

Tumors of the Orbit: Case Report and Review of Surgical Corridors and Current Options

Abstract

Tumors that involve the orbit can be classified into two major groups: primary tumors of the orbit and tumors that extend into the orbit from other sites. The most frequent primary orbital lesions in adults include cavernous hemangiomas, lymphoid tumors, and meningiomas. The most common tumors that extend into the orbit are meningiomas, followed by sinonasal carcinomas. In this article, we report a case of intraconal orbital lesion operated at our center and a review of the surgical approaches to the orbit.

Keywords: *Cavernous angioma, endoscopic approach to orbit, intraconal/extraconal orbital tumors, orbital anatomy, orbital tumors, surgical approach to the orbit*

Introduction

Orbital tumors are generally divided into three categories based on their location within the orbit: (1) intraconal (within the extra-ocular muscle cone), (2) extraconal, and (3) intracanalicular (within the optic canal), with differing features based on these locations.^[1]

Tumors that involve the orbit can also be classified into two major groups: primary tumors of the orbit and tumors that extend into the orbit from other sites. The most common primary orbital tumors in adults include cavernous hemangiomas, lymphoid tumors, and meningiomas whereas dermoid cysts, capillary hemangiomas, and rhabdomyosarcoma predominate in children.^[2]

Modern diagnostic methods help to determine their location, extent, and position relative to the blood vessels, nerves, and muscles in this region. Direct approach to various parts of the orbit is now possible with the help of modern equipment and the development of surgical techniques. In this article, we report a case of intraconal orbital lesion operated at our center and a review of the surgical approaches to the orbit. The representative case is a difficult and challenging case.

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Case Report

A 51-year-old male presented to our outpatient clinic with a history of double vision. No history of headache, vomiting, any other cranial nerve involvement, no motor weakness, or sensory disturbances was reported. Clinical examination was grossly normal; visual acuity, visual fields, and fundus were within normal limits. Plain computerized tomography (CT) of the brain revealed well-defined regular lobulated hyperdense lesion in the medial aspect of the left orbit; on magnetic resonance imaging (MRI), lesion was found to be hypointense on T1 and hyperintense on T2 within the extraocular muscle cone mildly displacing the optic nerve, with homogenous contrast enhancement on gadolinium as shown in Figure 1. With a provisional diagnosis of cavernous angioma, the patient was taken up for surgery. In this case, the transcranial approach was chosen because of our familiarity with the approach and previous experiences with the same. Alternative options are endoscopic endonasal approach and supraorbital keyhole approach.

The patient was positioned in the supine position, with head fixed in Mayfield head holder in slight flexion and turned to the right side. Using a bicoronal flap, a left frontal craniotomy was performed,

and then, using an extradural approach, the floor of the anterior cranial fossa

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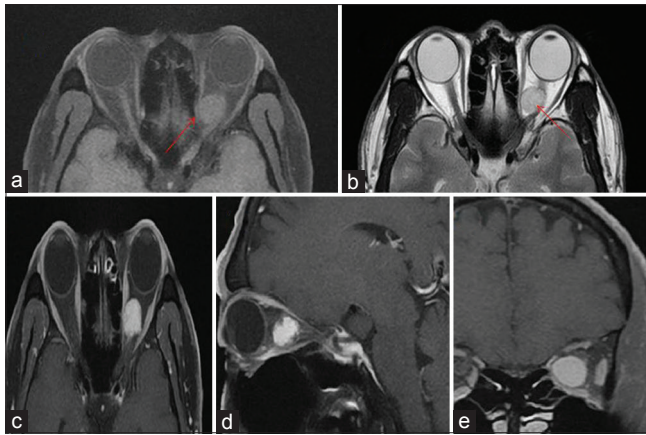


Figure 1: (a) T1-weighted showing hypointense lesion in the left orbit, (b) T2-weighted showing hyperintense lesion in the left orbit. (c-e) Postgadolinium contrast sagittal, coronal, and axial images showing homogeneous contrast-enhancing lesion in the left orbit. The characteristic popcorn calcification is not seen

was exposed by retracting the basifrontal duramater. The supraorbital margin was drilled flush with floor of the anterior cranial fossa, then followed by extradural drilling of the orbital roof entering into the orbit exposing the periorbital pad of fat. Under high magnification on dissecting the periorbita and lateralizing the rectus muscle, the lesion was visible. It was firm in consistency with well-defined margins [Figure 2a]. The lesion was attached to the inferior division of the oculomotor nerve; hence, near-total resection was performed. Orbital roof reconstruction was done with titanium mesh and screws. Gross specimen was firm, reddish white lesion [Figure 2b]. Postoperative MRI showed no gross residual tumor [Figure 2d]. In the immediate postoperative period, the patient had temporary blepharoptosis, improving with medications. Figure 2c showing cut section. Histopathological examination was suggestive of cavernous angioma [Figure 2e].

