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Case Report

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Infraorbital approach for retrobulbar orbital neurofibroma: a case report

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ABSTRACT

Retrobulbar orbital tumours are rare and treatment is challenging. These tumors include cavernous hemangioma, neurofibroma, A-V malformations, glioma etc. A variety of approaches have been used in resection of these tumours. The various approaches in literature include endoscopic transethmoidal and sphenoidal, lateral orbitotomy, transmaxillary infraorbital, and cranial and sub cranial approaches. Such tumours located inferior to the optic nerve can be reached through infraorbital approach avoiding traction on optic nerve or pressure on globe. We are reporting a retrobulbar orbital neurofibroma in a middle aged lady who presented with unilateral loss of vision, episodic giddiness, severe left sided headache, left orbital pain and epiphora on exposure to sunlight. MRI showed well defined 1.8×1.8×1.7 cm enhancing lobulated solid left intraorbital mass extending along the substance of inferior aspect of left optic nerve with mass effect. With transmaxillary infraorbital endoscope assisted approach, the retrobulbar tumor was exposed and enucleated. Floor of orbit was reconstructed with tensor fascia lata and nasal septal cartilage graft. The patient had uneventful recovery and immediate post operative visual acuity was perception of light and likely to improve further. Histopathology revealed neurofibroma. This case report highlights the advantages of infraorbital approach as it is a simple approach along the suture lines through the orbital floor with the help of endoscopic guidance. It has the advantages of avoiding traction on optic nerve or optic chiasma, no retraction of brain, no communication with cranial cavity, no pressure on globe and easy reconstruction of orbital floor.

Keywords: Infraorbital approach, Retrobulbar orbital tumor, Neurofibroma

INTRODUCTION

Retrobulbar orbital tumors are rare and encountered in elderly individuals. These tumors include cavernous hemangioma, neurofibroma, A-V malformations, glioma etc. Cavernous hemangioma is most common among them and is frequent cause for proptosis. Neurofibromas account for 0.6-5% of orbital tumors.[1] They may present with loss of vision and pressure effects. Resection of these tumors is challenging due to the complex and delicate anatomy. Various surgical approaches have been used to resect such tumors. Most commonly used approaches are endoscopic transethmoidal and sphenoidal, lateral orbitotomy, transmaxillary infraorbital, sub cranial approaches, fronto-zygomatic approach and supraorbital cranial approaches.[2]

We are reporting a case of middle aged lady who presented with left orbital retrobulbar neurofibroma causing pressure effects on optic nerve. She had no evidence of neurofibromatosis. We used transmaxillary infraorbital endoscope assisted approach to access and enucleate the retrobulbar tumor situated inferior to optic nerve. This is a simple approach along suture lines in the

orbital floor. In comparison to other surgical approaches infraorbital approach has advantages of avoiding traction on optic nerve or optic chiasma, no retraction on brain, no communication with cranial cavity, no pressure on the globe and easy reconstruction of the orbital floor.

Aim

To report the outcome of enucleation of retrobulbar orbital tumour by endoscope assisted transmaxillary infra orbital approach with minimum morbidity.

CASE REPORT

A 55 years old well built lady presented with history of loss of vision in left eye of one month duration. It was associated with severe left sided headache and left orbital pain and excessive lacrimation from left eye. There was no history of seizures, vomiting, diplopia or trauma. She had no other comorbidities and was not on any medications.

