Romanian Neurosurgery | Volume XXX | Number 4 | 2016 | October - December

Article

Hyperostotic sphenoid wing meningioma en plaque: proptosis management

Wael K. Zakaria, Ahmed N. Taha
EGYPT

DE GRUYTER OPEN

DOI: 10.1515/romneu-2016-0079

DOI: 10.1515/romneu-2016-0079

Hyperostotic sphenoid wing meningioma en plaque: proptosis management

Wael K. Zakaria, Ahmed N. Taha

Neurosurgery Department, Mansoura University, EGYPT

Abstract: *Background*: En plaque sphenoid wing meningioma is morphological unique in comparison with other intracranial meningiomas, characterized by a carpet-like usually small soft tissue component which invade the dura and extensively involve the bone specially the sphenoid wing and orbit causing significant hyperostosis. *Patients & Methods*: A retrospective analysis of the clinical data, neuro-radiological features, and operative techniques of eighteen patients underwent transcranio-orbital approach sphenoid wing meningioma presented with proptosis during the period from September 2011 to April 2014 in the neurosurgery department, Mansoura University. Patients age ranged from 38 years to 54 years and there was sex males and twelve females. Chief complaints were progressive proptosis and visual acuity deficits. All patients were operated up on using a fronto-temporal approach with orbital decompression. The extent of tumor resection and postoperative complications were investigated. *Results*: Total removal was achieved in fourteen cases (77.8%) over a mean follow-up period of 36 months. Pathological examination showed that twelve patients (66.67%) were meningothelial meningiomas. After surgery, proptosis improved in all patients, visual acuity improved in fifteen patients (83.3%). Cerebrospinal fluid leakage was found in one patient. There were no operation-related deaths or other significant complications. Four patients had residual tumor (22.2%); two of them underwent surgical re-attack of the tumor and the other two cases were sent for gamma knife radio-surgery. *Conclusions*: Sphenoid wing meningioma en plaque, mainly meningothelial meningiomas, are characterized by the associated bony hyperostosis that gives them a distinct radiological appearance. The bony hyperostosis is of neoplastic nature and is responsible for many of the clinical manifestation of such tumors and hence should be totally drilled to achieve cure and avoid recurrence. Extensive tumor removal is crucial for correction of proptosis and adequate visual decompression to achieve satisfactory cosmetic and functional outcome.

Introduction

Intracranial meningiomas can be classified into two morphological types; global and flat. The terms "meningioma en masse" and "meningioma en plaque" was initially described by Cushing and Eisenhardt (1938) to differentiate between them. (15) The most frequent type encountered is meningioma en masse while meningioma en plaque is a rarer subgroup characterized morphologically by a thin, widespread, carpet like mass infiltrating the dura with variable degrees of bone invasion causing progressive hyperostosis which may involve the bone orbit causing proptosis. Meningioma en plaque almost always involve the sphenoid wing. (2, 4, 17) Sphenoorbital or en plaque meningiomas is considered as a distinct entity which is different from sphenoid wing meningiomas. Such morphological characteristic of these tumors put them in differential diagnosis with others osseous skull lesions as fibrous dysplasia, osteoma and some metastatic lesions. (2, 3, 4, 15)

Hyperostosis is a common phenomenon occuring in different meningiomas with incidence ranging from 25 to 49% of meningiomas.it is most commonly seen in sphenoid wing and convexity meningioma. The biological behavior and growth pattern of spheno-orbital meningiomas is variable and unpredictable. In some patients; the tumor is slowly growing and symptoms are minimal while in others; the disease progression is rapid with marked affection of vision, significant proptosis, and disfiguring. (2-7, 11-19)