Discussion

Relevant anatomy

The orbits are paired structures; each orbit is a bony pyramid with four walls: a roof, lateral wall, floor, and medial wall with a posterior apex, anterior base, and a medially tilted axis with an approximate volume of 30 cm.^[3] The base is quadrangular, with its widest dimension just posterior to the orbital rim. The apex is formed by the optic canal and superior orbital fissure with its contents as described below, namely the superior and inferior divisions of the oculomotor nerve, the trochlear nerve, the ophthalmic branch of trigeminal, the abducent nerve, the superior ophthalmic vein, and sympathetic fibers from the cavernous sinus.

The orbit is made up of seven bones, namely – frontal, ethmoid, lacrimal, sphenoid, zygomatic, palatine, and maxilla.^[4] There are six true extraocular muscles responsible for movements of the globe. It comprises of four rectus

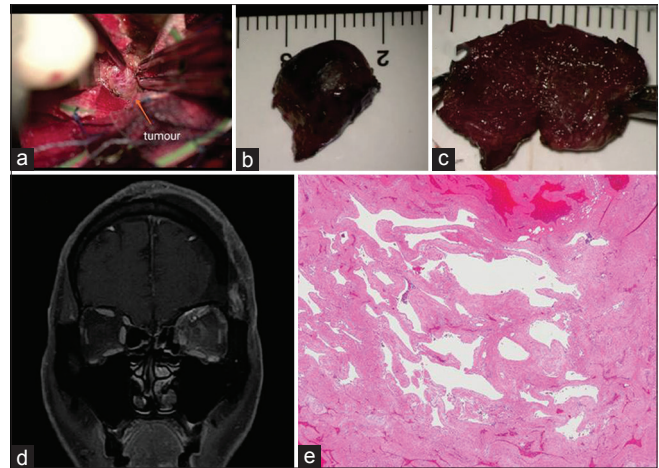


Figure 2: (a) Tumor dissection, (b) gross specimen, (c) cut section, (d) postoperative magnetic resonance imaging showing no gross residual lesion, (e) microphotograph showing vascular channels

muscles, which arise from the tendinous ring - annulus of Zinn at the apex of the orbit and insert into the sclera about 4–8 mm behind the limbus and two oblique muscles (superior and inferior) whose tendons approach the globe from the front and insert into the posterior aspect of the sclera. In addition, there is the levator palpebrae superioris, which originates at the orbital apex and inserts into the tarsal plate and upper eyelid.^[4,5]

Structures passing through the annulus are the optic nerve and ophthalmic artery, the oculomotor and abducens nerves, and the ophthalmic division of the trigeminal nerve. The trochlear nerve and the frontal and lacrimal divisions of the trigeminal nerve course outside the annulus.

The ciliary ganglion is situated on the inferolateral aspect of the optic nerve and on the medial side of the lateral rectus muscle. It receives three branches: the motor (parasympathetic) root from the inferior division of the oculomotor nerve, the sensory root from the nasociliary nerve, and sympathetic fibers from the plexus around the internal carotid artery (ICA). The sympathetic fibers arise in the cervical sympathetic ganglia and pass through the ciliary ganglion without synapsing. The short ciliary nerves pass from the ciliary ganglion to the globe. Sympathetic fibers to the globe and pupillary dilator are carried by the long ciliary nerves.

The superior ophthalmic vein drains the eyeball, the superior portion of the orbit, the ethmoid sinuses, and the forehead into the cavernous sinus. Other veins that drain the orbit are the supratrochlear, supraorbital, angular, and even the maxillary and retromandibular veins.

