

Orbit Conference: 9.28.15

The Not So Incidental Finding



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31 yo female with recurrent headaches

- CC: Recurrent headaches and sparkles in her vision
- HPI: 31 yo F was being evaluated for recurrent headaches and visual disturbances that increased with exercise in the setting of a prior traumatic brain injury after a snowmobile accident.

- PMH: von Willebrand disease, migraine headaches, TBI, depression, hernia repair, and prior knee surgery
- POH: None
- FH: No known ocular disease.
- SH: Lives in Utah. Former smoker.
- Meds: Multivitamin, citalopram
- Allergies: NKDA

Eye Exam

- VA: 20/15 OU
- Pupils: reactive, no RAPD
- EOM: normal motility OU
 - Alignment: ortho
- Confrontational fields: full
- No proptosis
- Color vision: normal OU

Additional Tests

- Multiple Humphrey visual fields were performed and normal
- MRI orbits wwo contrast: to be presented by Dr. H. Christian Davidson
 - Mass within left orbit

By the time she presented to the
Moran...

- 1.5 mm proptosis OS evident on exam

Differential Diagnosis

- Capillary hemangioma
 - Hemangiopericytoma
 - Lymphangioma
 - Cavernous hemangioma
 - Orbital metastasis
 - Orbital lymphoma
- **Will discuss radiographic findings later

