Abstract:
Hemangiopericytomas are tumors whose origin is still being debated. Their occurrence is rare and they are reported as case reports. Their occurrence in orbit is very few. They are malignant tumors capable of recurrence and metastasis. Their radiological features are identical to meningiomas. Transcranial transorbital approach is particularly useful in selected cases like a tumor arising from roof of orbit. Complete excision with wide local clearance is necessary to prevent recurrence.

Keyword: Orbit, orbital tumor, hemangiopericytoma.

Introduction:
Hemangiopericytomas are rare tumors occurring commonly in adults. It can affect any part of the body including the soft tissues of face and orbit. Radiological appearance is very similar to meningioma. For orbital tumors transcranial approach is excellent for exposure with minimal complications. For hemangiopericytoma complete removal with wide local clearance is essential to prevent recurrence and metastasis.

Case report:
A 27 year old lady presented with complaints of progressive swelling of left eye for 3 years. Over last 6 months there was protrusion of eye down and out. She felt that the eye became more prominent on bending down and on activities of exertion like coughing, sneezing. She had no diminished vision, double vision, watering of eyes or congestion. There was no facial pain, numbness or weakness over face. On examination, she was conscious and oriented. Her higher mental functions were normal. First cranial nerve was normal. Visual acuity, field, and color vision were normal on examination. Both fundi were normal. Both pupils were equal and reacting to light normally. On primary gaze, there was minimal deviation of the left eye inferiorly and laterally. The extraocular movements were not restricted inspite of the exotropia. Sensation over the face was normal. Other lower cranial nerves, spinomotor system and sensation were normal. Cerebellar and autonomic functions were
normal. Radiological investigations revealed a 2X2X2 cm sized left sided intraorbital extraconal lesion arising from the superomedial aspect of orbit displacing extraocular muscles and eyeball inferolaterally. It was uniformly hyperdense on plain CT. The lesion was isointense on both T1 and T2 weighted images and uniformly enhancing on postcontrast images.

Fig 1: Non contrast CT coronal images showing intraorbital extraconal tumor arising superomedially displacing the extraocular muscles and eyeball laterally and inferolaterally. Incidentally chronic maxillary sinusitis was also noted.

Fig 2: MR images of the tumor. (a)T1 axial, (b)T1 coronal (c)T2 coronal, (d)contrast axial and (e&f)sagittal images showing a well circumscribed 2x2 cm sized lesion near roof of orbit and displacing the orbital contents inferolaterally. The lesion was isointense on T1 and T2 weighted images and uniformly enhancing on contrast administration.
The patient underwent left transcranial orbitotomy and removal of tumor. The transcranial orbitotomy uses a frontal craniotomy with removal of a portion of the orbital roof to expose the orbital apex or superior orbit. In most cases, the supraorbital rim over the involved side is removed en bloc with the frontal bone flap. The anterior one-half or two-thirds of the orbital roof breaks free with removal of the rim and frontal bone flap, and the remaining posterior portion of the roof can be removed with rongeurs or air drill. The transfrontal approach was first described by Jones in 1970[1]. Jane and colleagues proposed the current technique in 1982[2]. Refinements have been discussed by Maroon and Kennerdell[3] and Housepian[4]. A coronal skin incision was made 2 cm behind the hairline. A frontal scalp flap was raised, elevating pericranium off the frontal bone. The supraorbital nerve was identified and, as it exited the skull through a foramen, it was chiseled out, mobilizing the nerve. The subperiosteal plane was followed over the superior orbital rim, elevating periorbita from the orbital roof and lateral orbital wall. The temporalis muscle was dissected anteriorly out of the temporalis fossa to expose the temporal bone. After these soft tissues have been separated from the bone, a burrhole osteoplastic bone flap was raised. The dura was freed from the undersurface of the bone flap and was elevated superiorly, and the orbital roof was cracked off. The frontal bone, orbital roof, and supraorbital rim were broken off in one piece. The brain was retracted superiorly, and the remaining orbital roof was removed with bone rongeur. After removal of the bony roof, the periorbita was visible. The tumor was found to be well defined and infiltrating the roof of orbit. It was firm, vascular and pinkish red in color. It was free from extraocular muscles. The tumor was bulging into the defect. Dura was retracted and tumor exposed. Intratumoral excision and complete removal of the tumor was done. Pericranium was introduced between dura and peri-orbita and orbital roof was reconstructed. Frontal bone was replaced, and scalp flap sutured back.

Fig 3: (a) Head fixed over Mayfield frame and incision line marked. (b) Bicoronal scalp flap elevated and Left frontal craniotomy done.
Fig 4: (a) Dura retracted and excision of orbital roof done and orbitotomy completed. (b) Tumor exposed. Bone and dura are seen in the field.

Fig 5: (a) Intratumoral excision done (b) After complete excision of tumor. Tumor bed is seen and hemostasis achieved with surgicel.