The ophthalmic artery is the major supply to the orbit; it is a branch of the supraclinoid portion of the ICA. The origin of the ophthalmic artery is usually medial to the anterior clinoid process, below the optic nerve. This vessel follows the optic nerve in the optic canal and orbit. At the optic canal, the artery has already passed lateral to nerve;

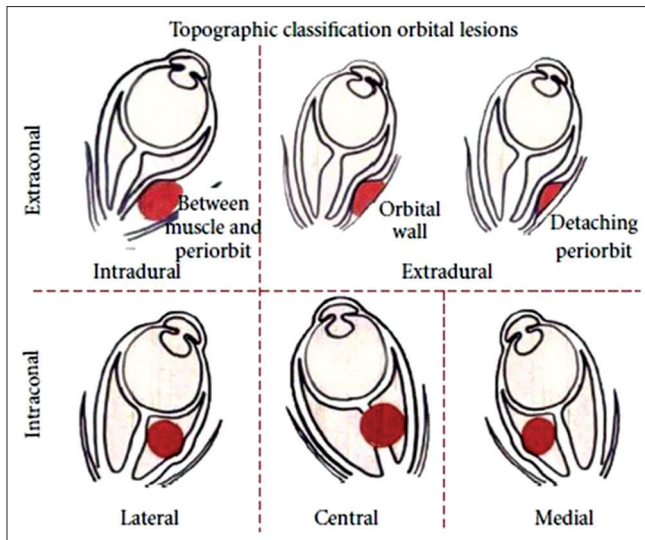


Figure 3: Topographical classification of orbital lesions from Martins *et al.*^[4] reprinted with permission

knowledge of this anatomy is essential during opening of the falciform ligament after removal of the anterior clinoid, to prevent iatrogenic lesion to this artery. Once inside the orbit, the ophthalmic artery passes in most cases, above the optic nerve. At this crossing point, it usually gives rise to the central retinal artery. Working along the lateral orbital space, it is vital to avoid blind coagulation once inadvertent coagulation of the retinal artery results in loss of vision.^[4]

Figure 3 topographical classification of orbital lesions from Martins *et al.*^[4] The presence of the periorbita allows classifying the orbital lesions into intradural (subdural) – when deep to the periorbita and extradural – when located between the periorbita and the bony orbit. The muscular cone divides the orbit into intraconal and extraconal spaces. While the intraconal lesions are always intradural (subdural), the extraconal lesion can be either intra- or extra-dural. The intraconal space also can be further subdivided in relation to the optic nerve into medial, central, and lateral orbital spaces. This understanding is paramount in classifying the orbital lesions and in adequately choosing a surgical path for its treatment.^[4]

Orbital tumors may arise primarily in the orbit or may invade the orbit from surrounding structures such as eyelids, sinuses, or the upper jaw.^[6] The most frequent primary orbital tumors in adults include cavernous hemangiomas, lymphoid tumors, and meningiomas whereas dermoid cysts, capillary hemangiomas, and rhabdomyosarcoma predominate in children.^[2]

The most common tumors that extend into the orbit are meningiomas, followed by sinonasal carcinomas.^[2,3]

The most common differential diagnosis as summarized by our senior author – Hasegawa^[7] is shown in Figure 4.

The most frequent initial symptom of an orbital mass is proptosis, followed by diplopia. Change in visual acuity is rare, may occur as a late finding, and indicates a tumor that is close to the orbital apex or optic nerve.^[2,3]

Intraconal tumors tend to cause early vision loss, impairment of ocular motility (manifesting as double vision), and axial proptosis. These effects result from direct compression of the optic nerve and impingement on extraocular muscles. Extraconal tumors cause proptosis as an early manifestation, and displacement of the globe and compression of the extraocular muscles can produce diplopia, sometimes only in certain fields of gaze. Intraconal tumors tend to cause early vision loss, impairment of ocular motility (manifesting as double vision), and axial proptosis. These effects result from direct compression of the optic nerve and impingement on extraocular muscles. Extraconal tumors cause proptosis as an early manifestation, and displacement of the globe and compression of the extraocular muscles can produce diplopia, sometimes only in certain fields of gaze.

Imaging of the orbit

MRI plain and contrast provides detailed information about the anatomical relationship among the extraocular muscles, the optic nerve, the globe, and the lacrimal gland as well as an idea of the state of tumor borders and potential encapsulation.

