Orbital Meningiomas

Meningiomas Orbitários

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ABSTRACT

Orbital meningiomas usually invade the orbit as an extension of the sphenoid wing meningiomas, clinoidal meningiomas, cavernous sinus meningiomas and tuberculum sella tumors. They also arise into the orbit originating from the optic sheath or as ectopical lesions. The authors present a review of clinical aspects and surgical treatment of the orbital meningiomas. **Material and methods**: *The authors present a literature review of the anatomical, clinical, and surgical aspects of the orbital meningiomas, add illustrative cases, pointing their principal concerns about the treatment of such tumors.* **Results:** *Exophthalmos and unilateral visual loss are the most common features of the orbital meningiomas. There are two important surgical routes to approach such tumors, which are extracranial (transorbital) and transcranial approaches. Recurrence is higher in sphenoorbital meningiomas and related to bone and neurovascular invasion.* **Conclusions:** *The anatomical structures involved by orbital meningiomas challenge the microsurgical resection. Total removal should be the goal at the first surgical approach. Orbital reconstruction should be performed in cases where the orbital floor is not preserved or in extensive lateral and roof removal with no preservation of the periorbit.*

Keywords: *Orbital tumors, orbital anatomy, orbital meningiomas, skull base surgery.*

RESUMO

Meningeomas orbitários invadem a órbita, na maioria dos casos, como uma extensão de meningeomas da asa do esfenóide, meningeomas do seio cavernoso, meningeomas da clinóide e do tubérculo da sela. Eles também podem ser originados do revestimento dural do nervo óptico ou como lesões ectópicas intraorbitais. Os autores apresentam uma revisão dos aspectos clínicos e cirúrgicos dos meningeomas orbitários. **Material e métodos:** *Os autores apresentam uma revisão da literatura dos aspectos anatômicos, clínicos e cirúrgicos dos meningeomas orbitários, com casos ilustrativos, apresentando suas principais preocupações no manejo destes tumores.* **Resultados:** *Exoftalmia e perda visual unilateral são os achados mais frequentes nos meningeomas orbitários. Existem duas importantes vias de acesso para tais tumores, que são a extracraniana (transorbitária) e a transcraniana. A recorrência no grupo dos meningeomas esfeno-orbitários é maior e relacionada a invasão do osso e das estruturas neurovasculares.* **Conclusões:** *As estruturas anatômicas envolvidas pelos meningeomas orbitários tornam estes tumores verdadeiros desafios para ressecção microcirúrgica, sendo a ressecção total o objetivo durante a primeira cirurgia. A reconstrução orbitária deve ser efetuada nos casos em que o assoalho orbitário não é preservado, assim como em casos de ressecções extensas da face lateral e do teto da órbita, sem a preservação da periórbita.*

Palavras-chave: *Tumores orbitários, anatomia da órbita, meningeomas orbitários, cirurgia de base de crânio.*

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INTRODUCTION

Orbital tumors are classified according to their original site in: primary lesions, arising from the orbit itself; secondary lesions, originating from surrounding structures; and metastatic lesions. Orbital meningiomas comprise 3 to 10% of all intraorbital tumors. [6,8] They usually invade the orbit as an extension of the sphenoid wing , cavernous sinus , clinoid and tuberculum sella tumors. They may also arise from the optic sheath or as ectopical rests of the arachnoid cells within the retrobulbar space [8]. The anatomical structures involved by orbital meningiomas make them formidable challenges for microsurgical resection. The authors present a review of clinical aspects and surgical treatment of the orbital meningiomas.

EPIDEMIOLOGY

Meningiomas represent 18 to 20% of all intracranial tumors. Primary orbital meningiomas comprise 0.4 to 2% of all meningiomas and secondary lesions present the distribution according to their initial sites. Sphenoid wing meningiomas comprise between 16 to 20% of the intracranial meningiomas and the suprasellar region originates 7 to 30% of all meningiomas. Such data evidence that orbital meningiomas are secondary lesions most of the time. Females are more affected, with 73 to 84% of the cases. Meningiomas are uncommon in children and neurofibromatosis is a possible risk factor associated to an orbital meningioma in infants. [5 6,8]

