Original Article

Outcome of Sphenoid Wing En Plaque Meningioma Surgery

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ABSTRACT

Background: Sphenoid wing en plaque meningiomas are complex tumours involving the sphenoid wing, the orbit, and sometimes the cavernous sinus. Complete removal is difficult, so these tumours have high rates of recurrence and postoperative morbidity. **Objective:** In this study the extent of tumour resection and postoperative complications in patients with meningioma en plaque were investigated. **Patients and Methods:** A retrospective study was made on clinical manifestations, neuroradiological features, and operative techniques in nine patients undergoing transcranio-orbital approach for sphenoid wing en plaque meningioma resection from January 2011 through December 2014. Patients ages ranged from 16 years to 67 years; 45.5 years in average. There were two males and seven females. The main complaints were progressive proptosis and visual acuity deficits. All patients were operated on using a fronto-temporal approach with orbital decompression. **Results:** Simpson grade II resection was achieved in three patients, Simpson grade III in six patients. Pathological examination showed that seven (77.8%) patients had meningothelial meningiomas. After surgery, proptosis improved in six patients (66.7%) while visual acuity improved in three patients (42.9%). Temporary ophthalmoplegia was found in three patients and cerebrospinal fluid leak was found in one patient. Duration of follow up was from 3 months to 3 years. Three patients showed progressive growth of the remaining tumor. There were no mortalities in this study. **Conclusions**: Sphenoid wing en plaque meningiomas are more likely to produce adjacent hyperostosis. All the hyperostotic bone of the great wing of sphenoid bone should be removed to minimise incidence of regrowth or recurrence. Extensive tumor removal with bony decompression at the orbital apex can produce satisfactory cosmetic and functional outcome.

INTRODUCTION

Sphenoid wing meningiomas with osseous involvement represent 9% of all intracranial meningiomas[1]. They may be considered a variety of en plaque meningiomas according to 1938 definition of Cushing and Eisenhardt[2]. Meningioma en plaque is characterized by a thin, sheet-like lesion that infiltrates the dura and invades the bone leading to significant hyperostosis. The sphenoid wing is the most common location of meningioma en plaque. Sphenoid wing en plaque meningiomas are also called spheno-orbital meningiomas and pterioneal meningiomas en plaque[3,4].

Differential diagnosis includes fibrous dysplasia, osteoma, and metastasis. Because of the rarity of this tumor, only a few published studies about surgical treatment of sphenoid wing meningioma en plaque are available in the literature [1,4-10]. Complete surgical resection is difficult because of the involvement of the orbital apex, superior orbital fissure and cavernous sinus, so it has a higher recurrence rates.

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PATIENTS AND METHODS

Between January 2011 and December 2014, nine patients underwent resection for sphenoid wing en plaque meningioma in Kasr Al-Ainy Hospitals, Cairo University.

All patients had proptosis. Visual acuity measurements of the affected eyes revealed, seven patients were less than normal: two patients were 0.5-1.0, three patients were 0.1-0.5, one patient was less than 0.1, and one patient had no light perception. Opthalmoplegia were present in two patients, mainly upside and abduction movement disorder. Perimetry was done in all patients to assess visual field defects.

All patients had brain and orbit computed tomography (CT) and magnetic resonance imaging (MRI) scans. Axial and coronal CT bone window scans were excellent for visualization of the hyperostosis. On CT scans, the typical features were significant hyperostosis of the great wing of sphenoid and this usually extended to involve the adjacent bone. This included the bones of the middle cranial fossa, the lateral orbital wall, the orbital roof and the walls of the sphenoid sinus. The edges of hyperostotic bone were rough and brush-like. On CT scans, the soft-tissue component of meningioma was carpet-like, isodense and homogeneously enhanced after contrast.

On T1 intensity MRI, the lesion was isointense or hypointense, while on T2 intensity, the lesion showed slight hyperintense signal. It underwent marked Gadolinium enhancement which is a typical features of meningioma. The hyperostotic bone showed different radiological picture and did not enhance **(Fig. 1)**. In those patients with tumor extending into the orbit, postcontrast fat suppression T1-weighted MRI was useful to evaluate the extension of dural enhancement and soft-tissue involvement of the orbit.