CT is used to assess the relationship of the lesion to the surrounding bones. MRI dynamic study shows characteristic nodular enhancement in cavernous angiomas as summarized by Hasegawa.^[7] Hasegawa *et al.* also summarized that CT arteriography with selective arterial catheterization clearly delineated the feeding artery territories and provides additional valuable preoperative information about the orbital tumor under investigation.^[8]

Approaches to the orbit

Paluzzi *et al.* suggested that the orbit can be viewed as a clock, with approach varying based on tumor position.^[9]

The principle when choosing a surgical corridor is to avoid working across or around nerves. Specifically, one should avoid crossing the plane of the optic nerve. Therefore, orbital pathology lateral to the optic nerve is accessed through lateral orbitotomies, and medial pathology is accessed through medial orbitotomies.

Traditionally, external approaches to the orbit provide excellent access to tumors that are superior and lateral to the optic nerve and orbit, usually are best accessed by a pterional or fronto-orbital temporal craniotomy with or without orbitozygomatic osteotomies.^[1,9,10] Tumors with lateral intracranial extension can also be approached by this approach. Another variant is the lateral orbitotomy, which provides excellent access for orbital tumors lateral to the optic nerve and apex.^[9,10]

For tumors located anteriorly in the orbit, an anteromedial micro-orbitotomy or transconjunctival approach is the

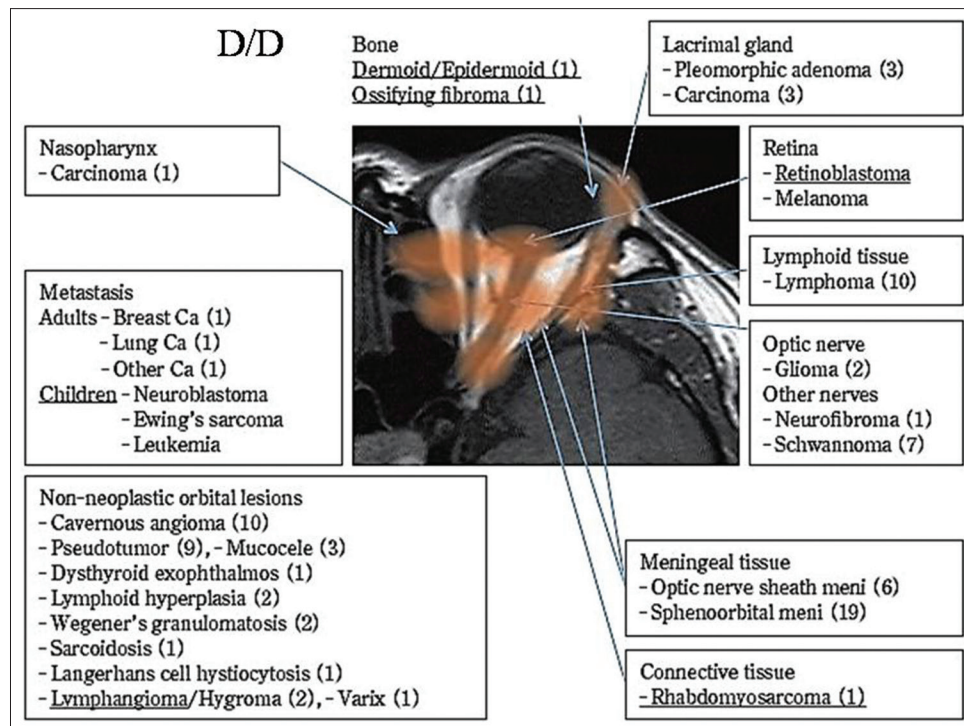


Figure 4: The most common differential diagnosis as summarized by our senior author – Hasegawa^[7]

traditional approach for resection. Endoscopic assistance through standard external approaches was used to improve visualization as early as the 1980s.

Extended endonasal approaches (EEAs) provide excellent access for intraconal and extraconal tumors that are medial and inferior to the optic nerve and can be applied to any medial intracranial extension, provided that key neurovascular structures (e.g., the optic nerve and ICA) remain lateral to the tumor. EEAs also provide access to most of the orbit, from the posterior globe to the orbital apex.^[9,11]

The main advantage of any EEA is its anterior and medial trajectory, which is most suitable for skull base lesions that are anteromedial to critical neurovascular structures. The key anatomic landmark is the optic nerve. Tumors that displace the optic nerve superiorly and laterally are usually excellent targets for an endonasal approach. There are three main endonasal approaches to the orbit: (1) the medial–inferior extraconal approach, (2) the transmaxillary extraconal approach, and (3) the medial intraconal approach.