CLINICAL ASPECTS

Primary and secondary orbital meningiomas present similar clinical presentation. Exophthalmos and unilateral visual loss are the most common features described in the literature. [(5)] Diplopia, pain, swelling and headaches are frequently observed in such cases. Nausea and vomiting are symptoms associated to intracranial hypertension. Optic nerve atrophy can be observed as a result of direct compression of the nerve. Diplopia is secondary to direct involvement of the cranial nerves or extraocular muscle disfunction. In large secondary orbital meningiomas, intracranial mass effect and hypertension can produce papilledema in both optic nerves. [5]

ANATOMICAL CONSIDERATIONS

ORBITAL BONE ANATOMY

The orbit is composed by 7 bones: frontal, ethmoid, sphenoid, maxilla, zygomatic, lacrimal and palatine. The superior border of the orbital opening is formed by the frontal bone, which presents a notch or foramen for the supratrochlear and supraorbital vessels and nerves. The medial border is formed by the frontal bone superiorly and the frontal process of the maxilla inferiorly. Lateral border is formed by the frontal process of the zygomatic bone and a small portion superiorly by the zygomatic process of the frontal bone. The inferior border is composed by the maxilla medially and the zygomatic bone laterally.[16] (Fig. 1)
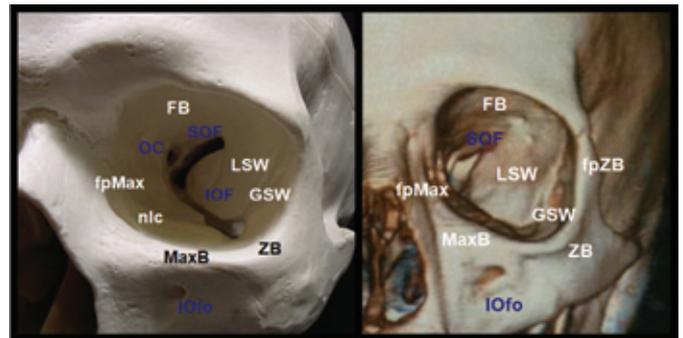

Fig. 1. - Orbital Bone Anatomy (frontal view). Left - Skull; Right - 3D CT. FB - frontal bone; SOF - superior orbital fissure; OC - optic canal; LSW - lesser sphenoid wing; IOF - inferior orbital fissure; GSW - greater sphenoid wing; fpZB - frontal process of the zygomatic bone; fpMax - frontal process of maxillar bone; nlc - nasolacrimal canal; MaxB - maxillar bone; ZB - zygomatic bone; IOfo - infraorbital foramen.

The orbital roof is composed by the orbital plate of the frontal bone and the lesser wing of the sphenoid bone. The orbital floor is formed by the orbital plate of the maxilla, the orbital surface of the zygomatic bone and the orbital process of the palatine bone. The lateral wall is formed by the greater sphenoid wing and the frontal process of the zygomatic bone. Posteriorly, the inferior orbital fissure (IOF) separates the lateral wall and the orbital floor. The superior orbital fissure (SOF) separates the lateral wall and the roof of the orbit. The nasolacrimal canal perforates the anteromedial portion of the orbital floor.

At the posterior orbital portion there are three bone apertures, which allow the neurovascular structures to access the orbit from the temporal fossa, cavernous sinus, suprasellar region and infratemporal fossa. The cavernous sinus is directly related with the orbit and the cranial nerves III, IV, V and VI enter the posterior portion of the orbit through the superior orbital fissure. (Fig. 2)
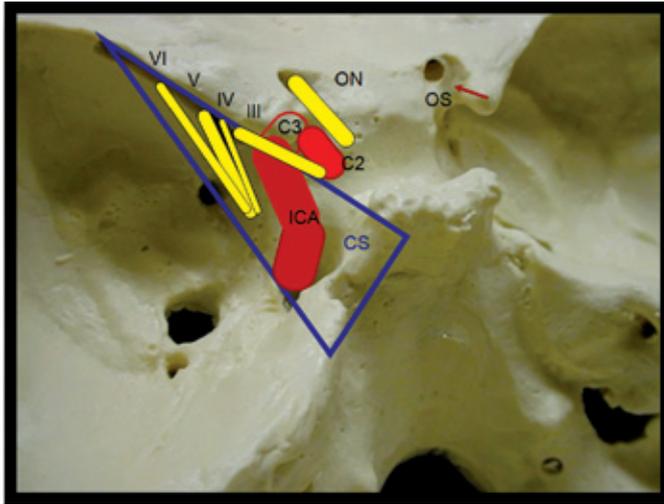
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Fig. 2. - Cranial nerve anatomy at SOF and cavernous sinus - digital schematic representation. ON - optic nerve; ICA - internal carotid artery; CS - cavernous sinus; C2 and C3 - portions of the ICA; III - oculomotor nerve; IV - trochlear nerve; V - trigeminal nerve; VI - abducens nerve; OS - optic strut.

