Case Report

Cryo Assisted Minimally Invasive Surgery for the Treatment of Orbital Cavernous Haemangiomas

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Purpose: To study cryo assisted minimally invasive surgery for the treatment of orbital cavernous hemangiomas.

Material and Methods: We present a case series of six patients having orbital cavernous haemangiomas. All patients underwent minimally invasive cryosurgical extraction of the tumour at the Armed Forces institute of Ophthalmology, Rawalpindi.

Results: All the tumours were successfully removed en bloc with the help of cryo extraction. The vision was improved in all but one case. The recovery was uneventful and histopathological reports confirmed the diagnosis after more follow-ups of nine months, no recurrence was observed in any of the operated eye.

Conclusion: Cryo-assisted minimally invasive surgery offers an exciting approach for management orbital cavernous hemangiomas, with good cosmetic results and early functional recovery.

Cavernous haemangioma is the most common primary benign vascular orbital tumour. It presents with painless unilateral proptosis that is reducible and associated with hyperopia. Most cavernous haemangiomas are intraconal and lateral in location. They result from new vessel formation, proliferation of tissue components of the vessel wall, and hyperplasia of cellular elements of vascular origin. Computed tomography (CT) and magnetic resonance imaging (MRI) are of particular importance in the diagnosis of orbital vascular lesions.

Surgical access to posterior orbit is difficult because of the presence of delicate structures including optic nerve, ophthalmic artery and veins, extra-ocular muscles and nerves. Surgical approaches to treat orbital disease should provide a good exposure of intra-orbital anatomical structures, and provide good cosmetic results. Different approaches to the intra-orbital space have been described in the literature.

In an attempt to maintain this philosophy and to avoid large incisions still providing an increased operative exposure, we describe cryo-assisted minimally invasive approach which mostly avoids bone removal or intraconal muscle sectioning. It provides access to the superior, medial, lateral, and inferior quadrants of the orbit, depending on the extent of the conjunctival and eyebrow incisions. Thus, complete exposure and resection of very large intra-orbital lesions is feasible with reduced morbidity.

MATERIAL AND METHODS

We describe the outcome of 6 patients with cavernous haemangioma of the orbit treated in our department between April 2008 and Feb 2010. Study Design was interventional case series. Patients underwent minimally invasive surgery through different approaches, completely removing the angiomas. All patients with intraconal mass and a strong clinical suspicion of cavernous hemangiomas underwent
minimally invasive cryosurgical extraction. Patients having any surgical intervention in the past were excluded from the study. The presenting features of all six cases are given in Table 1. Pre-op marking of site of incision and tumor was carried out. The tumor was reached either by lateral orbitotomy, a lid crease incision, or via trans-conjunctival approach depending upon location of tumor (Table 2). After securing the vascular connections, lesion was extracted en bloc with the help of cryo-probe and blunt dissection. Wound was closed in layers. Histopathology was carried out in the end to confirm the diagnosis.

RESULTS

The mean age of patients was 32 years with a range of 16-55 years. All the tumors were successfully removed en bloc with the help cryo extraction. The vision improved in all but one cases, and Table 3 shows the comparison of pre-op and post-op visual acuity of the selected cases. The recovery was uneventful and histopathological reports confirmed the diagnosis of cavernous hemangioma. After a mean follow-up of nine months, no recurrence was observed in any of the operated eyes.

CASE 1

A 32-year-old female presented with a 07 years history of painless progressive proptosis and impaired visual acuity of her right eye. She denied history of trauma. In the past, she was given a course of systemic steroids and biopsy was attempted elsewhere. Her systemic and medical history was not contributory. She was admitted in another institute where she was given a course of systemic steroids and biopsy was taken which was non-diagnostic because the sample was inadequate. She was using only lubricants on admission to our hospital. On examination the best corrected visual acuity (BCVA) was 6/36 in right eye and 6/6 in left eye. Right eye showed proptosis, diplopia in primary gaze, chemosis and restriction of extraocular movements in all gaze positions. Examination of right fundus showed choroidal folds. CT scan orbit and brain with coronal sections showed a well circumscribed intraconal lesion in right orbit. A plan for excisional biopsy was made.

The mass in her right orbit was approached via a subciliary incision. Extensive adhesions were encountered after opening the orbital septum, probably from her previous biopsy. These were lysed and the rest of the mass was extracted en masse using a cryoprobe and blunt dissection. The recovery was uneventful. Histopathology revealed an encapsulated cavernous haemangioma. Her vision improved to 6/9, the choroidal folds resolved and ocular movements recovered in all gaze positions (Fig. 1 and 2).