Spheno-orbital meningiomas usually involve important areas including the orbit, superior orbital fissure, optic canal, the cavernous sinus. Tumor commonly compress the optic nerve and the cranial nerves withen the cavernous sinus. (1, 2, 6, 9, 10) Extensive dural, bone, and orbital involvement makes total resection of such tumors not achievable and hazardous hence some authors in previous report reccomended conservate approach in dealing with these tumors and surgery should be restricted to rapidly progressive lesions. Total resection of these tumors necessiate extensive bone drilling to the sphenoid and orbital bone with may cause significant cranial deformity and cosmotic difigurement and hence adequate orbital and skull reconstruction is essential for good functional and cosmotic outcome. Early and aggressive surgical resection is considered by many authors the only to cure the patient and to achieve sound cosmotic outcome and to correct proptosis as longstanding proptosis is not surgically correctable. (1-4, 6-8, 11-19) Radiation therapy either conventional or radio-surgey has been described as an alternative or adjuvant to non radical surgery to achieve tumor control but the proximity to optic pathway increase the risk for progressive visual deterioration. (2, 13, 15)

Clinical material and methods

Patients characteristics:

Between 2011 and 2014, eighteen patients with hyperostotic sphenoid wing meningiomas underwent surgery at the neurosurgery department, Mansoura University. Patients with extensive

hyperostosis, en plaque dural invasion, minimal intracranial tumor were included in the study. We did not include patients with nonhyperostotic sphenoid wing meningiomas, hyperostotic sphenoid wing meningiomas with a moderate or large intracranial tumor portion, or clinoid or primary optic nerve sheath meningiomas.

The most common presentation (Table I) was proptosis. Other symptoms and signs included deficits of visual acuity (twelve patients (66.67%)), headache (eight patients) and seizure (one patient).

Radiographic studies

All patients received pre-operative 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 bone and involving adjacent bone, including the middle cranial fossa bone, the lateral orbital wall and the orbital roof, the walls of the sphenoid sinus and ethmoidal cells. The edges of hyperostosis were rough and brush-like. On CT scans, the soft-tissue component of meningioma was carpet-like, isodense and homogeneously enhanced after contrasted.

MRI showed T1 intensity was isointensity or hypointensity, T2 intensity was hyperintense. Gadolinium enhancement showed typical features for meningioma. The dura in the soft-tissue component was homogeneously enhanced, but the hyperostotic bone was not enhanced. Postcontrast fat suppression T1-weighted MRI was useful to evaluate the extension of dural enhancement and soft-tissue involvement of the orbital content in those patients with tumor extending into the orbit. Post-operative follow up imaging studies (both computed tomography (CT) and magnetic resonance Imaging (MRI) scans) were performed to assess the extent of tumor resection and to plan for any subsequent management.

Surgical technique

Sixteen patients were operated up on via the classic pterional approach and in two patient the orbito-zygomatic approach was performed. Ipsilateral fronto-temporal craniotomy under general anesthesia was used for all patients. Patients were placed in a supine position with the head tilted 20 degrees to the contralateral side. The scalp incision began 1cm superior to the anterior aspect of the auricle and curved anteriorly, ending at the hairline, 1 cm from the midline. Care was given to avoid injury to the temporal branch of the facial nerve. Initially we removed the extracranial soft tissue part of the tumor which was fequently encountered then we did extensive drilling of the all invaded bones of the lesser and greater sphenoid wings. In most cases; it was necessary to remove the anterior clinoid process and optic canal was opened when it was involved by the hyperosteosis. The infilterated dura was removed totally in all cases and we removed the small intradural soft tissue component. The component of the tumor in the cavernous sinus and the superior orbital fissure were left not to endanger the important structures to avoid significant postoperative morbidies. Dural graft and abdominal fat was used to achieve watertight dural closure. Orbital reconstruction was done using muscle and split calvaric bone.

Results

Our series included 12 women and 6 men, ranging in age between 36 and 54 years. The clinical manifestations present at the onset of disease in these cases are summarized in Table 1. The most common presenting symptom was proptosis, which was observed in 15 patients (83.3%), followed by progressive visual loss in 12 patients (66.67%), and headache in 8 patients (44.4%). Preoperative neuroradiological evaluations demonstrated hyperostosis in all 18 patients. The sphenoid ridge was a constant location for hyperostosis (observed in all cases), followed by the lateral and superior walls of the orbit, in 77.78 and 33.3% of patients, respectively (Table 2).

