

Clinical Outcome of Surgical Excision of Spheno-Orbital Meningiomas: Local Experience

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Abstract

Background: Spheno-orbital meningiomas are considered as tumors involving the sphenoid wing, orbit, and cavernous sinus. Their complete surgical resection is difficult. Sphenoidal hyperostosis that results in incomplete resection makes these tumors prone to high rates of recurrence.

Aim of Study: The aim of this retrospective study was to investigate the outcome of twelve consecutive patients treated by microsurgical resection of spheno-orbital meningiomas.

Patients and Methods: This series consisted of twelve patients who were retrospectively analyzed over a 5-year period. Visual function was evaluated pre-and post-operatively in all patients. We used a standardized surgical approach through supraorbital-pterional approach with drilling of all sphenoidal hyperostotic bone. Surgical outcome, complications and recurrence were clarified.

Results: There were two men and ten women included in the study, with a mean age of 50.8 years (range 27-63 years). Pre-operative symptoms include proptosis (67%), blurred vision (58%) and diplopia. All patients underwent microsurgical resection through supraorbital-pterional approach. An excellent clinical result was observed in six patients. Recurrence was evident in four patients.

Conclusions: Spheno-orbital meningiomas are characterized by potentially extensive hyperostosis of the skull base. Successful resection necessitates drilling of the superior orbital fissure and orbital osteotomy within anatomical limitations. Complete resection was found to be limited by tumor remaining in the cavernous sinus and the optic canal.

Key Words: *Spheno-orbital meningiomas – Orbital tumor – Proptosis.*

Introduction

SPHENO-ORBITAL meningiomas are a complex category of intracranial tumors that present with hyperostosis of the sphenoid wing and direct orbital involvement. They have traditionally been defined as secondary tumors of the orbit, originating from the dura of the sphenoid wing bone; however, on

closer pathological examination there is a distinct periorbital component that is a defining feature of these tumors [1].

Sphenoid wing meningiomas may grow and spread in different directions, affecting such areas as the cavernous sinus, sella turcica and orbit [2]. A secondary involvement of the orbital cavity may occur through the bone, the optic canal, and/or the superior orbital fissure; although in most invasive cases the pathway of tumor spread cannot be defined. Intraorbital invasion is estimated to occur in 39% to 50% of sphenoid wing meningiomas [3-7]. Approximately 9% of all intracranial meningiomas have spheno-orbital localization [8,9].

Extensive orbital invasion with diffuse bone infiltration represents an important surgical problem. Because of their extensive dural, bone, and orbital involvement, adequate resection of spheno-orbital meningiomas has historically been difficult to achieve, leading to a high rate of recurrence, as high as 35 to 50% [10-13]. Some authors have reported that longstanding tumor-related proptosis cannot be surgically treated and therefore should not be attempted [10,13].

This study reviews twelve patients with spheno-orbital meningiomas operated on in the Neurosurgical Department of our institution. The aim is to discuss the surgical approach, the outcome, complications and recurrence of this type of tumors.

Patients and Methods

Between February 2010 and December 2015, we retrospectively followed a cohort of twelve patients with spheno-orbital meningiomas in Department of Neurosurgery at Suez Canal University Hospital. Information on the patients' clinical history and signs, surgical approach, pathology and outcomes were recorded.

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Patients with non-hyperostotic sphenoid wing meningiomas, cavernous sinus meningiomas with secondary orbital involvement, primary optic nerve sheath meningiomas, and clinoidal meningiomas were excluded from this study. None of these patients had undergone surgery or radiation therapy before their first presentation.

All patients underwent an evaluation of the neurological functions with detailed cranial nerve testing and the results of which were compared to the post-operative findings. Presenting symptoms and clinical signs were recorded. All patients underwent visual function assessment, including visual acuity and visual field tests.

Pre-operative neuroimaging investigations consisted of Computerized Tomography (CT) with tridimensional reconstruction and Magnetic Resonance Imaging (MRI) using a 1.5-tesla unit, with and without gadolinium contrast. High resolution CT scans provided the best images of hyperostosis of the sphenoid wing and neighboring structures. T1- and T2-weighted and contrast-enhanced MR imaging studies were performed to analyze the extent of intradural and intraorbital component. All patients had a tumor mass of variable size and location in the orbital cavity. Based on the tumor extent and location in the orbit, four types were identified:

- 1- Lateral and superolateral (seven cases).
- 2- Medial and infero-medial (two cases).
- 3- Orbital apex (two cases).
- 4- Diffuse (one case).