CASE 2

A 37 years old man presented with gradual painless proptosis with gradual loss of vision in his left eye for the last 08 years. BCVA in right eye was 6/6, while in the left eye was 6/60. Left eye showed axial proptosis of 35 mm and supero-temporal dystopia of 5 mm (Fig. 3). Extra-ocular movements were restricted in all gaze positions. There was no retropulsion or bruit. Fundus showed choroidal folds. Examination of right eye was unremarkable. CT scan showed a large well circumscribed intraconal lesion in left orbit (Fig. 4).

The patient was planned for orbitotomy under...
general anesthesia. After marking an extended lazy S incision till zygomatic arch, lateral orbital margin was removed and the lesion approached through blunt dissection. On opening the peri-orbita, a plump, nodular, encapsulated mass with vascular channels on its well defined surfaces was found. An apical vascular tag was identified and cauterized; to prevent the gush of blood. A cryoprobe assisted extraction was done. The lesion was completely excised. Wound was closed in layers. Visual acuity of patient was checked in the evening to exclude optic nerve compression. Histopathology confirmed the diagnosis of multi-lobulated, well encapsulated left cavernous haemangioma. Vision in the left eye improved to 6/24 six weeks after the surgery (Fig. 5).

CASE 3
A 35 years old female presented to our hospital with inferior displacement of right eye for last 04 years associated with gradual deterioration of vision. There was no history of trauma or any systemic illness. On ocular examination BCVA in right eye was 6/36 while that in left eye was 6/6. Right eye showed 6 mm inferior dystopia, 2 mm of proptosis compared to left eye and restriction of ocular motility more marked in vertical than in horizontal gaze (Fig. 6). There was no
retropulsion or bruit. Fundus showed choroidal folds. Examination of left eye was unremarkable. CT-Scan revealed a large extraconal, well circumscribed lesion in supero-nasal region of orbit (Fig. 7).

Patient was planned for orbitotomy under general anesthesia. Pre op. marking was made (Fig. 8). Lid crease incision extending temporally 15 mm lateral to temporal orbital margin was made. After penetrating the septum, the lesion was approached through blunt dissection which revealed a well encapsulated nodular mass. Vascular connections were secured and the lesion was removed en bloc with the help of cryoprobe (Fig. 9). It measured 35 x 22 mm. Wound was closed in layers. Histopathology confirmed the diagnosis of multi-lobulated, well encapsulated cavernous hemangioma. Vision in the left eye improved to 6/12 two weeks after surgery.

CASE 4

A 16 years old male presented to our hospital with inferior displacement of right eye for last 3 years associated with gradual deterioration of vision (Fig. 10). There was no history of trauma or any systemic illness. On ocular examination BCVA in right eye was 5/60 while visual acuity in left eye was 6/6. Right eye showed 07 mm inferior dystopia, 04 mm of proptosis compared to left eye and restriction of ocular motility more marked in vertical than in horizontal gaze. There was no retropulsion or bruit. Fundus showed choroidal folds. Examination of left eye was unremarkable. CT – Scan revealed a large extraconal, well circumscribed and encapsulated lesion in supero-nasal region of orbit.

Patient was planned for orbitotomy under general anesthesia. Lid crease incision extending temporally 13 mm lateral to temporal orbital margin was made. The lesion was reached through blunt dissection. The tumour was found to be plum coloured well encapsulated nodular mass. After securing the vascular connections, lesion was extracted en bloc with the help of cryoprobe. It measured 30 x 20 mm. Wound was closed in layers. Histopathology confirmed the diagnosis of multi-lobulated, well encapsulated left cavernous hemangioma. BCVA in the right eye improved to 6/12 six weeks after the surgery (Fig. 10).

CASE 5

A 65 years old man presented with gradually enlarging mass below right eye for last 3 years. On examination visual acuity in right eye was hand movement (HM) positive. He had relative afferent pupillary defect in the same eye. Extraocular
Fig. 11: Six weeks after the surgery.

Fig. 12: Mass bulging through inferior fornix causing mechanical ectropion.

Fig. 13: Well demarcated homogenous hyper dense mass involving inferonasal aspect of right orbit.