Table 1

Presenting symptoms in 17 cases of sphenoid wing meningioma

Symptoms	*Number of cases (%)*
Proptosis	15 (83.3%)
Progressive vision loss	12 (66.67%)
Headache	8 (44.4%)
Diplopia	2 (11.1%)
Eye swelling	1 (5.56%)
Seizures	1 (5.56%)
Ptosis	2 (11.1%)

Table 2

Correlation of neuro-imaging and histological findings of hyperostosis

Case No.	sex	Areas of Hyperostosis on Neuroimages	Histological Confirmation
1	F	sphenoid ridge, lat & sup walls of orbit	Yes
2	M	sphenoid wing, lat wall of orbit	Yes
3	F	sphenoid ridge, optic canal	Yes
4	M	sphenoid ridge, lat & sup walls of orbit	Yes
5	F	sphenoid wing, lat wall of orbit	Yes
6	M	sphenoid ridge	Yes
7	F	sphenoid ridge, lat & sup walls of orbit	Yes
8	F	sphenoid wing, lat wall of orbit	Yes
9	F	sphenoid wing, lat wall of orbit	Yes
10	M	sphenoid ridge	Yes
11	F	sphenoid wing, lat wall of orbit	Yes
12	F	sphenoid ridge, lat & sup walls of orbit	Yes
13	F	sphenoid wing, lat & sup walls of orbit, ant wall of middle fossa	Yes
14	F	sphenoid wing, lat wall of orbit	Yes
15	M	sphenoid ridge, lat & sup walls of orbit	Yes
16	M	sphenoid ridge	Yes
17	F	sphenoid wing, lat wall of orbit	Yes
18	F	sphenoid wing, lat wall of orbit	Yes

Table 3

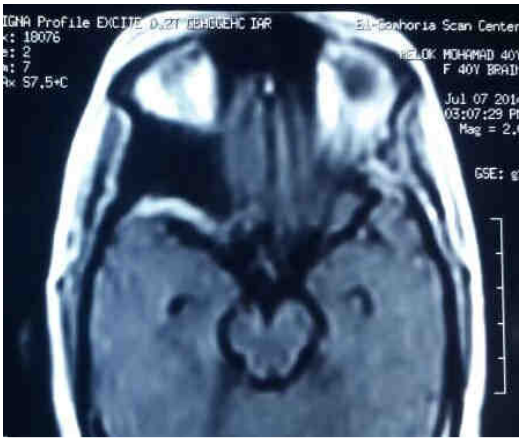
Histological tumor growth patterns and pathological bone invasion

Case No.	*Tumor histology*	*WHO grading*	*Bone invasion*
1	meningotheliomatous	**I**	**Yes**
2	meningotheliomatous	**I**	**Yes**
3	meningotheliomatous	**I**	**Yes**
4	transitional	**I**	**No**
5	meningotheliomatous	**I**	**Yes**
6	chordoid	**II**	**Yes**
7	meningotheliomatous	**I**	**Yes**
8	chordoid	**II**	**Yes**
9	meningotheliomatous	**I**	**Yes**
10	meningotheliomatous	**I**	**Yes**
11	meningotheliomatous	**I**	**Yes**
12	transitional	**I**	**Yes**
13	mixoid	**I**	**Yes**
14	meningotheliomatous	**I**	**Yes**
15	meningotheliomatous	**I**	**Yes**
16	meningotheliomatous	**I**	**Yes**
17	Meningotheliomatous	**I**	**Yes**
18	transitional	**I**	**Yes**