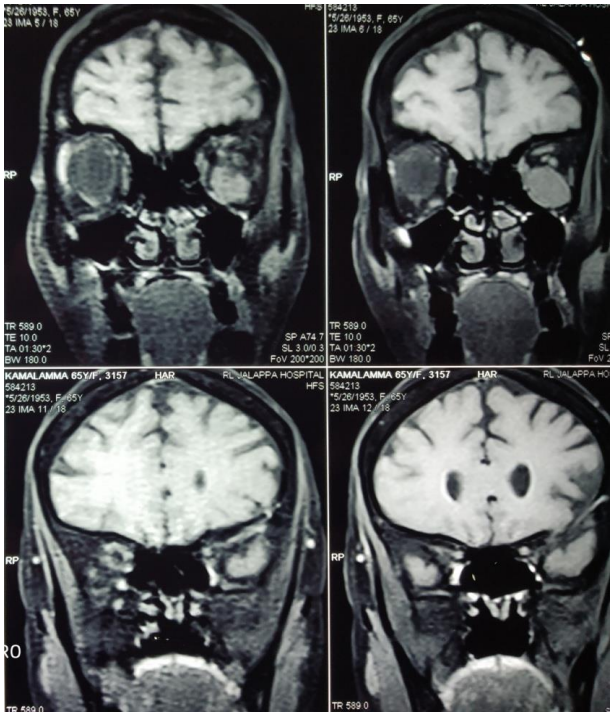

Figure 1: MRI shows well-defined 1.8×1.8×1.7 cm enhancing lobulated solid left intraorbital mass extending along the inferior aspect of left optic nerve with mass effect (stretching and lifting) on optic nerve.

On examination, patient was conscious, well oriented and clinically stable. Ocular examination showed visual acuity on affected side as finger counting in front of eyes. Ocular movements and examination of conjunctiva, cornea, anterior chamber, iris, pupil and lens was normal in both the eyes. Fundoscopy in both the eyes was normal. There were no other neurological deficits.

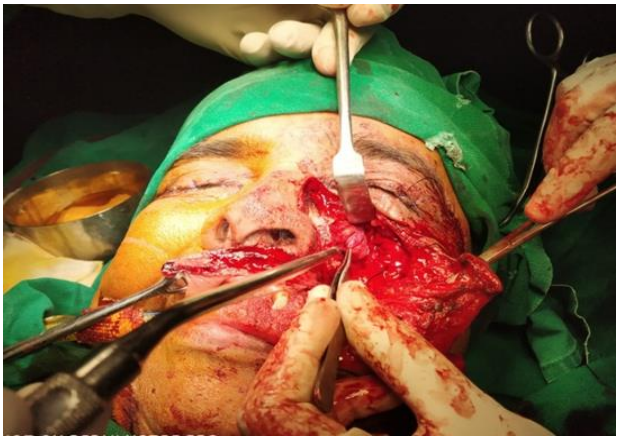

Figure 2: Infraorbital approach cutting infraorbital ridge and floor of orbit between nasolacrimal duct and anterior root of zygoma.

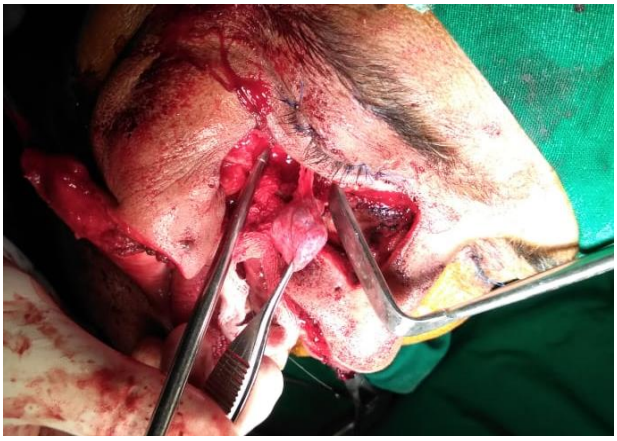

Figure 3: The globular, well demarcated, 2×2 cm firm tumour with prominent capillaries being delivered.

MRI showed well-defined 1.8×1.8×1.7 cm enhancing lobulated solid left intraorbital mass extending along the inferior aspect of left optic nerve with mass effect (stretching and lifting) on optic nerve (Figure 1). Patient was taken up for surgery under general anesthesia. Weber Ferguson incision was taken with diffinbach extension. Cheek flap was raised and infraorbital nerve sacrificed. Orbital periosteum was lifted from orbital floor. Periorbital fascia was intact. Infraorbital ridge and floor was cut lateral to nasolacrimal duct and medial to anterior root of zygoma (Figure 2). Orbital floor was cut along the natural suture lines in orbital floor. Middle third of orbital floor was removed and further resection was done till 1.5 cm in front of orbital apex. The retrobulbar tumor was approached from inferior aspect. The 2×2 cms globular, well demarcated, firm tumour with prominent capillaries adjoining and along the axis of optic nerve was enucleated (Figure 3). Hemostasis was secured. Orbital floor was reconstructed using tensor fascia lata graft and a piece of nasal septal cartilage (Figure 4). Wound was sutured in layers (Figure 5). Patient was administrated antibiotics and systemic steroids for one week. Post operative visual acuity was perception of light on the

operated side and is improving.Ocular movements were normal and patient was relieved of pain in the eye, giddiness and epiphora. Histopathology of the specimen showed neurofibroma.