CT scans with bone window were also obtained 24h after surgery to identify immediately the presence of postoperative hematomas, pneumocephalus, or other surgical complications. Post-operative MRI was performed three and six months after surgery, and then annually, if necessary.

Surgical procedure:

Surgery is performed under general anesthesia, with the aid of an operating microscope and microsurgical instrumentation in all cases. Pre-operative antiepileptic loading is mandatory. All cases had been operated through the supraorbital-pterional approach. The patient was positioned supine and fixed with Mayfield and the patients head was rotated 30 degree to the contralateral side.

A large frontotemporal incision is made. Sub-fascial dissection is performed to elevate the scalp flap, protecting the frontal branch of the facial

nerve, and exposing the superior and lateral rims of the orbit and the zygomatic arch. While elevating the temporal muscle, tumor involvement in the muscle must be carefully noted and excision must be performed when involvement is seen. The area of hyperostosis is encountered early at this stage of dissection, or even earlier as there can be tumor infiltration of the temporal muscle as well. The entire area of thickened hyperostosis is then removed using a series of cutting burrs, which results in a craniectomy exposing the frontal and temporal dura and the periorbita. A zygomatic osteotomy was performed in two cases with a significant amount of orbital involvement. During this bone drilling, the superior orbital fissure should be carefully identified because it is often distorted and severely compressed. Further drilling around the superior orbital fissure is performed under, high magnification of the surgical microscope after completing the craniotomy, which provides access to the convexity dura that may be involved over a wide area and bone drilling is continued up to the inferior margin of the superior orbital fissure. The anterior clinoid process, which is often hyperostotic, is removed and the optic canal is unroofed in its entire course.

Involvement of the orbit is directly through the periorbita. Starting anteriorly, the periorbita is incised carefully, protecting the lacrimal gland and the lacrimal nerve, and resected in a piecemeal fashion) with careful dissection of the extra ocular muscles and the annulus of Zinn. Tumor is resected up to the superior orbital fissure.

Results

Clinical and radiological findings:

Twelve patients with lesions located at the sphenoid bone and orbit were identified and analyzed. The patients were ten females and two males, with a male/female ratio of 1:5. The patient age ranged from 27 to 63 years (average, 50.8 years). For patients' characteristics, see (Table 1).

The most frequent symptoms of clinical onset were proptosis (67%) and blurred vision (58%). Diplopia was present only in two cases (17%). Specific neurological symptoms, such as headache or seizures, were present in two cases (17%). At clinical examination, all but one patient (92%) had a variable degree of proptosis, and six (50%) had visual dysfunction; disturbances of eye motility were found in four cases (33%) and optic disc edema or pallor in four cases (33%). The neuro-radiological diagnosis was performed by MRI and

CT-scan in all cases. All patients had a tumor mass of variable size and location in the orbital cavity. Cases were classified according to the location of tumors into the orbit into four types (Table 1).

Histologically, all tumors were benign (WHO I) meningiomas, of the meningothelial (six cases), transitional (four cases), fibrous (two cases) type.

Extent of tumor resection:

An apparently complete tumor resection, based on intraoperative observation and postoperative CT or MRI studies, was thought to have been achieved in eight out of twelve patients (66.7%). This group includes both Simpson grades I (four cases or 33%) and II (four cases or 33%), because it was difficult in some cases to exactly define the entity of the dural resection. Simpson grade III removal was obtained in three patients (25%) and grade IV in one (8.3%).

The analysis of the extent of the tumor resection according to the type of intraorbital location has shown that a seemingly complete resection was achieved in all lateral or superolateral tumors. On the other hand, in diffuse orbital tumors, the removal was mainly incomplete. The most frequent location of residual tumor in incomplete resections was the superior orbital fissure. In all cases aggressive surgery was deliberately not performed to avoid further ophthalmological and neurological deficits.