Pathology Microscopic examination showed neoplasm composed of numerous capillaries of varying sizes some of which showed hyalinized vessel wall separated by dense sheets of round to spindle shaped cells with compressed vasculature. Some foci showed dilated vascular spaces with adjacent foci showing sclerosed blood vessels with endothelial hyperplasia. A diagnosis of hemangiopericytoma was made.
Fig 6: Histopathological Photographs of the tumor.
(a) Low power image showing highly cellular tumor with regions of cystic changes and necrosis. The vessels appear compressed.
(b) staghorn appearance.
(c) Tumor showing sclerosed vessels with endothelial hyperplasia.
(d) Hyalinized vessel wall separated by dense sheets of spindle shaped cells with compressed vasculature.

Follow up:
Patient recovered well in the postoperative period. There was minimal ptosis of upper eyelid due to postoperative edema which resolved completely. Ocular movements were full and vision was normal. The patient is on regular follow up and is doing well. There is no tumor recurrence.

Fig 7: Postoperative images of the patient.
(a) POD-7. Bicoronal incision sc is seen. (b & c) No restriction of extraocular movement in the postoperative period. There was minimal drooping of eyelids which resolved soon.
Fig 8: Postoperative images. (a) Roentgenogram showing left frontal craniotomy bone flap. The orbital defect can also be made out. (b) CT images of orbit showing the postsurgical defect.

Fig 9: (a,b,c,d) Axial CT images of orbit 1 year later showing no tumor recurrence.

Discussion:
Hemangiopericytomas are rare occurrences in the orbit. As like most of the extraconal lesions they arise from the superior part of the orbit. Though our patient had discomfort in her eye for 2 years, she sought medical help only after complaints of proptosis. The volume of orbit is 30 cm$^3$ and that of the globe is 7 cm$^3$. The lesion was of a significant size (~4 cm$^3$), causing mild proptosis. She did not have any other complaints. Hemangiopericytomas can be locally aggressive tumors, but in our case it was well defined and did not breach the periorbita and lying extraconally. As there was no invasion into the extraocular muscles or nerves supplying them she had no restriction of eye movements [5]. Few case reports of intraorbital tumors of various causes with similar history and course were recorded [6-8].

Hemangiopericytomas develop from the soft tissues of the orbit, nasopharynx, tongue, nose and paranasal sinuses, maxilla and mandible, pharynx, auricle and neck, and at skull base.
Macroscopically hemangiopericytomas are attached to the dura either in a broad based manner or by a narrow pedicle. Some may have a dural tail. Most tumors are greater than 4 cm in diameter and are multilobular. Their consistency varies from soft to firm and is highly vascular. Tumor may not be well encapsulated. There may be signs of invasion. Bony erosion is common [9].

Hemangiopericytomas are cellular, malignant tumors. They are composed of a uniform population of medium sized elongated or polygonal cells with indistinct cell membranes. The nuclei are hyperchromatic and oval to carrot shaped. The cells are tightly packed in solid masses or sheets, often arranged around branching staghorn like blood vessels. Blood vessels vary from gaping sinusoids to collapsed capillaries. They are rich in reticulin, the meshwork being produced by the tumor cells and corresponding to basal lamina, outlining the vascular channels and surrounding the individual cells. Some may have areas of anaplasia, crowding of hyperchromatic nuclei and brisk mitotic activity. Hemangiopericytomas are positive for vimentin, and negative for EMA. They are CD-34 positive but not consistent as in solitary fibrous tumors. They are negative for GFAP and S-100. They are positive for factor XIII and HLA-DR (expressed in normal pericytes). Extracellular basement membrane surrounds most cells on electron microscopic examination. Cytoplasmic intermediate filaments associated with dense bodies are demonstrated. Dilated rough endoplasmic reticulum and pinocytic vesicles are also demonstrated. There are conflicting views if this is a variant of meningioma or a separate entity as the histological observation of papillary pattern in both. But ultrastructural, immunocytochemical, molecular genetic and tissue culture evidences now favor the view that they are different. This tumor resembles smooth muscle, endothelium and fibroblast more than meningothelium. Molecular analysis does not reveal abnormalities of chromosome 22, mutations of NF-2 gene. Homozygous deletions of CDKN2a/p16 gene occur in hemangiopericytoma but not in meningioma. The treatment of choice is wide local excision of the tumor along with the infiltrated tissues [10]. It may not be always possible to resect the tumor completely. Preoperative Onyx embolization [11] or radiotherapy [12] may facilitate the complete removal of the tumor. Intraoperatively, external carotid artery occlusion [13] may limit the blood flow and reduce blood loss. As the tumor is locally invasive [14] it has a tendency to recur [15] and metastasize [16]. There is a case report of tumor recurrence 33 years after initial removal [17]. Postoperative radiotherapy is found to decrease the rate of tumor recurrence [18]. Conventional or proton beam irradiation has been advised for recurrence.

Reference:


