Endoscopic Endonasal Approach for Intra- and Extraconal Orbital Pathologies

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Abstract

Keywords
► Endoscopic Endonasal Approach
► endoscopy
► orbit
► orbital
► cavernous hemangioma

Endoscopic endonasal approaches offer an important alternative in the management of posterior inferomedial orbital pathology. Beginning with endoscopic orbital decompressions for Graves' disease, the endonasal corridor for the management of intra- and extraconal pathologies has continued to evolve. Endonasal removal of orbital cavernous hemangiomas is well described in the literature; however, many other benign and malignant pathologies of the medial orbit can be accessed through this approach. Advantages of the endonasal approach include improved visualization and decreased manipulation of orbital contents in the medial and posterior orbit. Additionally, for tumors that extend from the paranasal sinuses into the orbit, this corridor may be ideal for concurrent management. The current literature for this approach will be reviewed including the oncologic results, complications, limitations, and reconstructive needs along with pertinent anatomy. In addition, data from our own institution will be reviewed.

Introduction

The orbit remains a complex anatomical area and intraorbital pathology benefits from the input and expertise of multiple specialists including ophthalmology, otolaryngology, and neurosurgery. External approaches including lateral and medial orbitotomies as well as the orbitozygomatic craniotomy are the mainstay for treatment, especially for intraconal tumors. With the proliferation of endoscopic skull base surgery, endoscopic endonasal approaches (EEAs) to orbital pathology have become increasingly utilized.[1–8] Posterior inferomedial orbital lesions are best suited for this approach. In the setting of favorably located pathology, the EEA offers a more direct approach with increased visualization and reduced morbidity when compared with open approaches.[9,10]

Pearls and Tips

- The optic nerve is generally considered the lateral limit of the endonasal approach.
- Intraorbital lesions require exposure and dissection through the medial wall of the orbit.
- Intraconal inferomedial lesions are typically accessed through a corridor between the medial rectus muscle (MRM) and inferior rectus muscle.
- MRM retraction can be performed in a variety of ways. Isolation and retraction with vessel loops at the globe via a transconjunctival approach are a quick and efficient technique, best applied in concert with an ocular surgeon.
- The benefit of reconstruction is under investigation. In larger defects with significant intraconal dissection, reconstruction may have substantial benefit.

Typical Pathologies Treated

Tumors of the orbit are exceedingly rare, with an incidence of three to five tumors per 1 million people per year.[11] Endonasal orbital surgery is well suited for benign soft tissue tumors such as hemangiomas or lymphangiomas[12]; however, there is a wide breadth of pathologies that are amenable to this approach. Both primary tumors of the orbit located medially as well as tumors that extend from the orbital bone, paranasal sinuses, and anterior skull base into the medial orbit could be managed through an EEA.

In addition to curative resection, the EEA can be used with diagnostic intent or for symptom alleviation. In the setting of an unknown intraconal or orbital apex mass, endoscopic access provides a good option for direct biopsy and decompression without compromising future surgical resection if needed. Other common benign lesions found in this area include fibrous dysplasia, osteoma, meningioma, orbital schwannoma, optic nerve glioma, and orbital pseudotumor. Malignant lesions in the orbit are commonly lymphoma or metastases. The role of the EEA is diagnostic in some of these situations that are primarily managed through nonsurgical options. Conversely, orbital extension in selected low-grade sinonasal malignancies can be chased via EEA with goal of negative margins. For example, resection of the medial periorbita for esthesioneuroblastoma could be considered in this setting to achieve negative margin status when only the orbital wall is involved. Finally, in rare selected incidences, subtotal resection remains an acceptable operative plan in the setting of symptom relief with optic nerve decompression.

Anatomic Location

The orbit contains several critical neurovascular structures and interfaces with both the paranasal sinuses and skull base. As such, understanding of this anatomy is vital (see Anatomical Relationship of Surrounding Structures by Peris-Celda in this issue).[13] Pathology within the orbit can be classified as either extraconal or intraconal based on its location with respect to extraocular musculature. The medial rectus muscle (MRM) is the first structure encountered in the endonasal approach and attaches to the annulus of Zinn along with the inferior, lateral, and superior rectus muscles. The superior oblique muscle (SOM) and inferior rectus muscle (IRM) are also frequently exposed in this approach. The SOM originates from the greater wing of the sphenoid above the medial margin of the optic canal, changes direction at the trochlea at the superomedial orbit, and inserts into the superior lateral surface of the globe.