Post-operative complications:

Deficits of the oculomotor nerve were the most frequent neurological troubles in six patients post-

operatively; this was due to drilling of the hyperostotic lesser sphenoid wing and anterior clinoid with remission within three months in four patients. One patient (8.3%) had transient paresis of the eye muscles. One patient (8.3%) had worsening of his visual acuity. A hyperpathic trigeminal sensation was noted in four patients. A subgaleal cerebrospinal collection was encountered in three patients. Permanent morbidities in our series consisted of temporal hollowing of moderate grade in three patients and a permanent third nerve injury in two patients.

Outcome:

The outcome was classified as excellent, good, moderate, and poor. An excellent clinical result, with disappearance of exophthalmos and without neurological sequelae, was observed in six patients (50%). Three patients (25%) had a good outcome, with an acceptable cosmetic result, reduction of the exophthalmos, and no neurological deficits. Three (25%) had moderate neurological sequelae.

Recurrence:

The overall rate of recurrence in the entire follow-up period was 33.4% (four patients). The recurrence rate according to the extent of the surgical resection (Table 1) was significantly lower for grades I and II (0%) than for grades III and IV (100%). The correlation between recurrence and initial tumor location showed a higher rate of recurrence for diffuse forms and for the orbital apex type, than for the lateral and superolateral forms.

Table (1): Patient characteristics and clinical findings in the study series.

Case No	Age/ Sex	Presentation	Tumor location in the orbit	Simpsons grade (tumor removal)	Complications	Outcome	Recurrence
1	32/F	Proptosis	Superolateral	Grade 1	• Transient 3 rd nerve palsy	Good	
2	47/M	Proptosis, eye pain	Lateral	Grade 2	–	Excellent	
3	53/F	Blurred vision	Orbital apex	Grade 3	• Transient 3 rd nerve palsy	Moderate	+
4	27/F	Headache, blurred vision, eye pain	Medial	Grade 3	–	Good	+
5	56/F	Proptosis, blurred vision, diplopia	Diffuse	Grade 4	• Permanent 3 rd nerve palsy, decreased visual acuity, ophthalmoplegia	Moderate	+
6	60/M	Proptosis, seizure	Superolateral	Grade 2	–	Excellent	
7	57/F	Blurred vision, diplopia	Orbital apex	Grade 3	• Permanent 3 rd nerve palsy	Moderate	+
8	45/F	Proptosis	Superolateral	Grade 1	–	Excellent	
9	63/F	Proptosis, blurred vision	Superolateral	Grade 2	• Transient 3 rd nerve palsy	Excellent	
10	44/F	Proptosis	Superolateral	Grade 1	–	Excellent	
11	39/F	Blurred vision, eye pain	Inferomedial	Grade 2	–	Excellent	
12	55/F	Proptosis,	Superolateral	Grade 1	• Transient 3 rd nerve palsy	Good	

Table (2): Patient symptoms and signs in the study series.

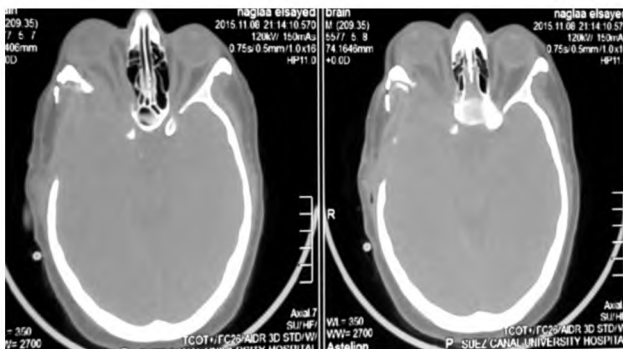
Symptoms and signs of clinical onset	Patients (12)
<i>Symptoms of clinical onset:</i>	
Proptosis	8 (67%)
Blurred vision	7 (58%)
Pain	3 (25%)
Diplopia	2 (17%)
Eyelid edema	1 (8%)
Tearing	1 (8%)
Other neurological symptoms (headache, seizures, etc.)	2 (17%)
<i>Signs at clinical examination:</i>	
Proptosis	11 (92%)
Afferent dysfunction	6 (50%)
Motility disturbance	4 (33%)
Optic disc pallor	3 (25%)
Optic disc edema	1 (8%)
Ptosis	1 (8%)
Cranial nerve dysfunction	1 (8%)