Neuroradiology by Dr. Davidson

**January
2015**



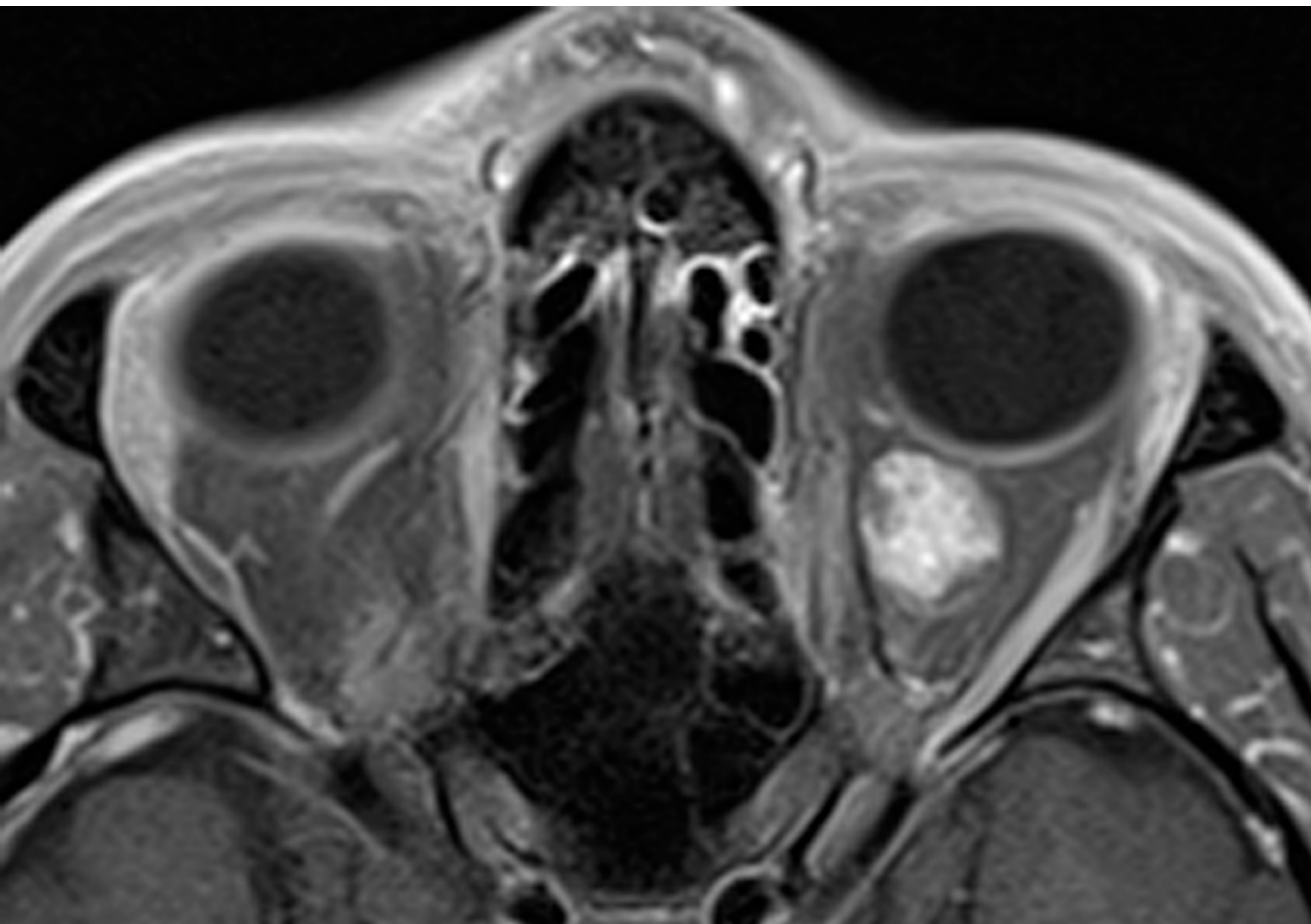
