical team with expertise in this type of surgery.[2] Visual prognosis of our patient was good reaching 6/6. There was also improvement of nasal visual field defect seen in Humphrey examination. This might be due to relatively early surgical management before further adverse clinical signs occur.

Some study considered spheno-orbital reconstruction after tumor resection unnecessary.[17] However, some other studies suggested reconstruction for structural and cosmetic reasons and also to provide landmarks for post-operative imaging.[2] Furthermore, if spheno-orbital reconstruction is not performed after lesion resection, several potential problems might occur such as meningocele formation, diplopia from extra-ocular muscle fibrosis, orbital pain, pulsating enophthalmos, and restrictive ptosis.[2,7,18] Our patient was initially planned to undergo reconstruction a few months after initial surgery, but unfortunately she was lost to follow up several months. On her last visit, we suggested the patient to have a regular follow up at Neurosurgery Department to decide whether sphenoid reconstruction is required or not.

CONCLUSION

Spheno-orbital meningiomas can be difficult to manage. Surgical resection can reduce the degree of proptosis and stabilise visual function. Transcranial supraorbital approach is an effective surgery for spheno-orbital meningioma removal as it offers excellent exposure. A radical resection though transcranial approach can be achieved with low morbidity, providing a significantly improved clinical outcome in long term period. In this case, the spheno-orbital meningioma was grossly resected totally with excellent visual outcome and acceptable cosmetic appearance.

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