Fig. 14: Lesion below right eye ball visible under conjunctiva and with bluish discoloration of lower lid.

Fig. 15: Brownish well encapsulated mass measuring 3.0 x 2.8 cm was extracted en bloc.

Fig. 16: Cosmetic improvement three weeks post operatively.
Table 1: Patient characteristics at presentation

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age and Sex of Patient</th>
<th>Duration of Proptosis</th>
<th>Previous Treatment</th>
<th>Medical History</th>
<th>Proptosis and Diplopia</th>
<th>Ocular Motility</th>
<th>Fundus</th>
<th>CT Scan Orbits</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>32 (F)</td>
<td>07 years</td>
<td>Systemic Steroids</td>
<td>Clear</td>
<td>Axial, Primary Gaze</td>
<td>Restricted in all gazes</td>
<td>Choroidal folds</td>
<td>Well circumscribed intraconal</td>
</tr>
<tr>
<td>2</td>
<td>37 (M)</td>
<td>08 years</td>
<td>Nil</td>
<td>Clear</td>
<td>Axial, Primary Gaze</td>
<td>Restricted in all gazes</td>
<td>Choroidal folds</td>
<td>Well circumscribed intraconal</td>
</tr>
<tr>
<td>3</td>
<td>35 (F)</td>
<td>04 years</td>
<td>Local steroids</td>
<td>Clear</td>
<td>Inferior, vertical gaze</td>
<td>Restricted in vertical gaze</td>
<td>Choroidal folds</td>
<td>Circumscribed Extraconal large mass</td>
</tr>
<tr>
<td>4</td>
<td>16 (M)</td>
<td>03 years</td>
<td>Non-specific eye drops</td>
<td>Clear</td>
<td>Inferior, vertical gaze</td>
<td>Restricted in vertical gaze</td>
<td>Choroidal folds</td>
<td>Circumscribed Extraconal large mass</td>
</tr>
<tr>
<td>5</td>
<td>55 (M)</td>
<td>03 years</td>
<td>Local injection</td>
<td>Clear</td>
<td>Superior, vertical gaze</td>
<td>Severe restriction</td>
<td>Optic atrophy</td>
<td>Homogenous mass in inferior orbit</td>
</tr>
<tr>
<td>6</td>
<td>30 (F)</td>
<td>10 ½ years</td>
<td>Nil</td>
<td>Clear</td>
<td>No proptosis or diplopia</td>
<td>No restriction</td>
<td>Healthy</td>
<td>Homogenous mass in inferior orbit</td>
</tr>
</tbody>
</table>

Table 2: Surgical approach used depending on tumour location

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Location in Orbit</th>
<th>Approach</th>
</tr>
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<tbody>
<tr>
<td>1</td>
<td>Inferior</td>
<td>Sub ciliary</td>
</tr>
<tr>
<td>2</td>
<td>Retrobulbar</td>
<td>Lazy S incision with lateral orbitotomy</td>
</tr>
<tr>
<td>3</td>
<td>Supero-medical</td>
<td>Lid crease incision</td>
</tr>
<tr>
<td>4</td>
<td>Supero-medical</td>
<td>Lid crease incision</td>
</tr>
<tr>
<td>5</td>
<td>Infero-medical</td>
<td>Trans conjunctival</td>
</tr>
<tr>
<td>6</td>
<td>Infero-medical</td>
<td>Trans conjunctival</td>
</tr>
</tbody>
</table>

Table 3: Decimal Equivalent of Pre-op ad Post-op visual acuities of cases (n=6)

<table>
<thead>
<tr>
<th>Patient</th>
<th>Pre op VA</th>
<th>Post op VA</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>0.7</td>
<td>0.7</td>
</tr>
<tr>
<td>2</td>
<td>0.25</td>
<td>0.25</td>
</tr>
<tr>
<td>3</td>
<td>0.5</td>
<td>0.5</td>
</tr>
<tr>
<td>4</td>
<td>0.02</td>
<td>0.02</td>
</tr>
<tr>
<td>5</td>
<td>0.08</td>
<td>0.08</td>
</tr>
<tr>
<td>6</td>
<td>0.25</td>
<td>0.25</td>
</tr>
</tbody>
</table>

movements were severely restricted. Mass was visible bulging through inferior fornix causing mechanical ectropion. (Fig. 12) CT scan showed a well demarcated homogenous hyper dense mass involving infero-nasal aspect of right orbit (Fig. 13). Cryo-assisted extraction through trans-conjunctival approach was done. Patient made smooth post operative recovery but his vision did not improve.