MRI orbits without contrast

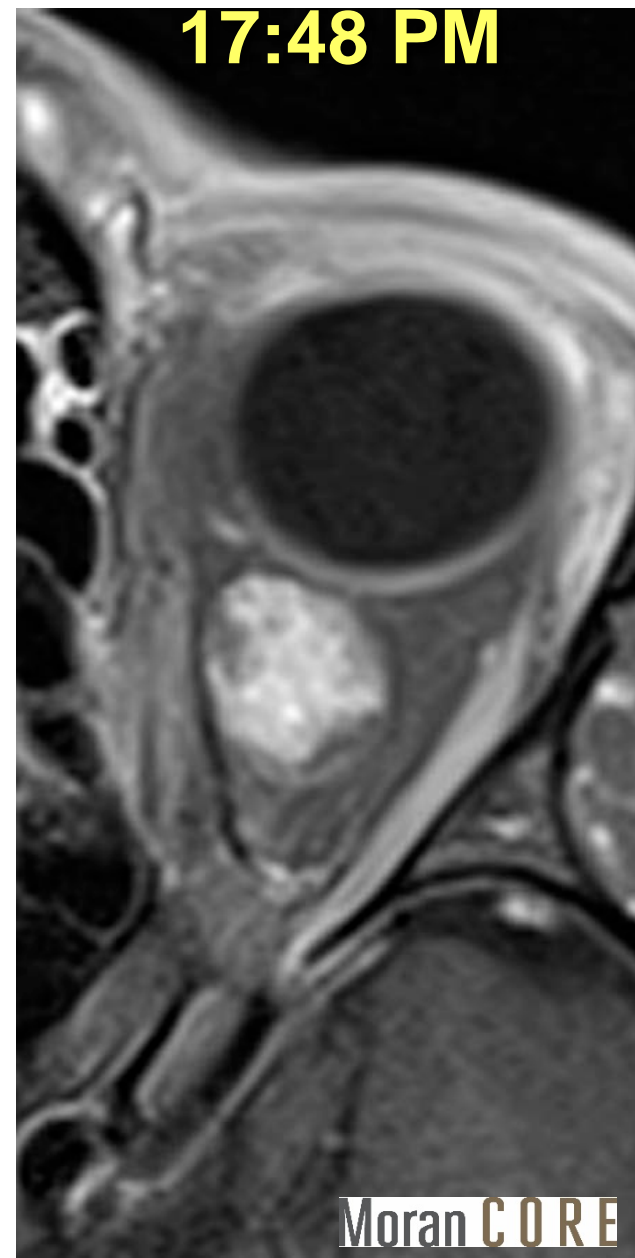
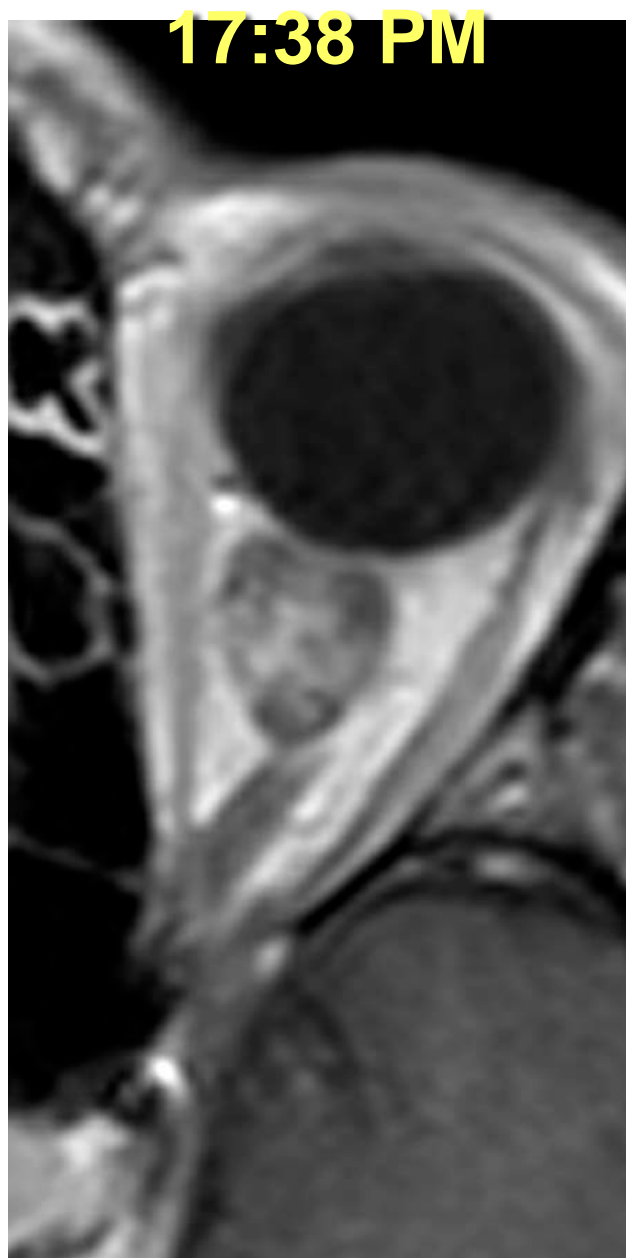
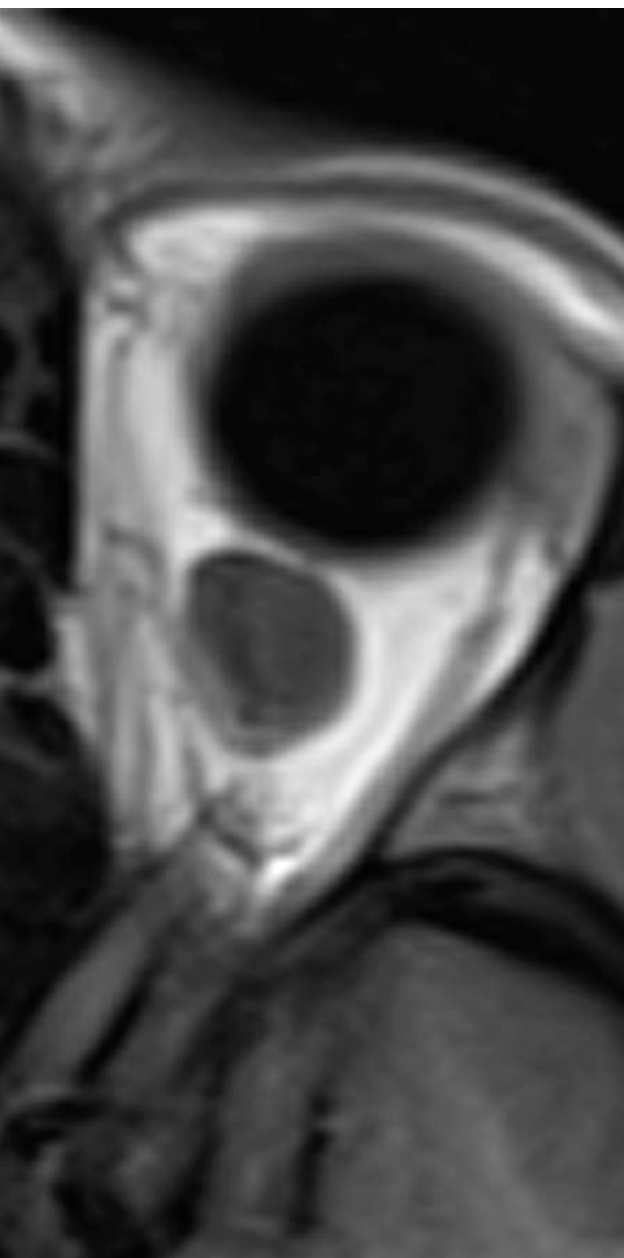


MRI orbits without contrast



MRI orbits without contrast

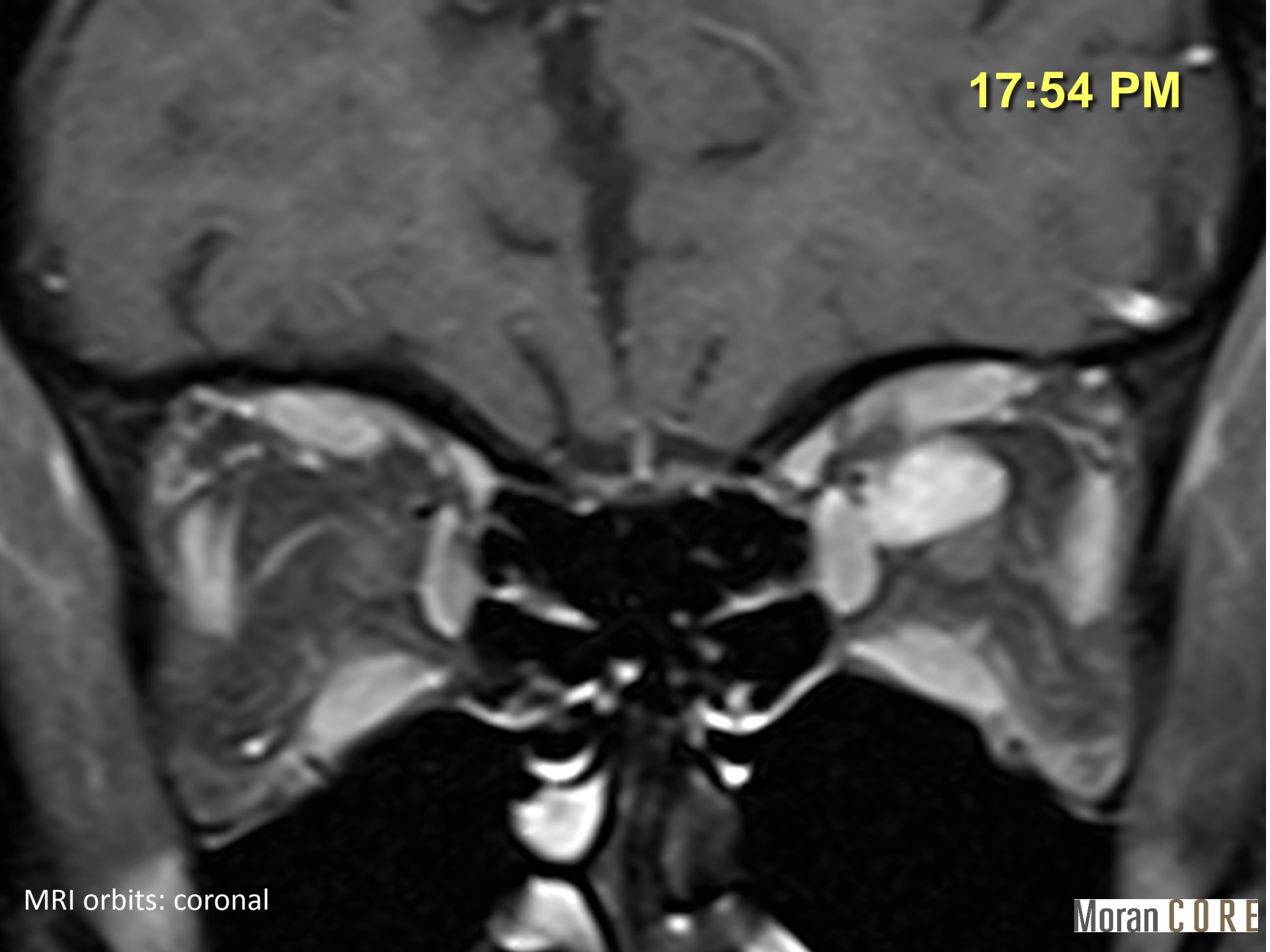




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MRI orbits with contrast: axial cuts

17:54 PM

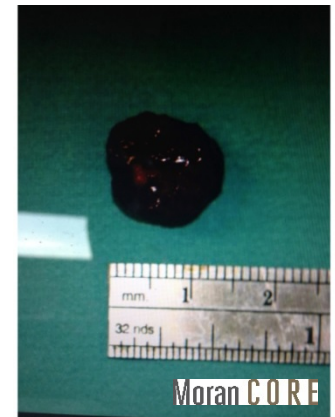


MRI orbits: coronal

Moran CORE

Excision

- Due to optic disk swelling and some visual field constriction, the left intra-orbital lesion was excised on 6.5.15 using an upper eyelid crease approach by Dr. Patel and sent for pathology.
- Histopathology to be presented by Dr. Guan



Histopathology

Dr. Jun Guan

Cavernous Hemangioma Histology

- Well-circumscribed and encapsulated in fibrous pseudocapsule
 - Can sometimes incorporate local structures into capsule
- Vascular malformation (versus benign neoplasm)
- Large vascular spaces lined by endothelium (CD31 positive)
- Thick septae of fibrous tissue
- Slow flow lesion
 - Separated serum
 - Thromboses