Lateral Approaches

Pterional (frontotemporal) craniotomy with or without orbitozygomatic extension

The pterional approach was first described by Naffziger in 1948 with modifications proposed by Hamby in 1964 and Yasargil *et al.* in 1987.^[12-14] A curvilinear incision starting just anterior to the tragus up to the midline apex

of the anterior hairline (so-called widow's peak) is usually sufficient to access the superolateral orbit. A soft tissue flap (scalp and temporalis muscle) is developed elevating the fat pad between the superficial and deep layers of the temporalis fascia to protect the frontalis branches of the facial nerve. A subperiosteal dissection should be carried onto the orbit and around its rim, dissecting the periorbita from the inner orbit. The supraorbital neurovascular bundle is either dissected from its notch or freed from its foramen with diagonal osteotomies (inverted V), directed away from the nerve. Next, a standard frontotemporal craniotomy is performed. The lateral bone of the greater wing of the sphenoid is then dissected free from the dura and removed with rongeurs and a high-speed drill until it is flushed with the orbit. Bone removal should extend posteriorly only as far as the orbital meningeal artery to prevent inadvertent injury to the contents of the superior orbital fissure. The orbital meningeal artery is an important landmark marking the "tip of the iceberg," with the superior orbital fissure lying beneath. At this point, the frontal dura can be dissected free from the roof of the orbit so that the orbital bone is freed from dura on one side and periorbita on the other before performing the orbital osteotomies. A reciprocating saw can be used for the orbital osteotomies while protecting the brain and orbit with malleable retractors. The saw is inserted into the orbit, and the cut is made from the orbit toward the frontal dura. The medial cut is usually at or just lateral to the supraorbital notch and should align with the medial edge of the craniotomy. The lateral cut is made by inserting the tip of the reciprocating saw into the inferior orbital fissure and completing an osteotomy from within the orbit at a level just

above or through the zygomatic prominence, as needed. The final posterior osteotomy is completed with a small drill bit from the cranial side while protecting the orbit with a ribbon. This cut is made from the posterior aspect of the medial osteotomy across the roof of the orbit, through the remaining sphenoid wing, and connected laterally to the lateral osteotomy in the inferior orbital fissure. These osteotomies should be extended as posterior as possible to prevent loss of orbital bone, which would require reconstruction to prevent enophthalmos. It is important that the posterior portion of the superior orbit is removed adequately because this bone can prevent adequate dural retraction and defeat any advantage of the superior orbitotomy. Depending on the location of the tumor within the optic canal, the roof of the canal may need to be thinned and then removed with a dissector or small curette. Continuous irrigation should be used to avoid thermal injury to the nerve from the diamond drill bit. Intraorbital tumor can be identified by digital palpation, image guidance, or intraoperative ultrasonography. The periorbita is opened in an anterior-to-posterior direction, and dissection is performed through the periorbital fat. If tumor is located in the medial or superior intraconal space or is affecting the optic nerve, the annulus of Zinn should be opened medial to the levator and superior rectus muscles to prevent injury to the oculomotor nerve. These muscles can be retracted laterally for improved exposure.^[9,12] Standard microsurgical techniques are used for tumor dissection and removal. Orbital reconstruction is performed using miniplates, and the orbital contents must be checked meticulously for entrapment of orbital contents during and after plating and in the postoperative period.

Supraorbital Keyhole Approach

The supraorbital keyhole craniotomy is a popular approach used by neurosurgeons. In the orbit, it can be used for a variety of intraconal and extraconal pathologies located superior to the optic nerve. In this technique, the incision is placed along the eyebrow, lateral to the supraorbital ridge to avoid injury to the supraorbital nerve. U-shaped pericranial flap is made from superior temporal line arching over to the supraorbital bundle, taking care to preserve this blood supply to the flap. A small amount of temporalis muscle can be reflected laterally and posteriorly from the lateral orbit to provide access for a McCarty burr hole. A craniotome is used to make cuts, flush with the orbital roof and arching as superiorly as the incision will permit.^[12,15,16] It is critical to drill the posterior aspect of the remaining orbital rim flush with the roof of the orbit to maximize access. Alternatively, orbital osteotomies can be performed as described earlier. Standard microsurgical techniques are used for tumor dissection and removal.