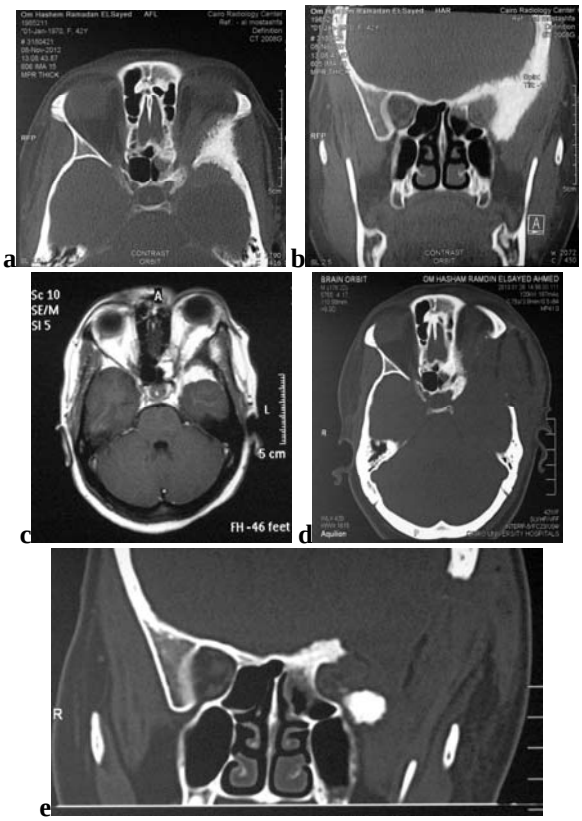

Fig. 1 a-e: A female patient with left sphenoid wing en plaque meningioma. a&b: Axial and coronal CT scans show typical hyperostosis of the greater wing of sphenoid and adjacent temporal bone, the edge of hyperostosis is rough and brush-like. c:T1-weighted MRI scan with gadolinium shows enhancement of the dura in the left anterior temporal area. The area of hyperostosis of the sphenoid bone including the lateral orbital wall is hypointense and did not change after gadolinium injection. d&e: Postoperative axial and coronal CT show the extent of bony resection.

All patients were operated upon using the pterional approach. The patients were put in the supine position, the head and the upper body was slightly elevated to reduce the cranial venous pressure. The head was tilted 30 degrees to the contralateral side and the neck was extended so that the cortex felt away from the sphenoid ridge. The scalp incision extended from the zygoma to 1-cm off the frontal midline following a curve behind the hairline. Care was taken during this part of the incision to avoid injury to the frontal branch of the facial nerve. In some patients, the extracranial soft tissue was involved. In those patient the extracranial soft-tissue tumor of the infratemporal fossa or the temporal muscle was resected first. After removal of the extracranial part of the tumor, a high-speed drill was used to remove any hyperostotic bone of the lesser and/or greater sphenoid wings. Removal of the lesser sphenoid wing opened the optic canal and upper part of the superior orbital fissure, removal of the greater sphenoid wing opened the lateral and lower parts of the superior orbital fissure, and removal of the lateral wall and roof of the orbit exposed the periorbita. The greater sphenoid wing was removed with a high-speed drill between the periorbita and temporal dura. It was drilled down to the opening of the foramen rotundum. The infratemporal fossa was then exposed. If there is a marked hyperostosis around the optic canal, the optic canal should be opened and decompressed. This was needed in three patients.

The blood supply of this tumor was usually from branches of the middle meningeal artery. During the greater sphenoid wing resection, those branches were coagulated and cut. It was suggested to resect the infiltrated dura 10 mm around the main tumor except in the superior orbital fissure and cavernous sinus where the residual tumor was coagulated. Intraorbital tumor nodules were resected under meticulous avoidance of intraorbital nerves. Areas of the periorbita involved by the tumor were also removed. The extent of resection was evaluated according to Simpson grading scale[11] **(Table 1)**.

Table (1): Simpson grading system for removal of meningiomas

Grade	***Degree of removal***
I	Macroscopically complete removal with excision of dural attachment and abnormal bone.
II	Macroscopically complete with endothermy coagulation of dural attachment.
III	Macroscopically complete without resection or coagulation of dural attachment or of its extradural extensions e.g. hyperostotic bone.
IV	Partial removal leaving tumour insitu.
V	Simple decompression (biopsy)

After tumor removal, the dura and periorbita were reconstructed with fascial grafts or artificial dura. In two patients, a muscle graft was required at the middle cranial base, augmented with histoacryl, to achieve a watertight closure and prevent cerebrospinal fluid leak. The superior and lateral orbital rims were important for a good cosmetic appearance, so it should be preserved or reconstructed. After bony resection of the orbital roof and lateral orbital wall, the eye bulb can remain in place if the superior and lateral orbital rims remain intact, so further bony reconstructions of orbital walls were not performed. The sphenoid ridge did not need reconstruction, but the temporal bone defect could be replaced with titanium mesh or bone cement.

RESULTS

Nine patients were treated surgically using transcranio-orbital approach to resect sphenoid wing en plaque meningiomas in Neurosurgery Department, Kasr Al-Ainy Hospitals, Cairo University between January 2011 to December 2014. Seven were females and two were males with a mean age of 45.5 years (ranged: 16-67 years). Average medical history was 31 months (range: 6 months to 8 years). Five lesions were found on the right side and four were on the left side. One patient had neurofibromatosis type II.