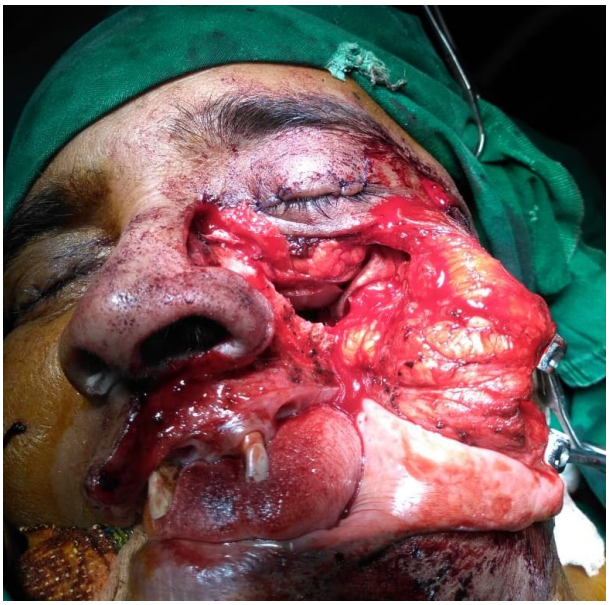

Figure 4: Orbital floor reconstructed using tensor fascia lata graft and a piece of nasal septal cartilage.

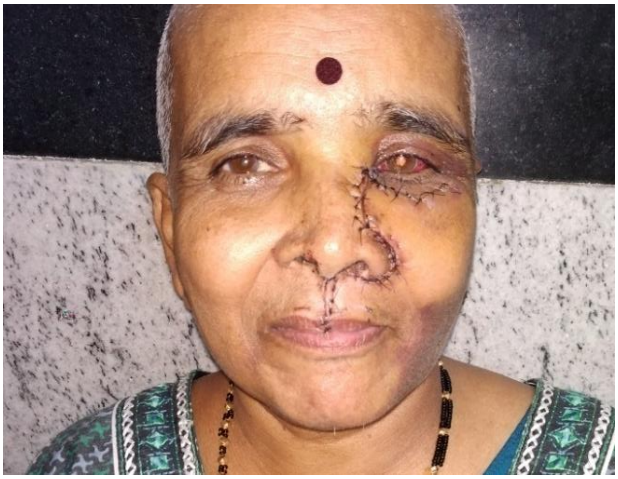

Figure 5: Post operative appearance of patient.

DISCUSSION

Retrobulbar orbital tumors are rare and encountered in elderly individuals. These tumors include cavernous hemangioma, neurofibroma, A-V malformations, glioma, etc. Orbital cavernous hemangioma is the most common among these accounting for 6% of all orbital tumors. Neurofibromas account for 0.6-5% of orbital tumors. Higher incidence is reported in neurofibromatosis-1.[1] Our patient had an isolated neurofibroma along the axis of optic nerve. They are asymptomatic initially and later present with axial proptosis or visual disturbance. Retrobulbar cavernous hemangiomas are the second most common cause of unilateral proptosis.[2]

The surgical approach should be selected for each case based on size, location, and relationship with neighboring structures like optic nerve as assessed by imaging. Less traumatic approaches are selected in this functionally and cosmetically important region.

The success of surgery depends on the size of the tumour and characteristics of the surgical site as well as experience of the surgeon and familiarity to orbital anatomy. The location of the tumor in relation to orbital muscle cone is very important to decide the surgical approach. Intraconal tumors have higher morbidity compared to extra-conal tumors.

Presenting symptoms include optic nerve compression resulting in loss of colour vision, diplopia, visual field defects and blindness.[2] Our patient presented with unilateral loss of vision, unilateral headache and lacrimation as reported in other studies.[2] Her visual acuity on affected side was finger counting in front of eyes. The retrobulbar tumour was identified and evaluated by MRI.