Lateral Orbitotomy

The skin incision is 3–4 cm long, and it extends from the lateral canthal angle and curves posteriorly in a line in the temple that could be covered by eyeglasses. The incision

should be limited to 2 cm lateral to the bony lateral canthus to avoid damage to the frontal branch of the facial nerve. The anterior margin of the temporalis muscle is dissected from the underlying bone, exposing the lateral wall of the orbit.^[9] A subperiosteal dissection of the orbital side of the bone allows retraction of orbital contents including the lacrimal gland. Two transverse osteotomies are made in the lateral orbital bone using a reciprocating saw, the first superior to the zygomaticofrontal suture and the second just above the origin of the zygomatic arch. This lateral orbital bone can be drilled, cracked with an osteotome, or fractured posteriorly by grasping the rim with a rongeur. The exposed sphenoid wing is removed with a high-speed drill and rongeurs as needed to reach the level of the orbital apex.^[9] The periorbita is opened parallel to the lateral rectus muscle depending on tumor location within the muscle cone. The lateral rectus muscle retracted with either a vessel loop or traction suture, helping to improve the lateral access into the orbit at the same time, previously the Weitlander–DeMartel retractor, which is designed for the lateral orbitotomy, was inserted to aid in retraction.^[17] If there is optic nerve involvement by the tumor, the optic nerve should be identified proximal and distal to the tumor. As with all approaches, standard microsurgical techniques are used for tumor debulking and resection.

The major transcranial–lateral approaches to the orbit with advantages and disadvantages are summarized [Table 1].^[12]

Medial Approaches

Anterior medial orbitotomy and transconjunctival approach

The approach was originally described by Galbraith and Sullivan using a three-pronged tracheal dilator to expose the medial orbit and has been modified to include better retraction and instrumentation with the use of the operating microscope.^[17] The procedure is done under general anesthesia and begins with an eye speculum being placed to retract the lids. A conjunctival peritomy is performed 90° around the cornea. The medial rectus muscle is isolated with a double-armed suture at its insertion site into the globe after relaxing conjunctival incisions are made superior and inferior to the muscle. The muscle is then severed from the insertion site, allowing the intermuscular septa and check ligament to provide retraction of the orbital fat. The lid retractor is removed, and a medial orbital self-retaining (Drucek-Mueller-DeMartel retractor) retractor is inserted. The retractor has an enucleation spoon that is placed medially on the globe for medial-to-lateral retraction.^[9,17] Cottonoids or cotton-tipped applicators are used for retraction of orbital fat. Performing a lateral orbitotomy can provide even more room for lateral globe displacement while avoiding compression of the orbital contents. As with all approaches, standard microsurgical techniques are used for tumor debulking and resection. The muscle is reattached to the original insertion site, and

Table 1: The major transcranial–lateral approaches to the orbit with advantages and disadvantages

Lateral approaches	Indication	Advantages	Disadvantages
Pterional	Tumors near superior and inferior orbital fissures and cavernous sinus; orbital decompression	Minimal brain retraction necessary in purely extradural surgeries	Detailed anatomic knowledge essential, smaller surgical field
Supraorbital	Intraconal and extraconal tumors superior to the optic nerve	Minimally invasive extradural approach with minimal manipulation of the brain and orbital structures, no limitation by tumor size, excellent cosmetic outcomes	Hypoesthesia secondary to supraorbital nerve damage, limited exposure, learning curve, limited exposure for large sphenoid wing meningiomas
Fronto-orbito-zygomatic	Tumors of orbital apex, optic canal, superior orbital fissure; tumors located dorsal to the optic nerve; lateral extraconal, and intraconal tumors	Broad exposure	Bicoronal flap. Scar exposure in patients with receding hairline

the conjunctiva is purse-stringed around the cornea with absorbable 6.0 suture material.

Endonasal Approaches to the Orbit

There are three main endonasal approaches to the orbit: (1) the medial–inferior extraconal approach, (2) the transmaxillary extraconal approach, and (3) the medial intraconal approach.^[3,9,11]