The optic canal is located inferiorly to the anterior clinoid process and the optic nerve leaves the canal medially to the C2 portion of the internal carotid artery. [4] (Fig. 2)

The optic canal opens into the superomedial portion of the orbital apex and transmits the optic nerve and ophthalmic artery. The canal is situated at the junction of the lesser wing with the sphenoid body, and it is separated from the superior orbital fissure by the optic strut, which is a bridge of bone originated from the inferior aspect of the anterior clinoid directed to the sphenoid body. [16] (Fig. 2)

The superior orbital fissure is situated between the lesser and greater wings of the sphenoid bone. The SOF has a triangular shape with a medial portion larger and posterior to the lateral portion (apex). (Fig. 3). The fissure communicates the orbit, the middle cranial fossa and the cavernous sinus. (Fig. 2 and 3)[16]

The four rectus muscles (superior, inferior, medial and lateral) arise from the annular tendon (tendineous ring) which it is attached to the upper, lower and medial margin of the optic canal and, laterally, to the lateral edge of the SOF. [16] (Fig. 3)

The inferior orbital fissure communicates the orbit and the pterygopalatine fossa, posteromedially. The anterolateral portion of the fissure communicates the orbit and the infratemporal fossa. The infraorbital nerve, zygomatic branches of the maxillary nerve (V2), some branches of the internal maxillary artery and inferior ophthalmic vein are the structures passing through the IOF. [16]

CRANIAL NERVES ANATOMY

The oculomotor nerve originates two roots at the SOF, which enter the orbit inside the annular tendon with the extra-ocular muscles and vessels, at the central portion of the SOF. Such central region is called oculomotor foramen. [4,16] (Fig. 3)
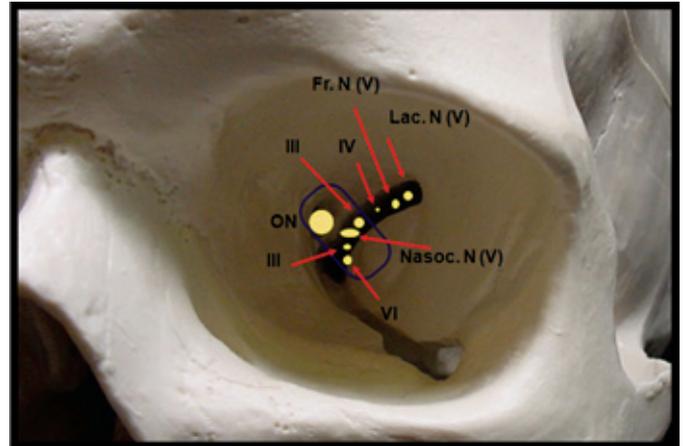

Fig. 3 -Orbital Cranial Nerve Anatomy (frontal view - Superior Orbital Fissure) III - oculomotor nerve; IV - trochlear nerve; Fr. N - frontal nerve; Lac. N - lacrimal nerve; Nasoc. N - nasociliary nerve; V - trigeminal nerve; VI - abducens nerve; ON - optic nerve. The blue line corresponds to the annular tendon.

The trochlear nerve enters the superior portion of the SOF, outside the tendinous ring, medially to the frontal nerve. [4,16] (Fig. 2)

The ophthalmic division (V1) of the trigeminal nerve originates three roots at the superior orbital fissure, which are: the nasociliary nerve, which enters the posterior aspect of the orbit inside the tendinous ring; the lacrimal and the frontal nerves, which enter the orbit outside the annular tendon. The frontal nerve is located between the trochlear nerve, medially, and the lacrimal nerve, laterally.[16] (Fig. 2)

The abducens nerve enters the inferior portion of the SOF, inside the annular tendon. [4,16] (Fig. 2)

VASCULAR ANATOMY

The anterior bend of the internal carotid artery (ICA) passes along the posterior edge of the SOF at the posterior surface of the optic strut. The artery turns upward in the subarachnoid space, passing medially to the anterior clinoid. (Fig. 2) The ophthalmic artery is the principal arterial root to the orbit. It arises just above the cavernous sinus, from the medial portion of the anterior bend of the ICA. In about 8% of the cases, the artery originates into the cavernous sinus. The artery crosses the optic canal inferior to the optic nerve passing through the annular tendon. After crossing the optic canal the ophthalmic artery passes above the optic nerve (85% of the cases), and originates several branches: central retinal, lacrimal, long and