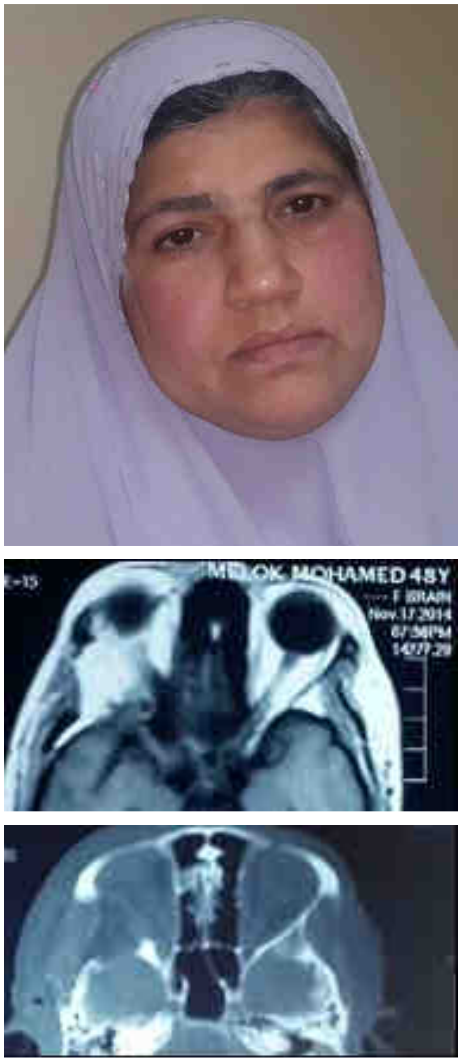
Surgical Results

There were no deaths related to the surgical procedures in this series of cases. Cerebrospinal fluid rhinorrhea happened in one patient and was improved after insertion of lumbar drain for three days. Preoperative proptosis improved in all patients (Figure 1), and 83.3% of patients had improved vision after surgery. One patient showed worsening of vision after surgery and hadn't improved on subsequent follow up.

Total tumor removal was achieved in 14 patients (77.8%). In four patient; we found residual tumor (22.2%) on follow up imaging. In two patient the residual tumor involved no drilled bone hyperosteosis and the tumor progressed on subsequent imaging and 2nd surgery was performed for both of them. The other two patient were refered for gamma knife radiosurgery as the residual tumor was left in the cavernous sinus.



Preoperative MRI showed extensive bone invasion with small intracranial soft tissue component



Postoperative CT and MRI showed tumor excision and proptosis subsidence on patient

Figure 1 - 48 female patient with excised large sphenoid ridge meningioma presented with progressive proptosis

Histological Results

The results of histopathological evaluation are summarized in Table 3. Bone invasion was confirmed histologically, but most of the hyperostotic bone was removed by drilling. In 16 of the 18 cases, the tumors were histologically benign (WHO Grade I); the other two tumors showed some chordoid features and were thus classified as WHO Grade II. Meningotheliomatous growth patterns predominated in most of the tumors (12 of 18). Bone invasion by the tumor was confirmed histologically in 17 of 18 patients. Histological examination did not show tumor invasion into the bone in one case, but this finding can be attributed to a loss of involved bone through aggressive drilling and the consequent evaluation of an inappropriate specimen because of the drilling.

Discussion

The cure for hyperosteotic sphenoid ridge meningiomas was difficult to be achieved as shown in different case series and this was attributed to their extensive bone, orbital, and neural involvement. They tumors usually grow following the contour of the inner table of the skull base with involvement of a large surface area. The characteristic hyperostosis is often disproportionate to the relatively small tumor. The cause for hyperosteosis is debatable in the literature but the widely accepted theory in most recent publications is that it's tumor invasion rather than bone irritation or just new osteoblastic bone formation. Hence, cure would not be possible without extensive bone drilling to remove all the involved bone. These tumors also insinuate themselves into the surrounding structures by way of natural openings. Extension into the orbit, superior orbital fissure or optic canal adds challenge to such tumors making the overall prognosis less favorable. (1, 2, 9, 10, 16, 18, 19)

The clinical presentation of such challenging tumors is mostly due to associated hyperostosis and the most common presentation in different series is unilateral slowly progressing proptosis. Other manifestations include headache, visual impairment, seizures and affection of ocular motility. In our series the clinical presentations of those lesions matches with what have reported in other case series. (2, 6, 7, 11, 15, 17)