The optic nerve passes through the optic canal in the annulus of Zinn, deep to the muscle attachments. Generally, the optic nerve is considered to be the lateral limit of dissection from an endonasal perspective; however, in certain circumstances, dissection beyond this border may be possible.[14,15] The oculomotor nerve (OMN) enters the orbit through the lateral compartment of the annulus of Zinn and immediately branches into superior and inferior rami. The inferior ramus ultimately gives off two branches that travel anteroinferiorly to innervate the IRM and inferior oblique muscle as well as one branch that passes medially below the optic nerve to innervate the MRM. The nerve branches to these muscles run medial to and insert on the medial muscle face. Understanding this relationship is critical when performing intraconal dissection. Most often, the OMN branch to the MRM inserts at the level of the sphenoid face.[15]

Key vascular structures in the medial orbit include the ophthalmic artery (OA), inferomedial muscular trunk (IMT) of OA, anterior ethmoidal artery (AEA), and the posterior ethmoidal artery (PEA). The OA gives off the IMT which supplies the medial and IRM. Similar to the associated nerves, the insertion of the arterial supply is predominately located along the medial mid-belly of the inferior and medial rectus musculature. The AEA can reliably be found passing between the superior oblique and MRM, while the PEA has a more variable course. A key endonasal anatomical relationship is the position of the optic canal relative to the ethmoidal foramina. The optic canal and the posterior ethmoidal foramen lie 6 mm apart, and the anterior ethmoidal foramen can be found 12 mm anterior to the posterior ethmoidal foramen.[16] Anatomic variants, such as duplication or absence of PEA, must not be forgotten when relying on this relationship.

Some institutions use IMT as a way to separate the orbit into anterior and posterior zones to facilitate risk assessment during dissection.[15,17] Anterior to the IMT lies the AEA, PEA, neurovascular supply to the extraocular muscle (EOM), as well as the muscles themselves. Posterior to the IMT, within the orbital apex, is the most technically challenging zone to access. The downside to this system is the inability to consistently identify the IMT well radiographically. In general, the orbital apex contains the confluence of critical structures, with a resultant higher risk intraconal dissection.

Recently, more has been written on anatomical considerations for orbital tumors with extension intracranially or into the skull base, pterygopalatine fossa (PPF), and infratemporal fossa (ITF); these lesions often require management with multiple approaches. In some instances, combining traditional EEA approaches to the skull base and PPF/ITF with endoscopic endonasal orbital approaches can successfully resect these challenging lesions (see "Combined Surgical Approaches In and Around the Orbit" article by Stefko in this issue).

Specialties Involved

The need for different specialties in an EEA to the orbit varies depending on the location, pathology, and complexity of the lesion. It should be stressed that the use of a multidisciplinary discussion benefits from the expertise of the individual specialties to determine the ideal approach and goals of surgery. Otolaryngology, ophthalmology/ocular surgery, and neurosurgery comprise the specialist team that is typically involved in the more complex lesions. This becomes especially important when dealing with lesions involving the orbital apex and those with intracranial, PPF and/or ITF extension. With benign pathology, the literature reports a relatively lower need for a multidisciplinary approach during the surgical resection. In a study of 23 patients with exclusively intranasal resection of orbital cavernous hemangioma (OCH), all cases had an

otolaryngologist present, while 26.1 and 21.7% had ophthalmology and neurosurgery, respectively.[8] At our institution, all three specialties are routinely involved in both the preoperative decision making and the surgical intervention. The expertise of otolaryngology in the adjacent paranasal sinuses and endoscopy is routine for these approaches. Neurosurgery is routinely involved with pathology that has skull base, dural, or intraconal involvement. Ocular surgery is vital to the identification for critical structures, orbital manipulation, aspects and techniques of dissection, and their expertise in the management of these tumor types. Intraconal pathology necessitates the involvement of all three specialties due to the complexities of access and dissection.

