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Surgical management of bony orbit tumors

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Abstract

Background: A variety of primary tumors affect the bony orbit. They include, sphenoid ridge meningioma, fibrous dysplasia, ossifying fibroma, osteoma and aneurismal bone cyst. Controversies in the treatment strategies still exist. This study is retrospective study that included 20 patients operated upon in the last three years—at neurosurgery department in Minia University Hospital—with tumors related to the bony orbit. We studied the data, to see what was done, what should have been done. CT and MRI were used for all patients preoperatively and for postoperative follow-up.

Results: Fourteen patients had sphenoid ridge meningiomas, two patients had osteomas, two patients had fibrous dysplasia, one had ossifying fibromas and one aneurysmal bone cyst. Proptosis improved in all cases. But one patient had deterioration of vision. There were no postoperative cases of enophthalmos or pulsating exophthalmos.

Conclusions: Early surgical intervention is mandatory before the tumor becomes less amenable. Both CT and MRI should be done preoperatively. Opening the bony superior orbital fissure is a mandatory step in all cases, whereas the optic canal should be drilled out open only if the optic nerve is compromised within the canal. Thorough knowledge of the skull base operative techniques is a must.

Keywords: Orbital tumors, Sphenoid ridge meingeoma, Bony orbit, proptosis

Background

Although tumors of the bony orbit represent a clear neurosurgical entity, they are diverse and not very common. Primary tumors affecting the bony orbit include sphenoid ridge meningioma, fibrous dysplasia, ossifying fibroma, osteoma, aneurysmal bone cysts, and other rare tumors like giant cell osteoclastoma, intra-diploic dermoids, and invasive schwannomas [1].

Due to the large controversy in the treatment strategy that still exist, these tumors are managed differently by every neurosurgeon. This controversy ranges from not to operate at all in some cases, to the other end of the spectrum, that is, the need for early intervention with aggressive bony drilling of the superior orbital fissure, the optic canal, the supraorbital rim, with extensive opening of the relevant dural coverings. As far as we are aware, there were no studies to validate any given management strategy for these tumors [2].

Aim of this work is to assess the visual and cosmetic outcome of patients with orbital bony tumors postoperatively and to address the best surgical management regarding timing and procedure.

Methods

This is a retrospective study—done in Neurosurgery Department at Minia University Hospital—that included 20 patients operated upon in the last three years with tumors related to the bony orbit.

All patient's records were collected, and standardized according to the same inclusion criteria, which included both CT and MRI as preoperative investigations, documented preoperative and serial postoperative ophthalmological evaluation for the degree of proptosis, visual acuity, and ocular motility. The operative data, postoperative radiological investigations, and follow-up notes were collected and categorized according to the pathological

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nature of each group of patients. We studied the data, to see what was done, what should have been done.

We used two approaches, the one-piece "FTOZ" approach. Using MacCarty keyhole and inferior orbital fissure which acts as a naturally existing burr hole connecting the orbit to temporal fossa [3]. However, in this approach, pathological involvement of the greater wing of the sphenoid bone in some patients with en plaque meningiomas prevented "controlled fracture" of the bone flap and led to "uncontrolled fracture" and/or excessive bleeding. So, with extensive involvement of the sphenoid greater wing at the floor of the middle cranial fossa, we used the 2-piece FOTZ approach starting with classic pterional approach followed by orbito-zygomatic osteotomy removing the orbital rim, superior and lateral orbital walls. After removing the bone flap, the pathological bone is drilled out in the elevated bone flap, floor of the middle cranial base, and orbital walls as well. The endpoint of drilling is removing all pathological bone in the elevated FTOZ bone flap, satisfactory decompression of SOF and optic nerve, orbital contents, and restoration of the orbital cavity [4].

Results

The study originally included 22 patients with tumors related to the bony orbit, of these only 20 patients were operated upon. The remaining two patients were not operated upon due to poor expected prognosis in one patient with an aggressive secondary from a prostatic carcinoma, and a second patient who refused surgery. Both cases were excluded from the study.

Out of the 20 patients included, sphenoid ridge meningiomas were the most common pathological entity affecting the bony orbit, with a total number of 14 patients. All patients included in the study above the age of 27 years were all cases with sphenoid ridge meningiomas, or osteomas. Two cases of osteomas were included. Fibrous dysplasia and ossifying fibroma were more common in the second decade with two patients for each. Only one case of aneurysmal bone cyst was included.

In a descending order, the common symptoms were diplopia peri/retro-orbital pain, visual deterioration, eyelid swelling temporal fossa swelling and temporal lobe epilepsy.

There were no mortalities. One patient had deterioration of vision following surgery during which de-roofing of the optic canal was done without preoperative evidence of its involvement. Two patients had CSF leaks that needed a second surgery with the use of fibrin glue. Two patients had a disfiguring scar following an extra-cranial inner canthus incision. One patient had a superficial wound infection which was managed accordingly. One patient had a recurrence that needed a second surgery.