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Revisão

short ciliary, meningeal, supraorbital, infratrochlear, supratrochlear, palpebral, dorsal nasal, anterior and posterior ethmoidal arteries. The central retinal artery is a terminal branch of the ophthalmic artery.[16]

The cavernous sinus is related with the posterior and medial portion of the SOF. (Fig. 2) The superior ophthalmic vein drains the superomedial portion of the orbit and the inferior ophthalmic vein drains the inferolateral area. These veins are connected by large anastomotic veins to the facial and angular veins at the anterior border of the orbit. The superior and inferior ophthalmic veins usually drain into a single vein, which drains to the cavernous sinus. [4,16]

Pathological Considerations

The primary orbital meningiomas are originated from the optic sheath and consist of the transitional subtype, and psammomatous changes are observed in some tumors. Histological subtypes as fibrous, papillary and anaplastic meningiomas are uncommon as primary orbital lesions[5]

Secondary meningiomas involving the spheno-orbital region present different histological subtypes and typically produce bone invasion and hyperostosis.[5]

According to Al-Mefty, hyperostosis represents bone invasion by the tumor. The author and colleagues analyzed the radiographic, surgical and histological aspects of the cranial base meningiomas and demonstrated such hypothesis.[14] Other theories about the origin of the hyperostosis described in the literature are: vascular disturbances of the bone, stimulation of the osteoblastic function in the normal bone by the tumor-secreting factors, tumor production of bone, previous trauma and bone reaction without tumor invasion.[5]

Surgical Considerations

"The best time is the first time."
Ossama Al-Mefty

As a general rule, the surgical treatment of meningiomas should consider the radical removal as the first and best alternative, in order to achieve the cure of the patients. In such cases, this goal should be seek during the first operation. In orbital meningiomas, radical removal of the bone involvement by the tumors is essential to achieve the cure of such lesions.

Considering that the bone hyperostosis represents tumor invasion, the surgical removal should include all orbital bone abnormalities. [1,2,7,9,10,11,12,18,19,20]

SURGICAL APPROACHES

The approaches are divided in two major groups: extracranial and transcranial approaches. The lesions located inside the periorbit in the anterior two-thirds of the orbit, can be removed by extracranial routes. The four principal transorbital approaches are: anterior orbitotomy with or without osteotomy; lateral orbitotomy; medial orbitotomy and a combination of medial and lateral orbitotomy.[3] Those meningiomas involving the orbital apex, medial to the optic nerve and lesions with intracranial extension should be removed by transcranial routes.[16] Some lesions with infratemporal and maxillary components should be managed with extended transcranial approaches.

The earlier transcranial approaches removed the frontal and frontotemporal bone flap, preserving the supraorbital rim. However, for larger tumors, after the development of skull base surgery techniques, the supraorbital and cranio-orbital zygomatic (COZ) approaches are more appropriate for radical removal.

Meningiomas of the optic sheath confined to the orbit should be removed by supraorbital approach. Cranio-orbital meningiomas, should be approached by COZ route.[1,2]

The two principal transcranial approaches are described as it follows:

SUPRAORBITAL APPROACH

The patient is placed supine, with head elevated 20 to 30 degrees, and kept straight. The head should be fixed in the Mayfield headrest, hyperextended to allow the frontal lobes to fall backward, using the gravity as a natural retractor. The skin incision is placed 1 cm anterior to the tragus and extends to the superior temporal line on the opposite side, behind the hairline for cosmetic reasons. The superficial temporal artery courses posteriorly and the facial nerve branches are located anterior to the incision. (Fig 4)

A large pediculated pericranial flap, vascularized by supraorbital and frontal vessels is reflected over the scalp flap. The pericranial flap should be prepared for dural and skull base reconstruction after the tumor removal. (Fig 5) The subfascial temporal dissection, posterior to the upper branches of the facial nerve is reflected anteriorly. The upper portion of the temporal muscle is reflected inferior and posteriorly to expose frontal, sphenoid and zygomatic bones.