In conclusion, a multidisciplinary team is ideal for the preoperative decision making and the exact constitution of surgeons in the procedure depends on the tumor characteristics and location. However, similar to skull base surgery, involvement of all three specialties may be a good way to develop team repertoire for tackling much more challenging cases in the future.

Operative Technique

Pre- and Perioperative Considerations

Preoperatively, patients undergo a comprehensive ophthalmologic evaluation as well as appropriate imaging including both computed tomography (CT) and magnetic resonance imaging (MRI) to assess the bony anatomy of the orbit and the soft tissue extent and imaging characteristics of the tumor. Intraoperative navigation is typically recommended, although the benefit within the orbit is less defined. Specific equipment is similar to other endoscopic endonasal surgeries and includes 0- and 45-degree endoscopes, an endoscopic bipolar, vessel loops for EOM retraction, and endoscopic instruments and dissectors. In addition, cottonoids or cotton tip applicators are always helpful for intraconal dissection and management of intraconal fat.

Endoscopic Endonasal Approach

Exposure to the medial and inferior orbital walls begins with a middle meatal antrostomy followed by sphenoethmoidectomy. Often, the ipsilateral middle turbinate is resected for improved visualization and working room for multiple instruments. Preservation of the middle turbinate may be considered if the approach is being performed for solely diagnostic purposes. The indications for a uninarial or binarial approach are largely surgeon and institution dependent. Our institution will often use a uninarial approach in the mid-orbit while converting to a binarial approach in the orbital apex and when optic canal exposure is required. If a binarial approach is indicated, a posterior septectomy is required. Within the sphenoid sinus, the optic nerve and internal carotid artery should be identified posteriorly along with medial opticocarotid recess and lateral opticocarotid recess. Skeletonization of the lamina papyracea proceeds inferiorly to the level of the maxillary antrostomy, posteriorly to the sphenoid face, anteriorly to nasolacrimal canal, and superiorly to the ethmoid roof.

Once the sinus exposure is completed, the lamina papyracea should be carefully fractured away from the medial periorbita without violation of its integrity to prevent inadvertent orbital fat herniation. The degree of exposure often depends on the tumor size and location. It is typically performed in an anterior to posterior manner with tools such as a Cottle elevator. In rare cases with thicker bone, a 4-mm coarse diamond burr drill may be required. Bone removal//sulcus can be continued inferolaterally to the infraorbital foramen if needed. Periorbita is then incised from posterior to anterior in a direction parallel to the MRM. This can be tailored to the pathology location and size to minimize the degree of orbital fat herniation into the ethmoid cavity, especially anterior to the lesion as this can obscure access and visualization. Bipolar cautery is used to partially reduce orbital fat, although the management of the orbital fat for both dissection and visualization is one of the significant challenges of the technique.

For more inferomedial intraconal lesions, the dissection corridor is between the MRM and IRM. Retraction of MRM and IRM is performed with aid of oculoplastic surgery (►Fig. 1). The insertion of the muscle at globe is identified and retracted with vessel loops. With the aid of the tension (and anterior fat retraction when needed) this provides, the dissection corridor is further developed. When necessary, identification of the IMT and/or the optic nerve is performed with care. The tumor is identified within the orbital fat. Depending on the goals of surgery, the tumor can be resected or biopsied. In the case of OCH, an extracapsular dissection is performed often with a bimanual technique and endoscopic visualization (three-handed technique). Bipolar cautery is limited to the tumor capsule.