The surgical approach should avoid traction or devascularization of optic nerve. Lateral orbitotomy has been described as the preferred surgical approach in the literature.[2] But it has the disadvantages of leaving an external deformity- a visible depression in the anterior part of temporal fossa caused by detachment of the temporal muscle from its insertion also causing loss of support to the orbit.[2]

Retrobulbar tumors can also be accessed by sub cranial and cranial approaches which are invasive, will have communication with cranial cavity, may require retraction of brain, traverse important neurovascular structures, and will require highly skilled surgeon.[3]

Endoscopic endonasal approach is used for orbital tumors and lesions within Meckels's cave. Endoscopic transnasal approach was first described in 1999.[4] This is the most common approach used in the management of orbital tumors located inferior and medial to the optic nerve.[4,5] It requires complete sphenoethmoidectomy, partial middle turbinectomy, medial maxillectomy, transposition of nasolacrimal duct, lamina papyracea removed and periorbita incised horizontally.[4] Residual tumor may require frontozygomatic approach. Transient diplopia and dyschromatopsia can result after resecting intraconal tumors by this approach.[4]

For tumors situated in lateral aspect of retrobulbar orbit, the working angle will be difficult and limited space for handling the instruments.[6] Ethmoidectomy exposes the orbit to nasal infections and affects the recovery of the patient.

Combined endoscopic transmaxillary transnasal approach has been described in the literature for the management of extreme lateral lesions in sphenoid sinus, pterygoid region and posterior orbit.[6] These anatomical areas are difficult to reach as they are posterior to maxillary sinus and close to skull base. However this approach provides working space and maneuverability of instruments and avoids post operative nasal crusting and adhesions. It can result in infraorbital hypoesthesia and facial edema.[6]

Retrobulbar space can also be accessed by a transconjunctival approach with lateral canthotomy and transient extraocular muscle severing without lateral orbitotomy. It is rarely used for orbits because of insufficient exposure during the surgery and can result in post operative cicatricial entropion, ectropion of the eyelid, lower eyelid retraction, lateral telecanthus, lower eyelid detachment, canalicular laceration, chemosis, and lacrimal sac laceration.[7]

Osteoplastic maxillotomy approach has been described for the management of infraorbital nerve schwannomas where the tumor is accessed by Weber Fergusson incision with a subciliary extension followed by en bloc osteoplastic maxillotomy and zygomatic osteotomy. However, diplopia can result due to displacement of the ipsilateral eye at the level of the inferior orbital rim due to scarring of the skin and loss of the inferior palpebral fat. Infraorbital nerve anesthesia can be permanent sequelae of this procedure.[8]

Endoscopic transnasal transorbital approach to lateral orbital apex involves resection of lamina papyracea and the medial portion of the orbital floor followed by incision of the periorbita along the inferomedial aspect of the orbit. Medial and inferior rectus muscles may require retraction or detachment to visualize the optic nerve.[9-12] Lesions that extend superolaterally are a contraindication for endoscopic transnasal orbital surgery.[13]

We accessed the tumor by removing middle third of infraorbital ridge thereby preserving the nasolacrimal duct. Middle third of orbital floor was cut along the natural suture lines in orbital floor. This approach provided relatively avascular field and avoided pressure on the globe or stretch on the medial and lateral canthal ligaments. It gave an excellent exposure and retrobulbar tumor was easily approached from inferior aspect and enucleated dissecting from orbital fat. There was no traction on the optic nerve. Similar transmaxillary approaches have been adopted by few other authors.[3,6]

We reconstructed the defect in orbital floor using tensor fascia lata graft and a piece of nasal septal cartilage. Post operative visual acuity was perception of light and is gradually improving. Ocular movements were normal and patient was relieved of pain in the eye and epiphora. Similar relief of symptoms was seen in other case reports where retrobulbar tumours were accessed by tramsmaxillary approach. Histopathology of the specimen revealed neurofibroma.

CONCLUSION

Transmaxillary infraorbital approach for retrobulbar tumors is a novel approach which minimizes pressure to the globe or traction to optic nerve, provides excellent exposure, avascular field, reduces chances of post operative orbital contamination, avoids communication with cranial cavity and ensures good cosmetic outcome by easy reconstruction of the defect in orbital floor and infraorbital rim.

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