The supraorbital bone flap begins with the keyhole burr, located at the frontosphenoidal junction, posterior to the zygomatic

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process of the frontal bone. The second burr hole is made above the supraorbital rim, medially to the supraorbital foramen. This hole should be as small as possible for cosmetic reasons. The two burr holes are connected with two bone cuts. The first cut goes through the frontal bone, 4 cm superior to the supraorbital rim. The second cut connects the two burr holes crossing below the supraorbital rim. This cut may be made using fine drill or Gigli saw. The cut using Gigli saw requires protection of the orbit contents with a brain spatula and the orbital rim and zygomatic process are cut together. [1]
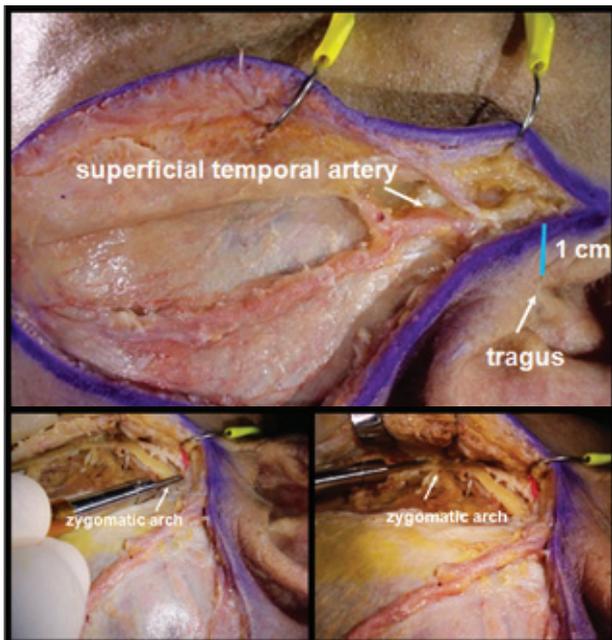

Fig. 4 - **Left -** Anatomical specimen dissection - Preservation of the superficial temporal artery and facial branches. **Right -** Cutting zygomatic arch in the anterior and posterior portions with preservation of the masseter muscle.
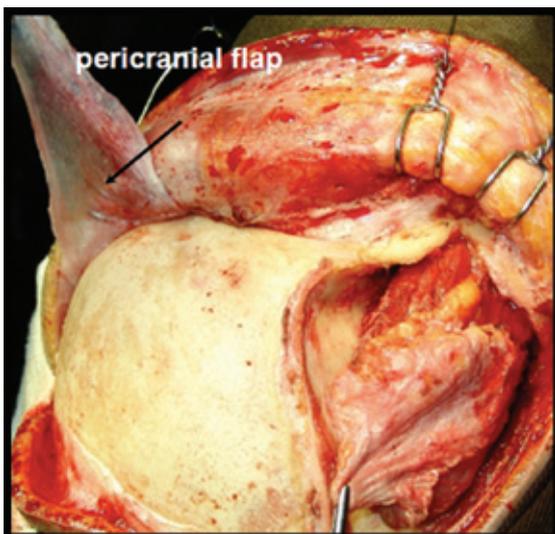

Fig. 5 - Exposition of the zygomatic arch and preparing the pericranial flap

CRANIO-ORBITAL ZYGOMATIC APPROACH

The patient is placed supine, with head elevated 20 to 30 degrees. The head is fixed in the three-point headrest rotated 30 degrees to the opposite side of the tumor. The skin incision begins 1 cm anterior to the tragus and extends behind the hairline to the contralateral superior temporal line, preserving the superficial temporal artery and superior branches of the facial nerve. (Fig.4) The pericranial flap is dissected to expose the supraorbital nerve at the supraorbital notch or foramen, which is drilled away to release the nerve. The zygomatic arch is dissected and the zygoma is sectioned with oblique cuts in the anterior and posterior portions. (Fig. 4) The masseter muscle is not detached from the zygoma in order to avoid postoperative pain and limitation of the mouth opening. The temporal muscle is incised posterior to the preserved temporal artery and it is reflected downward with the free zygomatic arch.