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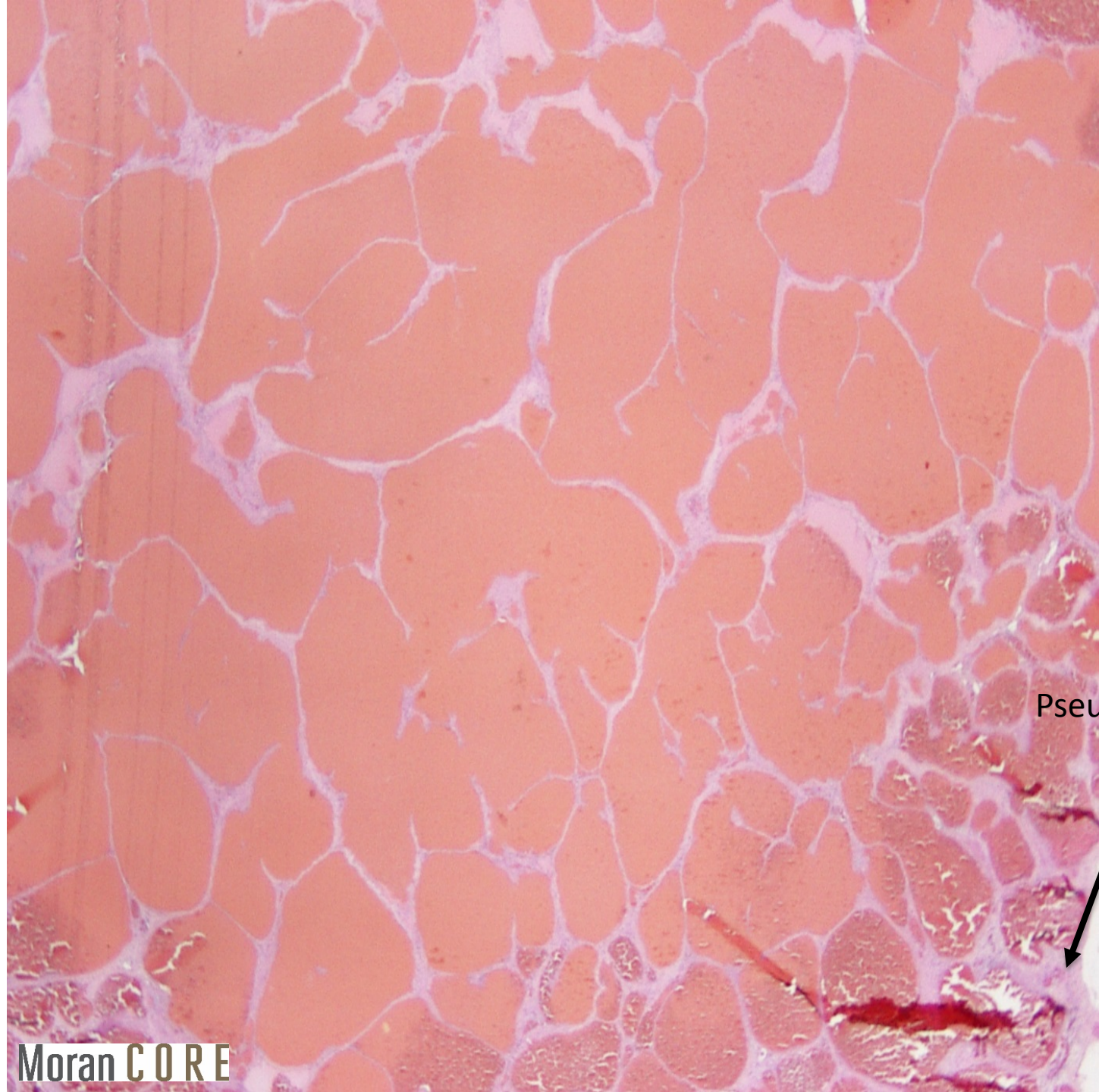
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OPHTHALMOLOGY