Medial–Inferior Extraconal Approach

All endonasal approaches to the orbit have similar initial phases. After undergoing endotracheal intubation and induction of general anesthesia, the patient is placed in a three-point head fixation system in a neutral or slightly extended position with the head turned 5°–10° toward the surgeons. Image guidance is used in all cases. A 0° endoscope is used for most of the operation; however, a 45° endoscope often becomes necessary for working near the orbital apex, particularly inside the muscle cone, or anteriorly in the orbit. The initial phase includes a complete uncinectomy, wide maxillary antrotomy, anterior and posterior ethmoidectomies, and sphenoidotomy to expose the medial and inferior orbital walls. The sphenoid sinus is opened from the natural overall survival and extended medially, laterally, and superiorly, as needed, for visualization of the bony landmarks overlying the ICA and optic nerve, the planum, and sella. Next, the lamina papyracea is removed in an anterior-to-posterior direction. At this point, the periorbita, optic nerve, and ICA are well defined. A single nostril approach with preservation of the middle turbinate can often be used, particularly in the case of small extraconal tumors in the medial orbit. A binostril approach is used, when dissection of the orbital apex or intraconal work is required. During a binostril approach, the middle turbinate ipsilateral to the pathology is removed, the posterior nasal septum is resected, and the sphenoid sinus rostrum is opened widely, which transforms the posterior sinonasal cavities into one single space for ease of dissection. It should be noted that if a bi-nostril approach is planned, a naso-septal flap is needed for reconstruction.^[3,9]

This approach has the most established record for an endonasal approach to orbital pathology; this approach is commonly used for sinonasal carcinomas that involve the orbit, meningiomas (both intradural and extradural), and juvenile nasal angiofibromas.

Transmaxillary extraconal approach

The initial phase is similar as mentioned above; in addition, a medial maxillectomy is extended anteriorly to give access to the floor of the orbit, inferior rectus muscle, and the orbital contents above. This approach is generally used for tumors that involve the middle fossa or Meckel's cave and extend to compress the cone, such as meningiomas, juvenile nasal angiofibromas, or schwannomas that have extended into the infratemporal fossa, pterygopalatine fossa, or maxillary sinus. The transmaxillary extraconal approach is one of the most advanced endonasal approaches, and such approaches should be reserved for experienced surgical teams.^[3,9]

Medial intraconal approach

This approach is used for medial–inferior and posterior intraconal lesions, including nerve sheath meningiomas, extraocular nerve schwannomas, hemangiomas, and metastases. This approach overcomes the limitation of posterior intraconal access associated with the anterior medial micro-orbitotomy approach. The dissection corridor is between the medial and inferior rectus muscles. The muscles are identified and isolated with vessel loop as they insert on the globe and then retracted. The periorbita is opened parallel to the medial rectus muscle. Once the periorbita is incised, the surgeon has access to the orbital contents. A cotton-tipped applicator can serve as an excellent retractor of orbital fat endonasally as well. The difficulty when working in the intraconal space is keeping the dissection corridor open because the orbital contents are all freely mobile and compressible. If required working in conjunction with an oculoplastic surgeon who can access and retract the rectus muscles through a transconjunctival approach can be done. This aids in the endoscopic identification of the muscles and

provide some anterior muscle retraction. Other variations for controlling the rectus muscles include detaching the medial rectus muscle from the globe, tagging it with a long silk suture, and passing the suture from within the orbit into the nasal cavity.^[9,18] Once the intraconal corridor is developed, standard microsurgical techniques are used bearing in mind that the optic nerve should be lateral and superior to the lesion, and the ophthalmic and central retinal arteries course medial to the optic nerve at the orbital apex. Then, the tumor is identified and removed with limited bipolar cautery and extensive sharp dissection.^[3,9,11]

Combined Approaches

Combined approaches have been proposed by various authors.

Koutourousiou *et al.* described a combined endoscopic endonasal transorbital approach with transconjunctival-medial orbitotomy for excisional biopsy of the optic nerve.^[19]

Campbell *et al.* described a combined transcaruncular and a transnasal endoscopic cryo-assisted approach to remove a cavernous hemangioma.^[20] Zhou *et al.* described Navigation-Guided Endoscopy Combined with Deep Lateral Orbitotomy for Removal of Small Tumors at the Lateral Orbital Apex.^[21]

Conclusion

Orbital tumors are a relatively rare and challenging group of tumors. Thorough knowledge of the anatomy of the extraocular muscle cone, key neurovascular structures is of paramount importance and helps to classify an orbital lesion and choose an ideal approach. Tumor location rather than tumor type determines the type of approach selected. Full 360° access to the orbit requires knowledge, expertise, and experience with a variety of open and endoscopic approaches. The same general neurosurgical principles and techniques can and should be applied throughout. In this case, the transcranial approach was chosen because of our familiarity with the approach and previous experiences with the same. Alternative options are endoscopic endonasal approach and supraorbital keyhole approach. A wide spectrum of reconstructive procedures helps to improve the functional and esthetic outcomes for patients.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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