For superomedial access, a corridor between the MRM and SOM can be developed. AEA typically is present in this corridor, so the artery is frequently sacrificed for access and to avoid increased risk of retrobulbar hematoma.
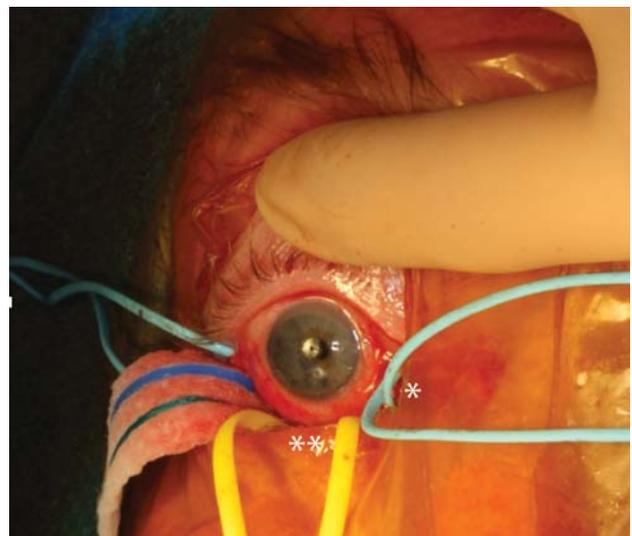

Fig. 1 Retraction of medial rectus muscle (*) and inferior rectus muscle (**) at their insertion site on globe with vessel loops.

Transcaruncular Approach

A transcaruncular approach can be used to supplement EEA as a way of gaining additional access to the medial orbit and aiding in fat retraction. In this approach, a medial conjunctival incision is made with scissors between the caruncle and plica semilunaris. The dissection is carried between orbital septum and Horner's muscle, and periorbita along the posterior lacrimal crest is incised with needle tip cautery. A periosteal elevator carries the dissection posteriorly, and the AEA and PEA are identified and cauterized. This allows transorbital access to the same medial portion of the orbit as the endoscopic endonasal orbital approach and allows for multiple corridors to be used simultaneously for retraction, dissection, and tumor removal. It also allows for a more direct approach for the anterior aspect of some tumors. At the end of the case, reapproximating sutures are not required for this approach.

Combined EEA and Endoscopic Transorbital Approaches

The application of these techniques to sinonasal and skull base tumors with orbital extension and/or orbital tumors with skull base extension is similar. When EEA represents the preferred approach for the management of the primary skull base pathology, it is easy to add an endoscopic endonasal orbital approach to manage both the extra- and intraconal extension of the disease. Generally, the intraorbital portion is performed after the skull base resection to minimize the operative time when orbital fat herniation is present. With purely extraperiorbital resection, the endoscopic endonasal orbital approach can be performed before the intradural portion of the procedure. The application of these complementary and similar techniques has significant benefit in the management of pathology that cross the boundaries between the paranasal sinuses, skull base, and orbit (see "Combined Orbital Approach" article by Stefko in this issue).

Reconstruction Needs/Options

The benefit of medial orbital wall reconstruction is still being explored. In extraconal tumors, reconstruction is generally not required; however, pathology that requires extensive intraconal dissection with significant bone and periorbita removal may benefit from reconstruction.

In one multicenter review which contained 23 patients with OCH, none of the patients with extraconal pathology underwent reconstruction. Of those patients with intraconal lesions, 37.5% underwent reconstruction with either a mucosal graft or fascia lata.[8] One systematic review of 17 cases of intraconal OCH found that orbital reconstruction was used in 23.5% cases and included nasoseptal flap (NSF), mucosal graft, bone fragments, and silicone sheet.[7]

While there is no consensus on the preferred reconstruction, the two most commonly used are the NSF and free mucosal graft. The NSF has been shown to provide coverage of the entire medial and inferior orbits[18] which may prevent excessive medial rectus scarring and limit postoperative diplopia and enophthalmos. This is a good option because of its familiarity to many skull base surgeons and the same

flap allows for more extended approaches if needed.[19] At our own institution, a free mucosal graft is commonly used and can often be harvested from the resected middle turbinate and carries minimal or no additional patient morbidity.