Table 1 Breakdown of the cases included in the study according to the pathological nature of the lesion

Pathology entity	N of cases included
Sphenoid ridge meningioma	14
Fibrous dysplasia	2
Ossifying fibroma	1
Osteoma	2
Aneurysmal bone cyst	1
Total	20

Table 2 Possible complications and their incidence

Complications	N of cases affected
Mortality	0
CSF leak	2
Disfiguring scar	2
Superficial wound infection	1
Recurrence	1
Enophthalmos	0
Pulsating exophthalmos	0

Proptosis improved in all cases. There were no postoperative cases of enophthalmos or pulsating exophthalmos (Tables 1, 2).

Discussion

Tumors of the bony orbit represent a clear neurosurgical entity. These tumors are diverse and not very common. Primary tumors affecting the bony orbit include sphenoid ridge meningioma, fibrous dysplasia, ossifying fibroma, osteoma, aneurysmal bone cysts and other rare tumors like giant cell osteoclastoma, intra-diploic dermoids, and invasive schwannomas.

Sphenoid wing meningiomas

They represent the most common pathological lesion affecting the bony orbit in the adult population. There is a strong female preponderance. Patients classically present with slowly progressive exophthalmos, with or without headache. The meningiomatous carpet could spread both intra-orbital and into the optic canal. In these tumors, it is true invasion of the bone by the tumor cells [2, 4–6]. It may form three types of bony growth: (a) Mere endostosis, (b) extensive hyperostosis (with globular tumor), and (c) en plaque hyperostosis (with only a thin tumor layer).

Fibrous dysplasia

The lesion is formed of a fibrous stroma displaying focal areas of osteoid tissue arrested at the stage of woven

immature bone. Osteoblasts are absent. Children and young adults are most affected. They present with headache, facial asymmetry, and very slowly progressive painless proptosis. Radiologically, it may mimic hyperostosis with sphenoid wing meningiomas (which very rarely occur in the pediatric age group). "Ground-glass" thickening of the adjacent frontal bone and sphenoid bone is characteristic on plain X-ray films. This lesion has a high percentage of recurrence [1, 2, 5, 6].

Ossifying fibroma

This tumor has created considerable confusion and controversy. It seems to be a distinctive lesion restricted to the facial and skull bones. The lesion is formed of a fibrous stroma displaying focal areas of osteoid that matured into lamellar bone. Osteoblasts are present. Ossifying fibroma is an osseous tumor encased in a sclerotic shell. It usually presents at the age of puberty. It is rapidly progressive, and more common in females. Typically, the lesion does not respect the supraorbital ridge [1, 7–9].

Orbital osteoma

It is a slowly growing lesion, more common near the ethmoid sinus, or the superior lateral part of the orbit. The supraorbital ridge is mostly spared. There is a slight female preponderance. Usually presents with proptosis without affecting the ocular motility [1].

Aneurysmal bone cyst

Patients present by rapidly progressive proptosis and a bony lytic lesion distinguish it radiologically. It usually affects children and young adults [1].

The diagnostic and treatment dilemma: Many neurosurgeons, even within the same department or institution, would differ in their decisions on several cardinal aspects in the management of these tumors. Although the goals are very clear, the strategy to reach such goals, regarding tumors of the bony orbit, differ greatly. The goals and objectives from the neurosurgical standpoint are: l—excision of the pathological process, 2—restore or improve function, and 3—maintain or improve the cosmetic aspect [10].

But several issues remain a matter of vast controversy regarding these tumors. We analyzed the procedures, results, and outcome in the 20 cases and came out with the following controversial issues, and conclusions.

Is surgery needed or not? Many neurosurgeons would agree that surgery is indicated in cases of sphenoid wing meningiomas and aneurismal bone cysts, but the controversy on whether to operate or not becomes very clear when dealing with ossifying fibroma, fibrous dysplasia, and osteomas of the orbit. These neurosurgeons argue that such lesions are slowly growing, could not be excised totally and have high tendency to recur after surgery. Authorities in orbital surgery clearly state the importance of early surgery [3, 7, 8, 11, 12]. Going back to these series of cases, it was clear that all the five patients with the above three pathologies had conflicting opinions from many surgeons before finally being admitted for surgery. These patients were all operated upon, and the proptosis improved in all cases. Recurrence took place in the one case of ossifying fibroma due to what was believed to be inadequate excision.

When to proceed with surgery? The timing when to proceed with surgery is also controversial, some neurosurgeons would argue about it claiming that due to the slow growth rate for most of these lesions. They will not see surgery needed except when the patient starts complaining of visual deterioration or persistent headache. Their argument will target specifically the en plaque meningioma with hyperostosis". We detected 3 cases where surgery was delayed on this assumption. With the result that the tumor became not amenable to total excision, thus surgery is transferred from a curable treatment modality into a palliative one [2].

Should one proceed with a trans-cranial or an extracranial approach? Some maxillofacial surgeons and few skulls base surgeons would advocate the extra-cranial approach as the better avenue to excise such lesions [8]. They advise using a curvilinear inner canthus incision. In this study two patients only were operated upon using this approach, and the inner canthus skin incision was totally managed by plastic surgeons. Both patients had the lesion excised successfully, but had a disfigured pigmented scar, and were not satisfied about the cosmetic outcome of the surgery, they felt the proptosis was exchanged for the disfigured scar.