The cranio-orbital flap is made using three guide holes. The first is the key hole, as described above. The second is placed at the temporal squama, just above the zygomatic root, and the third is placed superior to the supraorbital rim, near to the midline. In older patients, additional holes should be done in order to preserve the dura. The osteotomy is made connecting the inferior temporal hole with the frontal hole, and the frontal hole is connected with the key hole using the fine drill or Gigli saw. The inferior temporal hole and the key hole are connected with high-speed drill using foot attachment or rongeurs. The dura should be dissected free, and the bone flap is removed. [2]

In the case of lateral orbital bone involvement by the meningiomas, the cranio-orbital zygomatic osteotomy should remove the frontal process of the zygomatic bone. In such cases, the osteotomy should include the zygomatic arch and the frontal process of the zygoma as a single piece.[13,21] If the orbital roof is also involved the bone removal should include such portion. The optic canal opening and the radical removal of the sphenoid ring around the superior orbital fissure offer a superior control of the disease. [12]

ILLUSTRATIVE CASE 1

V. M. , female, 51 years-old, with previous partial removal of a right sphenoid wing meningioma 5 years before in another institution, submitted to complementary radiotherapy. The patient presented with headache, progressive right exophthalmos and visual disturbance. Preoperative CT scan and MRI evidenced intraosseous involvement of the greater and lesser wings of the sphenoid bone. (Fig. 6)

A radical removal of the bone and adjacent temporal dura was performed through a COZ approach. (Fig. 7) There was no adverse ophthalmological effect after surgery.
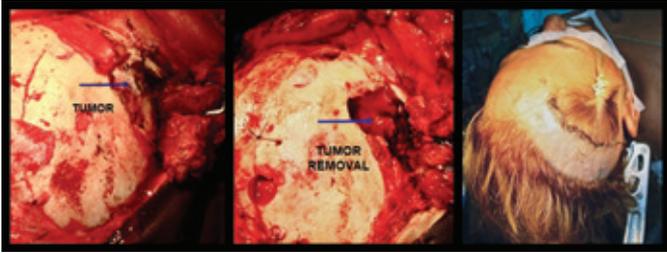
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Fig. 7 - Surgical removal through the cranio-orbito zygomatic approach. Note the extensive bone removal of the superior and lateral aspects of the orbit.

Postoperative 3D CT evidences the resection of the bone, with satisfactory cosmetic results. (Fig.8 and 9)

Fig. 8 - Above: frontal and lateral view of the bone involvement of the greater and lesser sphenoid wings.
Below: Surgical removal of the bone tumor involvement. TU: tumor; SR: surgical removal

Fig. 9 - Left - pre-operative aspect; Right - post operative (3 months follow-up)
Note the resolution of the exophtalmus with inferior dystopia of the right eye.

ORBITAL RECONSTRUCTION

The periorbit preservation is crucial for the ophthalmological cosmetic and functional restoration. According to DeMonte et al. [7], removal of the posterior portion of the roof usually is not associated with clinically significant ophthalmic disturbances. Also, the lateral and medial wall bone removal are well tolerated and produce discrete enophthalmos or dystopia. If the periorbit is preserved, the medial, lateral and roof solid reconstruction may be unnecessary. Otherwise, the orbital floor removal, especially if in more than two-thirds, is associated with higher levels of ophthalmic complications. Studies demonstrated the necessity of orbital floor reconstruction if more than 50% of the bone is removed, in order to avoid enophthalmos and hypoglobus, even if the periorbit is preserved. [7]

COZ approach and other orbitocranial approaches replace the superior and lateral borders of the orbit at the end of the procedures. Nevertheless, the floor and periorbital contents require special techniques and materials for reconstruction. Several alternatives for orbital replacement are described as follows: bone cement, split calvaria, iliac crest, rib grafts, titanium mesh, and other synthetic materials. The techniques include since simple flaps manually fashioned to complex computer-generated models. [15]

ILLUSTRATIVE CASE 2

C. J. A., female, 67 years-old, with multiple previous surgical resections of a right sphenoid wing meningioma, presented with amaurosis, exophthalmos, ocular pain, complete ocular paralysis and headache. A large right frontotemporal bone removal was evidenced with serious cosmetic disturbance. (Fig. 10 and 14).
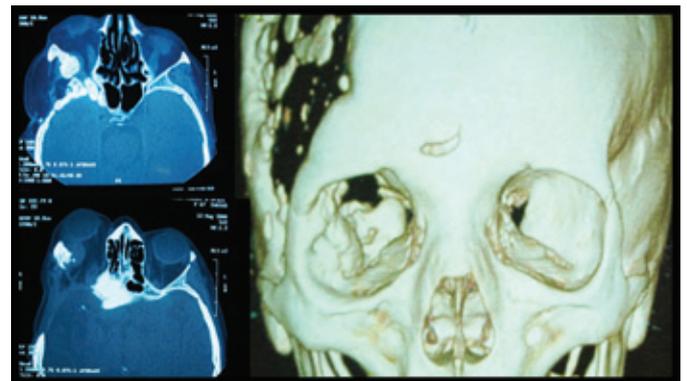

Fig. 10 - Left - CT pre-operative. Note the lateral orbital, temporal bone, the lesser and greater sphenoid wing involvement. Right - 3D CT presenting the large frontotemporal bone defect.