The development of proptosis is reported to occur in up to 100% of cases according to different series and it is explained either due to direct tumor invasion into the orbit, hyperosteosis of the bony orbit and / or cavernous sinus and superior orbital fissure invasion causing impairment of the venous drainage of the orbit. For correction of proptosis; it is necessary to do extensive excision of the invaded bone which should also remove the involved bone of the optic canal and superior orbital fissure to improve symptoms caused by cranial nerves compression especially visual impairment and ocular motility affection. Both magnetic resonance and CT images are essential to allow precise determination of anatomical relationships of these tumors and pattern of their extensions enabling planning of the surgical treatment. (2-6, 14, 15, 17-19)

The goal of surgery is to achieve total resection of the entire tumor both the soft and bony component. Hyperostotic bone should be extensively removed to prevent tumor recurrence. (2-6, 14, 15, 17-19) Most of the cases in our study were operated up on via the classic pterional approach while in 2 cases the orbito-zygomatic approach was performed. Drilling of the hyperosteotic bone improved the visualization making removing the zygomatic arch not necessary in most of the cases.

Despite aggressive bone drilling to achieve to resection of such tumors is usually attempted however total resection was not always feasible and there is still a potentiality for tumor recurrence. Tumor extension into the cavernous sinus and the superior orbital fissure hinders the attempt for total tumor resection. Meningiomas in the cavernous sinus encircle the cranial nerves and vascular structures within sinus and manipulation of such tumors inside sinus increase the possibility of permanent ophthalmoplegia and even vascular injuries. (1, 2, 4, 6, 11, 15, 16, 19) We have four cases in our study in whom there was residual tumor on subsequent imaging; two of them due to missed invaded bone on the initial surgery and were followed up and 2nd surgery was performed for tumor progression. The other two cases the tumor was left in the cavernous sinus and was sent for gamma knife radio-surgery. Furthermore; extensive drilling of sphenoid bone and orbit had a significant risk for postoperative cranial deformities and cranial and orbital reconstruction should be considered with surgical planning. On the other hand; aggressive bone drilling was not able to correct long standing tumor related proptosis. (2, 3, 7, 12, 15, 19)

The resection of the involved hyperostotic bone from the sphenoid and the orbit disrupts the structural integrity that the sphenoid wing provides to the orbit and the anterior and

middle cranial fossa. Lack or improper orbital reconstruction have its potential risk for aditional postoperative morbidities ranging from meningoceles, postoperative infection up to complete ophthalmoplegia and a nonfunctional globe. Hence, proper surgical reconstruction is as important as adequate tumor resection for achievement of a successful outcome. (2, 8, 12, 14, 18, 19) As in all the cases in our study, it was essential to remove all in invaded dura of the base, we considered thefirest step to achieve sound reconstruction and prevent the potential morbidities is to perform water tight dural closure. We prefer to use autologus dural graft either pericranium or fascia lata as needed in adition to autologus fat graft. Skeletal calvarial and orbital reconstruction is another crucial step that we performed in all our cases. Although many authors described various techniques and variable reconstructive materials, we prefered to do autologus skeletal reconstruction in all of our cases presented in this study. The result was acceptable without any significant functional or cosmotic sequalae and avoidance the potential risk of using synthetic materials to achieve such reconstruction. Although in other studies, many authors utilized different synthetic materials that proved to be safe and effective in achievement of cosmotically sound orbital reconstruction and their results are more favorable than we do but all must be individually fashioned pre-operatively to have the symetrical shape for the orbit that have to be reconstructed. Unfortunately, such technology wasnot feasible for us apart from the potential problem with management of potentially possible recurrence and the need for subsequent imaging studies. (2, 8, 12, 14, 15, 18, 19)

Conclusions

Early and radical surgery for hyperosteotic sphenoid ridge meningiomas is essential for both achievement of good cosmetic and functional outcome and prevention of tumor recurrence. However extensive bone involvement and tumor extension into cavernous sinus and superior orbital fissure hinder the capability for total resection and increase the potential risks for aggressive surgical interference. Radio-surgery is a useful alternative for residual tumor in the cavernous sinus. Adequate dural repair and orbital reconstruction is important to achieve good cosmetic and functional outcome.