The likelihood of immediate postoperative compartment syndrome is decreased as the NSF and free mucosal graft are soft initially allowing for blood and fluid to escape. Eventually, the flap contracts over time, creating a barrier similar to a patient's native periorbita. Packing around the flap at our institution is typically done with absorbable hemostatic agents. Nonabsorbable packing may provide more robust support but may be avoided to prevent postoperative increased intraocular pressure in the setting of increased risk of postoperative edema/bleeding.[20]

Intraoperative Neurophysiology

Intraoperative neurophysiology is not routinely used secondary to direct manipulation of muscles throughout the case; however, in select situations where tumors involve solely the orbital apex, neurophysiology monitoring (electromyography) can be a nice adjunct in the identification of cranial nerves (CNs) or branches. Intraoperative visual-evoked potentials are another option that is infrequently used to monitor when a patient is at a higher risk of postoperative visual deterioration.[21] This is not routinely used at our institution but is worth study to determine its sensitivity and specificity for nerve injury.

Complications and Tumor Outcomes

From some of the largest multicenter studies and systematic reviews, the most common complications of this approach include diplopia, enophthalmos, and incomplete resection, with rates being higher for intraconal versus extraconal pathology. Ina multicenter study with 23 patients from six centers with OCH, 25% of patients with intraconal pathology had diplopia and enophthalmos. In contrast, this complication occurred in 14.3% of patients with extraconal resection.[8]

More serious complications include optic nerve injury, cranial neuropathies, cerebrospinal fluid (CSF) leak, and retrobulbar hematoma. In one case series of varying pathology, transient CN III palsy occurred in 6.25% of patients.[3] Another article, 12 subjects with varying pathology, reports 16.7% decreased visual acuity and 8.3% CSF leak.[22]

The literature on complications for this approach in non-OCH pathology is scant and still developing. Our own institutional data had a wide variety of complex pathology that was not purely limited to the orbit and showed a CSF rate of 3.7%. Other complications from our series included postoperative orbital hematomas (2.5%), transient CN IV palsy which resolved by first postoperative visit (1.2%), epistaxis (1.2%), mucocele development(1.2%), and respiratory failure (2.5%).

One case series presented a mix of endonasal and combined approaches for benign, malignant, infectious, and inflammatory etiologies of the orbital apex. This study reported 72% of patients require adjuvant treatment which included predominantly radiation therapy. Other postoperative treatment

regimens included prolonged steroids, chemotherapy, and antifungal therapy.[22] As one begins to move away from benign lesions in a typical posterior inferomedial corridor, the ability to obtain gross total resection decreases and the need for adjuvant treatment increases. In these situations, one benefits immensely from having a multidisciplinary team involved both preoperatively and in the operating room. The differing pathologies that can occur in this anatomical location make the expertise of each specialist invaluable to patient care.

Approach Limitations

Limitations to this approach hinge most prominently on the relationship of the pathology to the optic nerve. An early

algorithm to guide surgical approach based on anatomical location of tumor used a clock model.[20] EEAs provide access to the mid- and posterior orbit along with the orbital apex from 1 to 7 o'clock (in a right eye). Lesions lateral to this plane of the optic nerve should employ a different approach to avoid undo manipulation.