CASE 6
A 30 years old female presented with gradual deterioration of vision and swelling below right eye ball for last one and half year. (Fig. 14) On examination her BCVA was 6/24 in right eye and 6/6 in left eye. There was a mass involving inferior aspect of right orbit but not causing any dystopia. Growth was visible under the conjunctiva in inferior fornix. Lesion was excised through trans-conjunctival approach. A brownish well encapsulated mass measuring 3.0 x 2.8 cm was extracted en bloc. (Fig. 15) Post operative recovery was uneventful and patient’s visual acuity improved to 6/9 three weeks post operatively (Fig. 16).

DISCUSSION
Cavernous haemangiomas are the most common benign, non-infiltrative neoplasms of the orbit and represents 9.5 to 15% of the primary expansive lesions of the orbit. They usually grow slowly between the extrinsic muscles and present as a mass effect on the globe. It is a vascular malformation characterized by the presence of sinusoids with fine walls, which
contain an accumulation of blood with no apparent arterial or venous inflow. Its incidence peaks between the ages of 40 – 50 years and women are affected more.

A slowly progressive painless proptosis is the typical presenting symptom. Impaired extraocular movements and visual function are seen with large lesions and with lesions located at the orbital apex. Most cavernous haemangiomas are located within the intraconal space. Nearly all patients with cavernous haemangioma can be correctly diagnosed by preoperative radiological studies. Orbital CT scan is the single most useful diagnostic test and shows a well circumscribed lesion with no osseous involvement. In our study, CT scan showed very well defined smooth mass, which enhanced with intravenous contrast.

Surgical treatment is recommended for optic nerve compression as evidenced by visual field defects, optic nerve swelling and pallor. Other indications include diplopia and bothersome cosmesis. Several therapeutic modalities and surgical approaches have been described, in order to preserve the normal orbital structures. Most approaches used to remove orbital tumours typically include bone removal (orbitotomies) with or without craniotomies. Complete excision is generally accomplished without recurrences as the tumour is well encapsulated with relatively few feeding vessels. Lateral orbitotomy has been widely employed for the removal of orbital tumors, being used in large sized hemangiomas, especially those located in the lateral compartment of the orbit and orbital apex.

Transconjunctival approach can be used successfully for anterior as well as retrobulbar intraconal cavernous hemangiomas. Anterior orbitotomy is useful in many cases, without significant complications and warrants more favourable consideration if combined with the use of cryoprobe and surgical microscope. Postero-inferior orbitotomy through the maxillary sinus can be used in small, well demarcated lesions in the posterior and inferior orbit near the apex. Endoscopic trans-ethmoidal approach of the orbit is a minimally invasive surgery for retro-bulbar orbital neoplasm, leading to excellent cosmetic results with less bleeding. The medial wall of the orbit, the orbital apex, and the optic canal can be exposed through a middle mental antrostomy, an anterior and posterior ethmoidectomy, and a sphenoidotomy. Lateral suprabrow, trans-nasal and the combined perional and orbitozygomatic approach are employed for removal of tumors affecting the optic canal.

Transcranial approaches offer an excellent surgical exposure and a good cosmetic outcome and should be considered for big lesions located superiorly or medially to the optic nerve, especially those involving the apex. Complications of surgery include ptosis, impairment of ocular movements, diplopia due to mechanical or vascular trauma and visual disturbances.

Careful preoperative workup including plan for minimal invasive approach based on CT scan offered excellent exposure and a rewarding cosmetic result in our cases and that this technique might also be considered for larger lesions Figure 1 – 4 show pre- and post-operative picture of one of cases with technique of en bloc removal of these tumours. Avoiding large incisions and osteotomies resulted in early rehabilitation and decreased morbidity. Additionally, there were no risks of cerebrospinal fluid leakage or significant blood loss during the procedure, no bone removal or reconstruction was required and no postoperative enophthalmos or temporal muscle atrophy was encountered.

CONCLUSION
Cryo-assisted minimally invasive surgical excision offers an exciting approach for management orbital cavernous hemangiomas. With improvement of neuroimaging techniques, it is possible to obtain a correct pre-operative diagnosis in almost every case of vascular orbital lesions. This can lead to excellent cosmetic results with complete functional recovery and minimal morbidity.

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