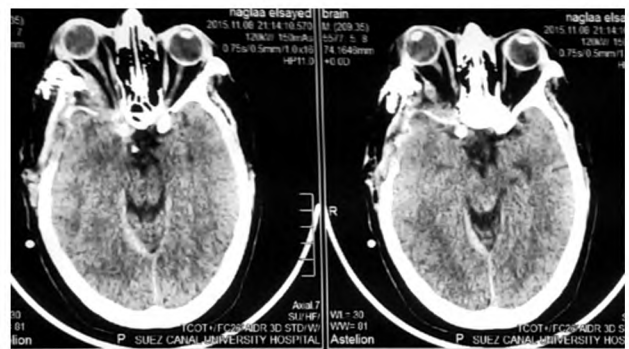
(A)



(B)



(C)



(D)

Fig. (1): (A) Female aged 32 years presented with right exophthalmos. (B) Axial CT scan showed a right superolateral sphenoorbital meningioma. (C, D) Postoperative axial CT bone window and brain after total removal of the tumor.

Discussion

Despite the progress of modern neuroimaging techniques and the technical advances in neurosurgery, meningiomas of the sphenoid wing involving the bone and the orbital structures are still a challenge for neurosurgeons. In fact, as stated by

Cophignon [14] some 30 years ago “to cure a patient from a sphenoorbital meningioma one has to remove the entire intradural tumor, all the involved dura, the orbital tumor, and the so-called hyperostosis, opening the intracranial and facial sinuses, if necessary”. Thus, removal is really complete only in some cases.

Several surgical controversies still exist concerning the management of sphenoidal meningiomas. These include the necessity of large bone flaps with resection of the zygomatic and malar bones, removal of tumor infiltration of the cavernous sinus and orbital apex, reconstruction of the orbital walls, and the opportunity to achieve radical resection in cases with more extensive orbital invasion. Also, the definition of complete removal is controversial for meningiomas of this region; in fact, the grading system according to Simpson cannot be applied without reservations, because of the often extensive dural and bone infiltration. Therefore, we may define complete removal as both Simpson grade I and II removals, because in many cases completeness of resection of the involved dura is difficult to define [15].

Many surgical approaches have been advocated for sphenoidal meningiomas, including lateral orbitotomy [16], supraorbital pterional [17], fronto-temporal-orbito-zygomatic [8,18-21], fronto-orbitomalar [1,22-24], and pterional [10,14]. The approach is chosen according to the extent of tumor invasion and the personal surgical experience. Although most sphenoidal meningiomas with osseous involvement extend to the orbital roof and/or lateral orbital wall, only about half of the cases in the reported series show an intraorbital tumor mass [6,7].

Sphenoidal meningiomas may be classified into 4 types (lateral or superolateral, medial or inferomedial, orbital apex, and diffuse) according to the location of the intraorbital tumor. In type I, lateral or superolateral, the tumor extends to the orbit through the sphenoid wing and the lateral orbital wall; the tumor mass is located mainly in the lateral or superolateral compartment of the orbit Fig. (1). The optic canal is preserved more often, although it may sometimes be involved. This type of sphenoidal meningioma may be approached through a lateral orbitotomy. In type II, medial or inferomedial, the tumor grows through the sphenoid bone and the floor of the orbital cavity; the intraorbital mass is located inferomedially. The optic canal may be invaded. These meningiomas can be removed through a supraorbital-pterional approach. In type III, orbital apex, the meningioma spreads through the optic canal, the superior orbital fissure, and the posterior orbital wall; the intraorbital tumor is located mainly in the orbital apex and the posterior part of the orbit. This type of meningioma can be approached through a supraorbital-pterional route, which allows the optic canal to be opened and the posterior orbital compartment to be controlled safely. In

type IV, diffuse, the tumor spreads to different intraorbital compartments and sometimes to the whole orbital cavity; thus, two or all three previous types may coexist. These meningiomas may be approached through a supraorbital-pterional approach, which allows adequate control of most of the tumor mass; a fronto-temporal-orbito-zygomatic approach may be necessary to improve the visualization of the tumor mass in cases of large inferomedial tumor components and invasion into the infra-temporal fossa. Thus, the surgical approach for removing sphenoidal meningiomas is decided based on the type of intraorbital tumor invasion. When using the supraorbital-pterional and fronto-temporo-orbito-zygomatic approaches the first surgical steps are entirely extradural, and involve drilling of the lesser sphenoid wing down to the superior orbital fissure and the lateral orbital wall. Then removal of the anterior clinoid process is performed and the optic canal is unroofed extradurally. If the periorbital is infiltrated, it is removed as extensively as possible. If there is infiltration of the extra ocular muscles, only the exophytic tumor is removed [25,26].