References

1.Abdel-Aziz KM, Froelich SC, Dagnew E, Jean W, Breneman JC, Zuccarello M, et al.: Large sphenoid wing meningiomas involving the cavernous sinus: conservative surgical strategies for better functional outcomes. Neurosurgery 54:1375–1384, 2004

2.Bikmaz, K, Mrak R, Al-Mefty O: Management of bone-invasive, hyperostotic sphenoid wing meningiomas. J Neurosurg 107: pp. 905-912, 2007

3.Boari N, Gagliardi F, Spina A, Bailo M, Franzin A, Mortini P.: Management of spheno-orbital en plaque meningiomas: clinical outcome in a consecutive series of 40 patients. Br J Neurosurg; 27(1):84–90, 2013

4. De Jesús O, Toledo MM: Surgical management of meningioma en plaque of the sphenoid wing. Surg Neurol 55:265–269, 2001.

5.Elder, JB, Atkinson R, Zee CS, Chen TC: Primary intraosseous meningioma. Neurosurg Focus 23: pp. E13, 2007

6.Honig S, Trantakis C, Frerich B, Sterker I, Kortmann RD, Meixensberger J.: Meningiomas involving the

sphenoid wing outcome after microsurgical treatment–a clinical review of 73 cases. Cent Eur Neurosurg;71(4):189–98, 2010

7.Honeybul S, Neil-Dwyer G, Lang DA, Evans BT, Ellison DW: Sphenoid wing meningioma en plaque: a clinical review. Acta Neurochir (Wien) 143:749–758, 2001

8.Leake D, Gunnlaugsson C, Urban J, Marentette L: Reconstruction after resection of sphenoid wing meningiomas. Arch Facial Plast Surg 7:99–103, 2005

9.Lee JH, Sade B, Park BJ: A surgical technique for the removal of clinoidal meningiomas.Neurosurgery 59:ONS108 ONS114, 2006

10.Margalit NS, Lesser JB, Moche J, Sen C: Meningiomas involving the optic nerve: technical aspects and outcomes for a series of 50 patients. Neurosurgery 53:523–533, 2003

11.Marcus H, Schwindack C, Santarius T, Mannion R, Kirollos R. Image-guided resection of spheno-orbital skull-base meningiomas with predominant intraosseous component. Acta Neurochir (Wien);155(6):981–8, 2013

12.Mirone G, Chibbaro S, Schiabello L, Tola S, George B: En plaque sphenoid wing meningiomas: recurrence factors and surgical strategy in a series of 71 patients. Neurosurgery 65:6 Suppl 100–109, 2009

13.Oya S, Sade B, Lee JH: Sphenoorbital meningioma: surgical technique and outcome. J Neurosurg 114(5):1241–9. 2011.

14.Pieper, DR, Al-Mefty, O, Hanada, Y, Buechner, D: Hyperostosis associated with meningioma of the cranial base: secondary changes or tumor invasion. Neurosurgery 44: pp. 742-746, 1999.

15.Ringel F, Cedzich C, Schramm J.: Microsurgical technique and results of a series of 63 spheno-orbital meningiomas. Neurosurgery;60 (4 Suppl 2):214–221; discussion 221–12, 2007.

16.Roser F, Nakamura M, Jacobs C, Vorkapic P, Samii M: Sphenoid wing meningiomas with osseous involvement. Surg Neurol 64:37–43, 2005.

17.Scarone P, Leclerq D, Heran F, Robert G. Long-term results with exophthalmos in a surgical series of 30 sphenoorbital meningiomas. Clinical article. J Neurosurg; 111(5):1069–77, 2009.

18.Schick U, Bleyen J, Bani A, Hassler W: Management of meningiomas en plaque of the sphenoid wing. J Neurosurg 104:208–214, 2006.

19.Shrivastava RK, Sen C, Costantino PD, Della-Rocca R: Sphenoorbital meningiomas: surgical limitations and lessons learned in their long term management. J or Neurosurgery 103 (3): 491-497, 2005.