The most common presenting symptom was proptosis (nine patients). Other symptoms and signs included deficits of visual acuity (seven patients, 77.8%), headache (four patients) and seizures (one patient) **(Table 2)**.

Table 2: Presenting symptoms of the patients

Presenting symptom	***No. of patients***	***Percentage***
Proptosis	9	100%
Declined visual acuity	7	77.8%
Visual field deficits	4	44.4%
Headache	4	44.4%
Temporal swelling	3	33.3%
Diplopia	2	22.2%
Seizures	1	11.1%
Blindness	1	11.1%

Three patients were graded II while six were graded III according to Simpson grading system for tumour removal. Total tumor resection was not achieved in any of our cases. Complete surgical resection was difficult because of the involvement of the orbital apex, superior orbital fissure and cavernous sinus. Six tumor residuals in the cavernous sinus and superior orbital fissure were treated by gamma knife therapy.

Histopathological examinations showed benign meningioma (WHO grade I) in all of our patients. Seven tumors (77.8%) were meningothelial meningiomas and two were fibroblastic meningiomas.

After surgery, proptosis improved in six patients (66.7%). Out of seven patients presenting with deterioration of their visual acuity, three patients (42.9%) improved, the other three patients remained unchanged and one patient worsened postoperatively. Three patients (33.3%) showed temporary ophthalmoplegia and two patients had abducent deficits. Those symptoms improved in 2–3 months. Cerebrospinal fluid leak was found in one patient and stopped after using continuous lumbar drainage for 1 week. There were no cases of intracranial hematomas, meningitis, or pulsating exophtalmos.

Patients were followed-up with an average follow-up time of 16 months (ranged: 3 months – 3 years). Three patients (33.3%) showed progressive growth of the remaining tumor. In two patients, the tumour volume was small and the rate of growth was slow, so close and regular neurological and ophthalmological follow-up was advised. The third patient with regrowth received gamma knife treatment. There were no operative deaths in this study.

DISCUSSION

Sphenoid wing en plaque meningiomas are a clinical and pathological subgroups of meningiomas defined by its particular sheet-like dural involvement and its disproportionately large bone hyperosteosis. They are presumed to account for 2%–4% of intracranial meningiomas[7]. They are located in a complex anatomical area between intracranial and orbital compartments. They usually involve the sphenoid wing and the orbit, so they are also known as spheno-orbital meningiomas[8].

The cause of associated hyperostosis in meningiomas at the sphenoid bone remains a point of controversy: specifically regarding whether this represents a secondary change of the bone without tumor invasion versus direct infiltration of the bone by a tumor[9,12]. Most authors agree with Cushing's conclusion that the infiltration of meningioma cells into bone stimulates osteoblastic activity with hyperostosis. New bone growth probably resulted from periosteal stimulation by tumor invasion. Histopathological examination of hyperostotic bone revealed tumor cells invading the Haversian system of the overlying bone, so hyperostosis is a part of the tumor. In all patients in this study, hyperostotic bone samples were sent for histopathological examination, and all of them were confirmed to have tumor invasion.

In this study, nine patients were operated for sphenoid en plaque meningiomas. Proptosis improved in six patients (66.7%). Visual acuity improved in three patients (42.9%) out of seven patients presented with impaired visual acuity. Ringel et al.[8] reported 63 patients of spheno-orbital meningiomas treated

surgically of which proptosis improved in 77% of patients and 64% of patients presented with declined visual acuity had improved.

MRI is the best modality to assess the extent of dural and intradural involvement while CT scan allows better visualization of bone involvement. Postcontrast fat suppression T1-weighted MRI is useful to evaluate the presence and degree of meningeal enhancement especially in those patients with tumours extending into the orbit[7]. Some CT features of hyperptosis are suggestive of en plaque meningiomas including periosteal pattern of hyperptosis, inward bulging of the vault lesion, surface irregularity of the hyperptotic bone and subtle intracranial changes[13].

The surgery of sphenoid wing meningioma en plaque poses a great technical challenge because of the complicated anatomical boundaries of the orbits, the cavernous sinus, and the cranial nerves and vascular structures in the medial aspect of the sphenoid wing[1,5]. Different surgical approaches have been proposed for the removal of sphenoid wing meningioma en plaque, including pterional, frontotempo-orbito-zygomatic, cranio-orbital, and combined transcranial-transmalar approaches[1,4,5,7-10]. All patients in this study underwent fronto-temporal craniotomy. This approach provides a sufficient access to the orbit apex and the base of the middle cranial fossa to resect the bone and soft tissue and decompress the superior orbital fissure and the optic canal. In some authors' opinion, removal of the zygomatic arch is not necessary, because additional space provided by removal of the hyperostotic bone can increases tumor exposure[3]. The greater wing of sphenoid bone is removed with a high-speed drill between the periorbita and temporal dura. The foramen rotundum is opened carefully. The foramen rotundum is an important anatomic landmark as the cavernous sinus is located medial to it[8].