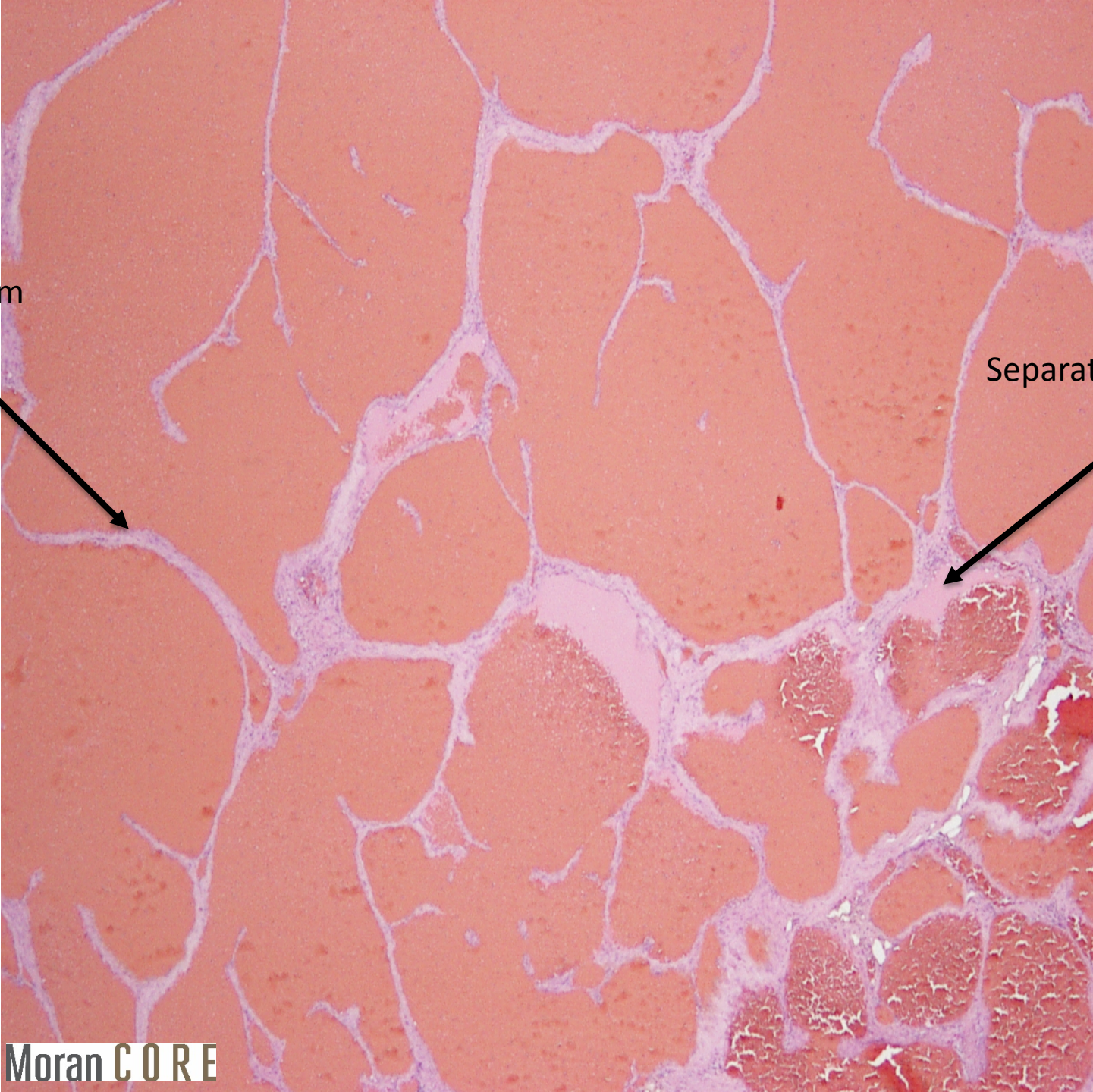
Gross cross section
of specimen



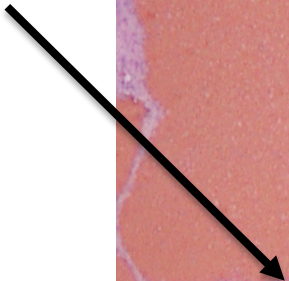
Pseudocapsule

H&E
20x

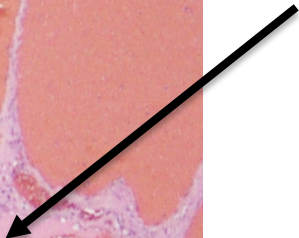
Moran C O R E



Endothelium



Separated serum



H&E
40x



Endothelium



Moran CORE

H&E
100x



Cavernous Malformation Mechanism

- Slow flow venous system
 - “Thrombosis and recanalisation”
- Fibrin clot formation
 - Layer of endothelium forms over matrix
 - Proangiogenic factors produced by activated platelets, endothelium, and PMNs
 - Endothelial growth
 - » Natural clefts in thrombus
 - New vascular channels form along fault lines
 - Monocytes recruited
 - Active neovascularization
 - Fibroblastic components and collagen deposition in following 4-8 weeks
 - Myofibroblastic infiltration in later stages

Vascular Lesion Differential Diagnoses

- Hemangiopericytoma
 - Can be malignant
 - Reticulin stain
- Lymphangioma
 - Unencapsulated
 - Infiltrative growth pattern
 - Lymphoid aggregates
- Capillary hemangioma
 - Plexus of capillary caliber vessels
 - GLUT-1 positive

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Cavernous hemangiomas

Epidemiology

- 4.3% of orbital neoplasms
- Male to female ratio debatable but historically felt to be predominately in women
- Usually presents in the 3rd to 5th decade of life
- Imaging findings: well-circumscribed intraconal mass, 90% can be diagnosed pre-operatively
- No factors predictive of lesion growth
- Hormones (i.e. pregnancy and menopause) may influence size of lesion and lesions have been shown to express progesterone receptors