The Cavernous Hemangioma Exclusively Endonasal Resection staging system is a newly published classification which focuses on anatomic location of OCH to stratify lesions based on complexity of resection.[23] Relationship to critical structures defines each stage, and specific focus is placed on the optic nerve, IMT, and EOMs, as well as extension into and/or through the optic canal, inferior orbital fissure, and superior orbital fissure. The classification ranges from stages
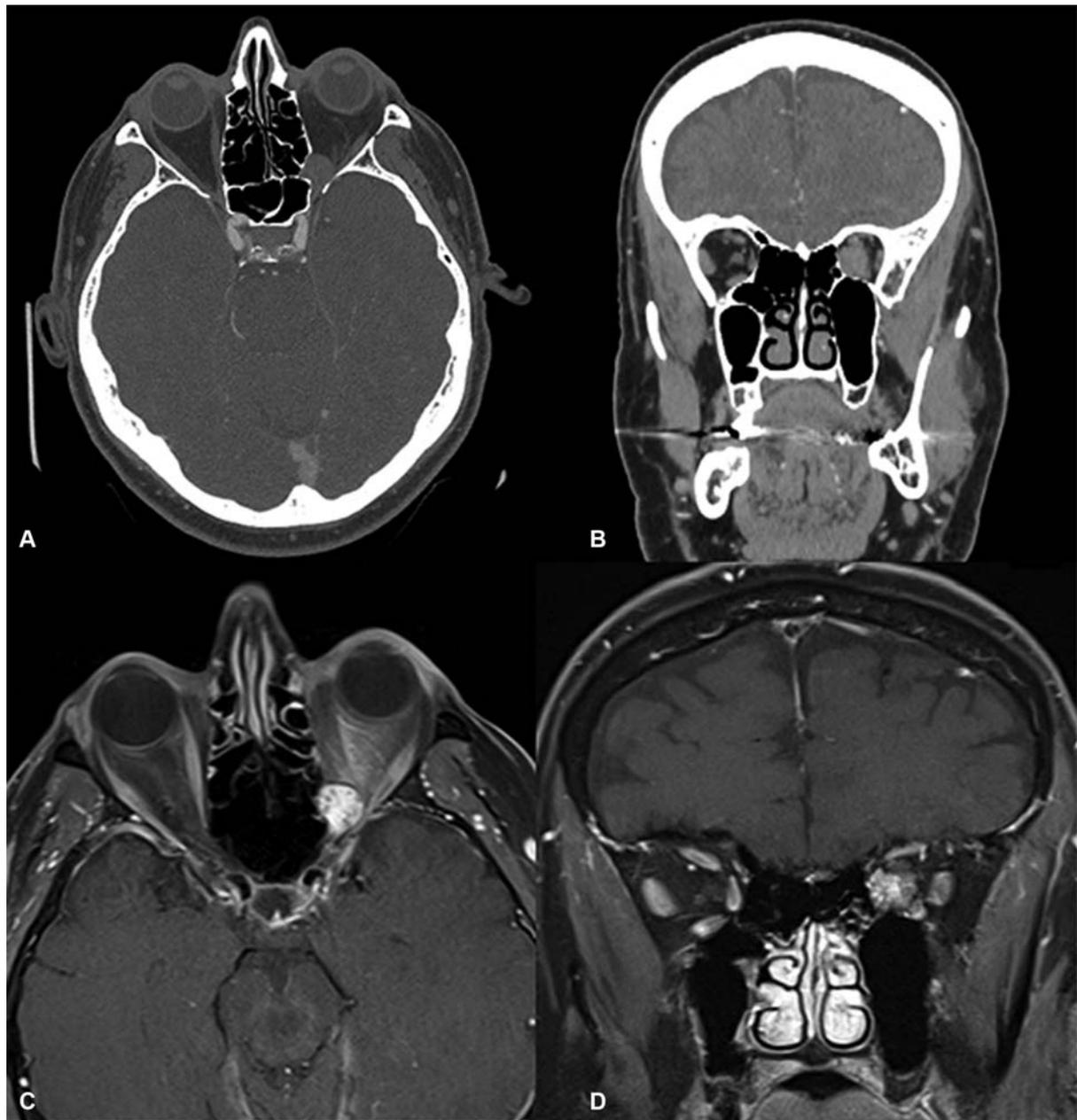

Fig. 2 (**A**) Axial and (**B**) coronal cut through CT showing enhancing left orbital apex lesion. (**C**) Axial and (**D**) coronal T1 MRIs with gadolinium of the same lesion highlight the enhancing lesion in the intraconal space. CT, computed tomography; MRI, magnetic resonance imaging.

I to V with stage V being the most technically challenging and representing OCH that extends beyond the orbit.

In that same article, a consensus of experts agreed on orbital locations which are favorable for endoscopic resection.[23] Tumors medial to a plane along the long axis of the optic nerve are amenable for approach. If tumor extends inferolaterally—if tumor remains below a plane from the contralateral naris through the long axis of the optic nerve

(plane of resectability)—it can still be resected with this approach.