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A radical removal was performed with a COZ approach: complete resection of the superior and lateral orbital borders and the total removal of the orbital roof, extending to the ethmoid and sphenoid bones, removal of the anterior clinoid and opening of the optic canal. (Fig. 11 and 12)
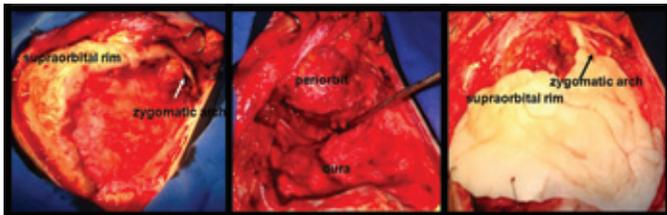

Fig. 11 - **Left -** Preparing COZ approach. Note the lateral involvement of the orbital border. **Central -** Radical removal of the superior and lateral orbital border. The roof, anterior clinoid process and ethmoidal bone were also removed. **Right -** Bone cement reconstruction of the superior and lateral orbital border and the frontotemporal bone defect.
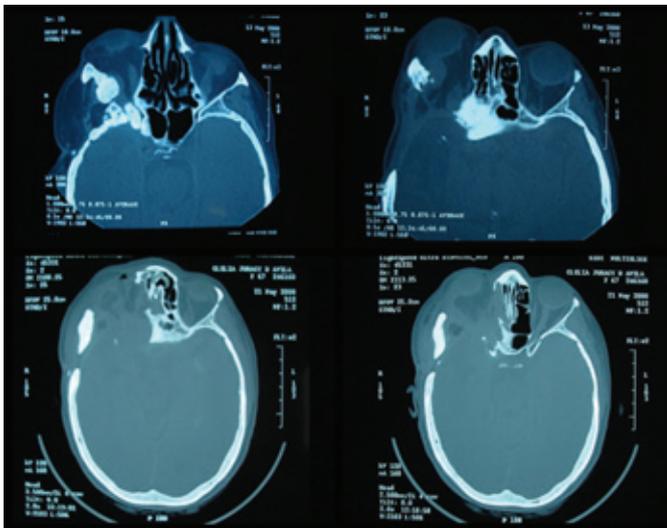

Fig. 12 - Illustrative case 2. **Above -** Right orbital recurrent meningioma. The extensive involvement of the lateral wall of the orbit, greater and lesser sphenoid wings promotes exophtalmos. **Below -** Radical removal of the bone involvement. Note the improvement of the exophtalmos on the postoperative CT.

Orbital reconstruction was performed in a single piece of bone cement, creating the superior and lateral orbital borders and the frontal and temporal bone replacement. (Fig. 11 and 13) There was no improvement of the ocular paralysis and amaurosis. The initial cosmetic result was satisfactory and the patient will be submitted to superior eyelid plastic reconstruction in order to correct the skin expansion caused by previous exophthalmus. (Fig.14)

Fig. 13 - **Left -** 3D CT pre-operative. **Right -** 3D CT postoperative

Fig. 14 - **Left -** Pre-operative cosmetic disturbance; **Central -** Postoperative aspect; **Right -** Postoperative at 1 year follow up.

Recurrence

Meningioma recurrence rates ranges from 10 to 23%. The recurrence of the spheno-orbital meningioma tends to be higher and varies from 14% to 50%. The bone involvement of the cranial base and of the superior orbital fissure or cavernous sinus invasion respond for such higher levels of recurrence. [12,17]
Both cases illustrated above were examples of subtotal removal during their first operations, followed by radiotherapy, with poor disease control.

Conclusion

The orbital meningiomas are formidable neurosurgical challenges. The higher recurrence levels are related to bone involvement and the structures of the cavernous sinus and superior orbital fissure. Radical removal should be the goal at the first procedure. Orbital reconstruction is recommended when the orbital floor is removed. Also, the extensive bone resection of the roof and lateral orbital wall, with periorbital disturbance, are indicative of orbital reconstruction.

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