In case of a trans-cranial procedure, how much of the orbital roof is drilled out? And should the supraorbital rim be drilled? Revising the preoperative and postoperative radiological investigations of the patients in this study, we noticed that the amount of orbital involved was mainly dependent upon the pathological nature of the lesion in case of fibrous dysplasia and sphenoid wing meningiomas, only the lateral two thirds were affected and needed to be drilled out. The entire orbital roof should be drilled out in ossifying fibromas, and only the affected part in osteomas and aneurysmal bone cysts. On the other hand, removing less than the lateral two thirds, in case of en plaque meningiomas or fibrous dysplasia, will not allow adequate regression of the proptosis [1, 2, 4, 5, 8, 9]. The supraorbital rim should always be preserved except in cases of ossifying

fibroma because in these cases it is involved in the relatively rapidly progressive hyperostotic process [1, 11].

When is opening the SOF necessary? The debate on whether opening the SOF is mandatory or not was confused by the fact that this crucial anatomical part has both a bony component and a dural component. A plethora of reports in the literature concluded that drilling the bony SOF should be done in all cases where the tumor affects the roof or the medial orbital wall; otherwise, proptosis will not improve [1–3, 9–11]. This is one of the main causes of failure to treat the proptosis occurring in these patients. This took place in one case in this series. There is no value in opening the dural covering of the SOF, except with a proved MRI evidence of a tumor carpet inside; otherwise, the risk of such a maneuver will be much higher than the assumed benefits [5, 6].

When is opening the optic canal necessary? Opening of the bony optic canal is not a maneuver without risk. Inadvertent injury to the optic nerve is well reported and could be a direct injury by the high-speed drill or indirectly due to the heat effect generated by the drilling [2, 7, 10, 13, 14].

In this series, one patient had deterioration of vision following surgery during which de-roofing of the optic canal was done without preoperative evidence of its involvement. We did not find significant evidence in the literature to support the notion that visual loss associated with fibrous dysplasia is the result of progressive optic canal stenosis, thus raising questions about the value of prophylactic optic canal decompression [6, 7, 9, 10]. Therefore, optic canal de-roofing should not be done except when the preoperative investigations clearly demonstrate its involvement as the cause for the visual deterioration.

When is zygomatic osteotomy needed? Zygomatic osteotomy should only be done if the bony orbital tumor extends to the floor of the bony orbit or to the orbital apex [10]. Failure to stick to this operative rule means that repair and reconstruction will be more difficult, and sometimes miniplates and screws would be necessary. On the other hand, if the orbital apex or floor is involved, it would be impossible to gain adequate and safe access without the zygomatic osteotomy.

Should the periorbita be repaired? The periorbita is a tough structure, especially its lower parts. Due to the configuration of the contents within the orbit. and the effect of gravity. herniation of its contents would take place only if its inferior portion is affected [1, 2, 10, 13]. Violation of the upper part of the periorbita could take place during the bony drilling or by the lesion itself. In this series, no repair of the upper part of the periorbita was done in all but three cases without any difference in the outcome. It should be repaired only if it was violated in its inferior parts [1, 9].

Who should be operated for orbital reconstruction? Which type of reconstruction is better? Most authors state that long term cosmetic results with bone cement reconstruction alone are not very impressive [1, 8, 9, 13]. They advise using bony autografts for better results. In our experience, we found that titanium plates, mesh with bone cement on top give the best results, but this again needs a longer follow-up period for proper credibility. And lastly, do these patients really improve? Looking carefully at the outcome of these 20 patients, one can confidently conclude that they do improve if early and aggressive surgery is performed. The first surgery is very crucial for the outcome.

Conclusions

Tumors of the bony orbit are diverse pathologically. Early surgical intervention is mandatory before the tumor becomes less amenable. Both CT and MRI should be done preoperatively. The first surgery is very crucial for the outcome. We did not find significant evidence in the literature, or in this series to support the notion that visual loss associated with fibrous dysplasia is the result of progressive optic canal stenosis, thus raising the question about the value of prophylactic optic canal decompression. Patients do improve if early and aggressive surgery is performed. Enophthalmos and pulsating exophthalmos are very rare to occur if the fore mentioned strategies are followed. The take home message is that early diagnosis and treatment of orbital bony tumors with good decompression of the optic canal and SOF results in great improvement of cosmetic and visual manifestations.

Abbreviations

CT: Computed tomography; MRI: Magnetic resonance imaging; SOF: Superior orbital fissure; CSF: Cerebrospinal fluid.

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Author contributions

All authors contributed and participated in all parts of the research and manuscript. All authors read and approved the final manuscript.

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Availability of data and materials

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Declarations

Ethics approval and consent to participate

An informed written consent was taken from each patient prior to the operation. This consent was done according to the guidelines of Faculty of Medicine

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Consent for publication
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Competing interests
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