Case Example and Case Series

Case 1 (Intraconal Tumor)

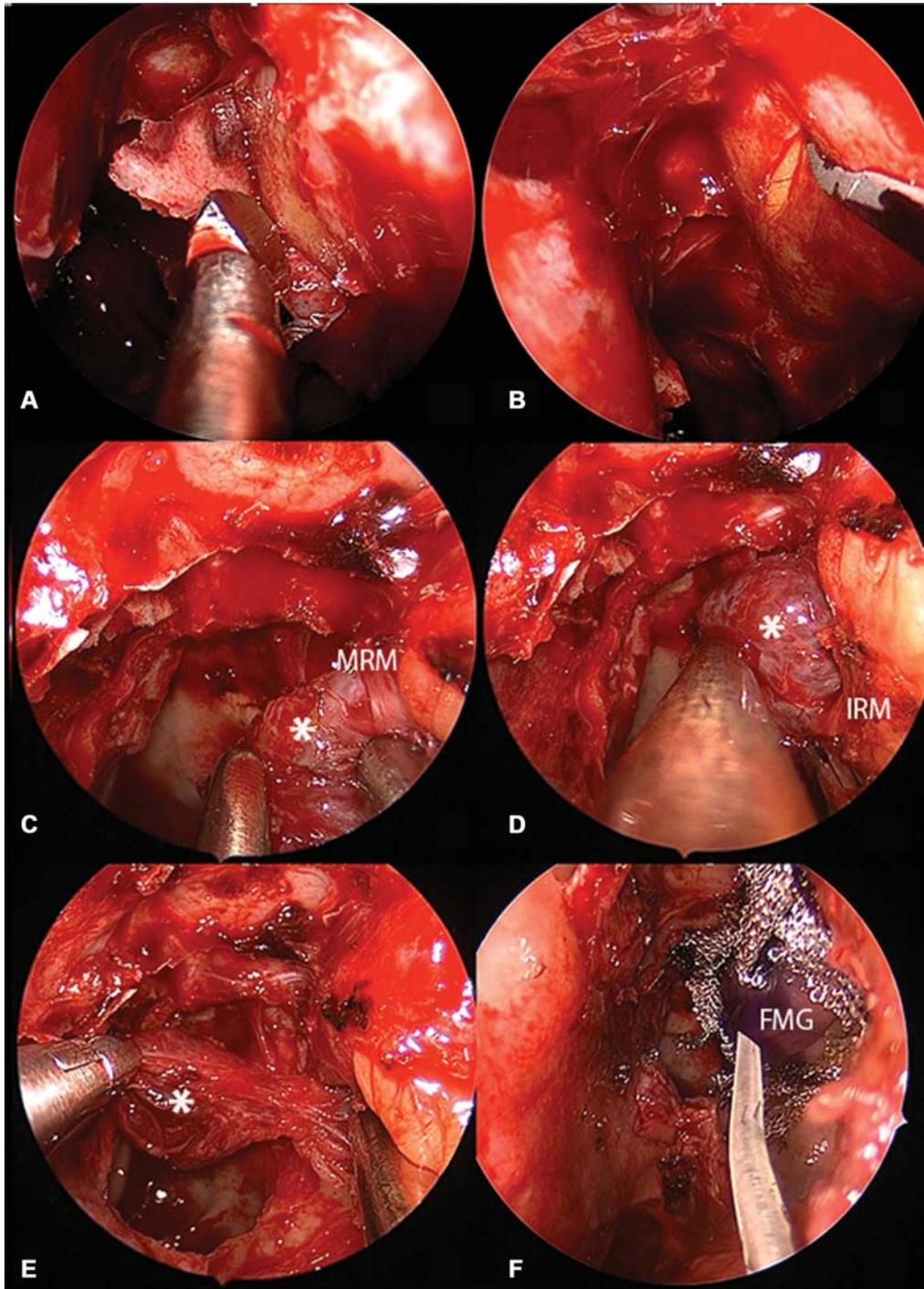Patient is a 66-year-old man with a left orbital lesion incidentally discovered 4 years prior. The decision was

Fig. 3 (**A**) Removal of lamina papyracea from periorbita with Cottle dissector. (**B**) Incision of periorbita in a posterior to anterior direction with a sickle blade. (**C**) Peeling vascular lesion (*) from medial rectus muscle (MRM). (**D**) Freeing inferior attachments of hemangioma (*) from inferior rectus muscle (IRM). (**E**) Removal of cavernous hemangioma (*) from orbital apex. (**F**) Placement of free mucosal graft (FMG) overlying medial orbital wall defect.

made by treating physician to observe. Over the past year, he experienced progressive worsening blurry vision and left retro-orbital headache. On exam, his corrected visual acuity was 20/50 in the left eye with decreased color vision and afferent pupillary defect. CT and MRI are shown in ►**Fig. 2**.