Symptoms

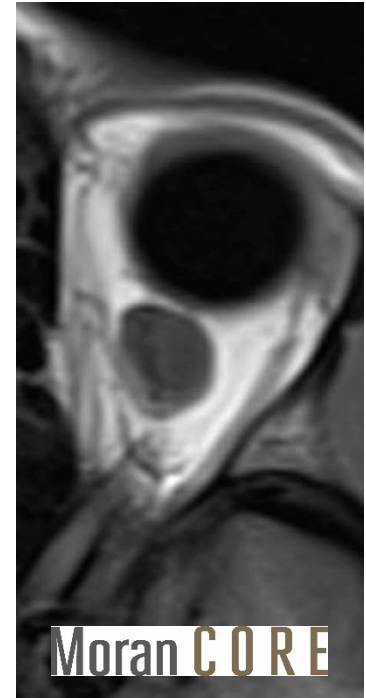
- Eyeball protrusion (90%)
 - Visual impairment (65%)
 - Double vision (20%)
 - Local Pain (18%)
 - Headache (12%)
 - Eyelid fullness/swelling (5%)
- **Usually unilateral but bilateral cases have been reported

Clinical Signs

- Unilateral proptosis
- Strabismus
- Choroidal folds
- Relative afferent pupillary defect
- Visual field deficits
- Lagophthalmos
- Elevated intraocular pressure
- Optic disk edema

Radiographic Findings

- 80% located within the intraconal compartment
- Usually round or oval in cross section and well circumscribed
- Frequently abut globe but do not deform it
- Maybe deformed by globe
- Hypoattenuating compared to muscle on CT



Treatment

- Observation
- Surgical excision if threatening vision

Issues with surgical resection

- Access to the orbital apex
- Morbidity associated with large resections (increased operative time, post-operative pain, recovery time)
- Surgical scars

History of orbital resections

- Lateral orbitotomy was first described by Dr. Rudolf Ulrich Kronlein in 1889 for dermoid resections that would become the basis for orbital decompressions
- Dr. Kronlein was a Swiss surgeon and chaired the Surgery Department at the University of Zurich
- First to perform an appendectomy (boy died 2 days later, however)
- Pioneer in neurosurgical interventions
- Lowered post-operative mortality to 3% by decreasing depth and length of anesthesia and using aseptic technique

Hildebrandt et al., 2012.

Revisions of Lateral Orbitotomy

- Then modified by Berke by extending canthotomy in 1953
- Stallard then further altered the approach by moving into lateral brow
- Access to the intraconal and extraconal space has gone through multiple iterations since that time

What is the neurosurgical approach to
resection???

Orbit viewed in clock hours to help guide approach

1-5 o'clock: endoscopic endonasal vs. medial orbitotomy approach

6-8 o'clock: Lateral orbitotomy with zygomatic osteotomy

8-10 o'clock: lateral orbitotomy

10-1 o'clock: Frontotemporal craniotomy/orbitotomy

****For further description of clock hour approach, please see Paluzzi et al., "Round-the-Clock" Surgical Access to the Orbit. J. Neurol Surg, 2015**

Newest techniques: Goal of decreased morbidity with equal or better access to orbit

- Gamma knife ablation described by Thompson et al., 2000
- Stereotactic-guided transcranial cryoextraction described by *Papalkar et al., 2005*.
- Endoscopic transnasal approach described by Stamm et al., 2005.
- Endoscopic transnasal transphenoidal approach by Chen et al., 2010
- Endoscopic transthemoidal with or without medial rectus detachment described in *Wencal et al., 2013*.
- Endoscopic transnasal septotomy to access contralateral orbital apex described in *Murray et al., 2013*.
- Excision without bony marginotomy described by Dr. *Goldberg et al., 2014* from Jules Stein to decrease post-operative morbidity in causes of benign lesions.

Discussion

- Consider cavernous hemangiomas in both men and women with painless unilateral proptosis in the 3rd to 5th decade of life
- Use imaging to help aide in diagnosis
 - Neuro-radiologists
- Multiple emerging ways to access orbital tumors
 - Balance visualization of tumor with morbidity
 - Use and enhance techniques of those before us

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