Recurrence rates for benign meningiomas are inversely related to extent of surgical removal of the tumour[14]. To prevent the tumor recurrence, all the tumor-infiltrated bone in the cranial base should be resected, especially the hyperostotic bone of anterior clinoid process[5]. If the hyperostosis around the optic canal is obvious, the optic canal should be opened and decompressed. It is recommended to open the dura to identify the optic nerve intradurally before unroofing the optic canal and incising the dural cuff of the optic nerve for a short distance[9]. The blood supply of the tumor is from branches of the middle meningeal artery, during the greater sphenoid wing resection, those branches are coagulated.

After completion of the bony removal, and removal of all the intracranial tumor, the dura is opened beyond the area of carpet-like tumor infiltration, and all infiltrated dura should be resected. This includes the dura over the sphenoid wing, and the temporal and frontal bone. Jesus et al. found that the hypervascular dura contained neoplastic cells more than 10 mm away from the main tumor, so he recommended that an extensive amount of dura be removed with the main tumor and the margins should be pathologically studied. An intraorbital tumor nodule displaces rather than infiltrates the orbital context, so areas of the periorbita involved by the tumor are also removed[7].

The best surgical outcome of sphenoid wing meningiomas en plaque is dependent on early diagnosis and complete removal[5,8,10,14]. Radical resection attempts carry a high risk of postoperative neurological morbidity especially in cases with orbital and cavernous sinus extension. This risk has to be considered when planning the surgical treatment, especially because meningioma en plaque is a benign tumour and there are other non invasive treatment options to deal with the residual tumour like radiation therapy. In these cases, the surgical aim should be the relief of leading symptoms rather than radical resection[8,15,16].

Recurrence rate of sphenoid wing meningiomas en plaque was reported to be about 35%–50% in the literature[10,17]. In this study, re-growth occurred in three patients (33.3%) in a mean follow up of 16 months. The growth of the residual was slow in two patients and rapid in a patient to justify radiosurgery. However, greater sample size and longer follow-up period are needed. Ringel et al.[8] reported 63 patients of spheno-orbital meningioma, 76% of which had tumor residuals. Follow-up data was collected for up to 17 years (median, 4.5 years), of which 61% were stable and didn't need further treatment while 39% were progressive. Maroon et al. reviewed recurrent spheno-orbital meningiomas and developed the following hypothesis to explain the high rate of recurrence: (1) failure to diagnose early because of symptoms similar to those of other clinical entities such as fibrous dysplasia, (2) inadequate resection secondary to the involvement of neurovascular structures, (3) surgeons' cautious behaviour to avoid iatrogenic morbidity and mortality[1]. In patients with tumour residuals, radiosurgery is effective in controlling tumor growth. Kondziolka et al. documented an overall control rate of 93% for patients with benign meningiomas managed with stereotactic radiosurgery with an overall morbidity rate of 7.7%[18].

Reconstruction has been a matter of controversy. Many authors propagate a firm reconstruction of the orbital walls to avoid enophthalmus, pulsating eye bulb, or oculomotor muscle fibrosis, which may result in ophthalmoplegia[8,10,19-21]. The superior and lateral orbital rim is important for a good cosmetic appearance, so it should be preserved or reconstructed[20]. The reconstruction of sphenoid wing bone is not needed, but the temporal bone defect can be reconstructed with titanium mesh or bone cement. After bony resection of the orbital roof and lateral orbital wall, the eye bulb remains in place if the superior and lateral orbital rims remain intact. Maroon et al. reported 200 patients of

orbital wall and roof resections without reconstruction, resulting in no permanent pulsating enophthalmus[1]. DeMonte et al. concluded that a partial or complete orbital roof resection, isolated or combined with lateral or medial orbital wall defects, does not require routine reconstruction if the periorbita remains intact[22]. Talacchi et al. stated that one of the goal standards of surgical treatment is normalization of proptosis[23]. This can be achieved by accurate resection of the superior and lateral orbital walls and careful reconstruction of the frontobasal dura. He stated that this is far superior to bony reconstruction. No patients in this study underwent reconstruction of the orbital walls and no patients experienced enophthalmus or pulsating eye during the follow up period.

CONCLUSION

All the hyperostotic bone of the great wing of sphenoid bone should be removed to decrease recurrence of sphenoid wing en plaque meningiomas and to relief the symptoms. Total excision must be done in case of resectable tumours. In those cases with involvement of the cavernous sinus and the orbital apex, a subtotal but extensive tumour removal with bony decompression of the cranial nerves at the superior orbital fissure and optic canal produces good functional and cosmetic results. Radiosurgery can be used to control the regrowth of the residual tumor.

Conflict of interests:

The authors declare that they have no conflict of interests.

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