Due to the location of this tumor, otolaryngology, neurosurgery, and oculoplastic surgery were present for the approach and resection. As discussed earlier, the approach was started with a maxillary antrostomy and complete sphenoethmoidectomy. The lamina papyracea was removed with a combination of a drill, Cottle dissector, and ball probe. The periosteum was incised with endoscopic scissors directly over the tumor. The fibrous capsule around the tumor was dissected and removed. The small periorbital defect was repaired with free mucosal graft (►**Fig. 3**).

Postoperatively, the patient had an improved visual exam with 20/30 vision in the left eye. The afferent pupillary defect resolved, and the patient regained full color vision in that eye. Pathology was cavernous hemangioma.

Design: Literature review, retrospective case series

Setting: Tertiary Medical Center

Participants: Patients who underwent EEA for access to orbital or orbital apex pathology from 2002 to 2014 were reviewed.

Main Outcome Measures: To review the varying orbital pathologies that can be approached by EEA while presenting postoperative outcomes and complication rate at our institution.

Results: Seventy-seven patients underwent 81 EEA procedures for resection or decompression of orbital pathology. Meningioma was the most common pathology (34.6%) followed by esthesioneuroblastoma (6.2%) and squamous cell carcinoma (6.2%). The average follow-up was 23.5 months and complications included CSF leak (3.7%) and postoperative orbital hematomas (2.5%). Overall 37.1% of patients had symptomatic improvement and 60.4% stabilized.

Case Series

Seventy-seven patients who underwent 81 endoscopic endonasal procedures for resection or decompression of symptomatic orbital pathology from 2002 to 2014 were retrospectively reviewed. The most common preoperative complaint was visual deterioration followed by exophthalmos and diplopia. Meningioma was the most commonly encountered pathology accounting for 34.6% of the cases followed by esthesioneuroblastoma (6.2%) and squamous cell carcinoma (6.2%). Other pathologies include osteoma, trauma, metastatic disease, hemangioma, fibrous dysplasia, adenoid cystic carcinoma, schwannoma, optic glioma, chondrosarcoma, and sinonasal undifferentiated carcinoma.

The average follow-up was 23.5 months and complications included CSF leak (3.7%), postoperative orbital hematoma (2.5%), transient CN IV palsy (1.2%), epistaxis (1.2%), mucocele development (1.2%), and respiratory failure (2.5%). Overall 37.1% of patients had symptomatic improvement, 60.4% remained unchanged, and 2.5% worsened. The pathology was broken into extraorbital (33.3%), intraorbital/extraconal (34.6%), and intraconal (32.1%). All patients who had extra-

orbital lesion had improved/stable symptoms, while intraconal lesions had the highest rate of visual complications (7.7%).

This series shows the diversity of pathology and location of pathology that can be treated with the EEA approach.

Conclusion

EEA to pathology in the posterior inferomedial orbit offers improved visualization and access to certain areas, especially the orbital apex, when compared with external approach. Patients have the benefit of no external scar in addition to the low rate of reported major complications. At our institution and many others, this approach is being applied to pathology that extends into the skull base and pushes the traditional lateral limit.

Conclusion

The EEA to pathology in the posterior inferomedial orbit offers decreased patient morbidity, improved visualization and access to this area as well as the medial orbital apex when compared with external approach. In addition, EEA serves as a window for more complex pathology that extends intracranially or into the PPF and/or ITF. This approach can also aid in resection of sinonasal or skull base pathology with orbital extension as demonstrated in our case series. In this complex field that is growing and expanding, a multidisciplinary team approach is strongly recommended.

Conflict of Interest
None declared.

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