Recurrence and regrowth are one of the main problems of sphenoidal meningiomas. The causes of recurrence include inadequate resection of the involved bone, dura, and periorbital, as well as infiltration of the cavernous sinus and nervous and vascular structures of the medial aspect of the sphenoid wing, and particularly the tendency of these tumors to grow diffusely into foramina, fissures and interstices of the orbit and skull base [14]. Most series [15,27-34] report a variable recurrence rate from 8% [35] and 16-17% [30,32] to 60% [28] and even 82% [34]. The estimated probability of recurrence is 34% at 5 years and 54% at 10 years [36,37] the recurrence rate in our series is 34%.

The analysis of the surgical data of our series allows a discussion of few points. Drilling of the hyperostotic bone allowed excellent visualization and access [21]. Radical excision of the involved bone, dura, and periorbital must be accomplished, when possible; on the other hand, an aggressive effort to remove tumor infiltration of the cavernous sinus, orbital apex and superior orbital fissure is, in our opinion, not advisable; in fact, it carries the risk of severe functional eye disturbances and does not significantly influence the recurrence rate. Reconstruction of the orbital walls, although advised by some authors [9,24,38-42], is unnecessary. Pulsating exophthalmos and meningoceles did not occur in our twelve patients, as was also the case in over two hundred cases treated by Maroon et

al., [31]. Furthermore, the cosmetic results were good or acceptable in more than 90% of our cases.

Conclusion:

Spheno-orbital meningiomas are characterized by potentially extensive hyperostosis of the skull base. Successful resection necessitates drilling of the superior orbital fissure and orbital osteotomy within anatomical limitations. Complete resection was found to be limited by tumor remaining in the cavernous sinus and the optic canal.

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Conflict of interest: None.

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النتائج الإكلينيكية لإزالة أورام سحائية وتداية مدارية خبرة مؤسستنا

تعتبر الأورام السحائية الودية المدارية فئة معقدة من الأورام داخل الجمجمة التي تظهر مع فرط التجلط في الجناح الودي والمشاركة المدارية. وقد تم تعريفها تقليدياً على أنها أورام ثانوية في عظام المدار، تنشأ من الأم الجافية لعظم الجناح الودي.

الورم السحائي الجناح الودي قد ينمو وينتشر في اتجاهات مختلفة، مما يؤثر على مناطق مثل الجيوب الكهفية، الجيب الحصاني أو المدار.

قد يحدث تورط ثانوي في التجويف المداري من خلال العظم والقناة البصرية و/أو الشق المداري الفائق، على الرغم من أنه في معظم الحالات الغازية لا يمكن تحديد مسار إنتشار الورم.

يقدر أن الغزو داخل الحجاج يحدث في 39% إلى 50% من الأورام السحائية الجناح الودي. (3-7) ما يقرب من 9% من جميع الأورام السحائية داخل القحف لها توطين مداري.

يمثل الغزو المداري المكثف مع تسلل العظم المنتشر مشكلة جراحية مهمة. بسبب كثرة نشاطها في الجافية والعظام والمشاركة المدارية، كان من الصعب تاريخياً إجراء عملية إستئصال كافية للورم السحائي الودي-المداري، مما أدى إلى معدل تكرار مرتفع، يصل إلى 30-50%. (10-13) لقد أفاد بعض المؤلفين أن الجراثيم المرتبطة بالورم منذ فترة طويلة لا يمكن معالجتها جراحياً، وبالتالي لا ينبغي محاولة القيام بها.

تستعرض هذه الدراسة إثني عشر المرضى الذين يعانون من الأورام السحائية المدارية تعمل في قسم جراحة الأعصاب في مؤسستنا، الهدف هو مناقشة النهج الجراحي، والنتيجة، والمضاعفات وإعادة إنتشار هذا النوع من الأورام.