PROPTOSIS in ADULTS

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ETIOLOGY

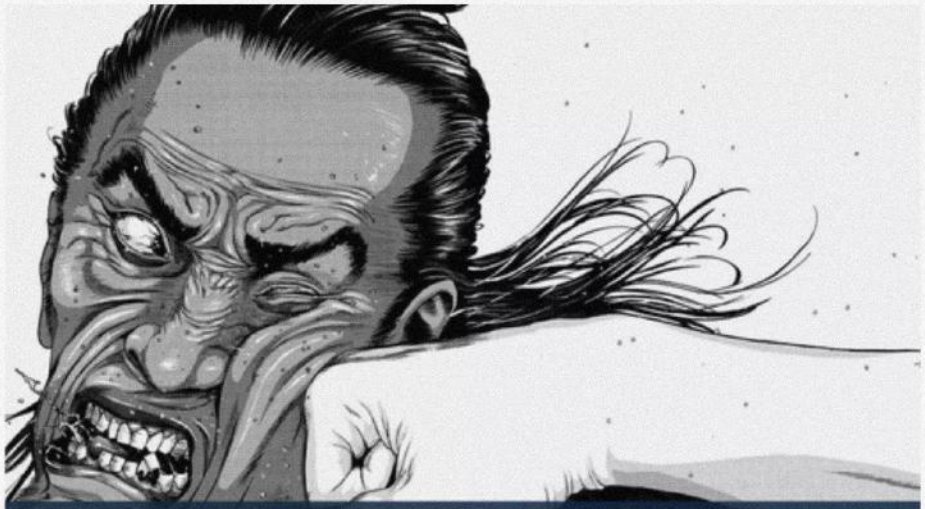
- *Endocrine*
- *Trauma*
- *Vascular*
- *Infective*
- *Inflammatory*
- *Neoplastic*
- *Others*

CATEGORY	SPECIFIC CONDITIONS
Endocrine	Thyroid eye disease
Trauma	Facial fracture, soft tissue swelling, retro bulbar hemorrhage
Vascular	Carotid cavernous fistula
	Cavernous Haemangioma
	Cavernous sinus thrombosis
Infective	Orbital cellulitis
	Mucormycosis
Inflammatory	IOID/Dacryoadenitis/Orbital myositis
	Tolosa hunt syndrome / Wegner granulomatosis
	Sarcoidosis
	Churg Strauss syndrome
Other	Paget disease/ fibrous dysplasia
	Langerhans cell Histiocytosis

NEOPLASTIC DISORDERS

1.	Lacrimal gland tumors
2.	Meningioma
3.	Optic nerve Glioma
4.	Schwannoma
5.	Neurofibroma
6.	Lymphoma
7.	Sino-nasal tumors
8.	Ossifying fibroma
9.	Orbital Osteoma
10.	Neuroblastoma
11.	Haemangioblastoma
12.	Acute leukemia
13.	Myeloid sarcoma
14.	Metastasis

ORBITAL TRAUMA



- Orbital trauma can damage the facial bones and adjacent soft tissues. Fractures may be associated with injuries to orbital contents, intracranial structures, and paranasal sinuses.

Assessment of Orbital Trauma

- Check ABC's
- History of mechanism of injury
- Signs & Symptoms:

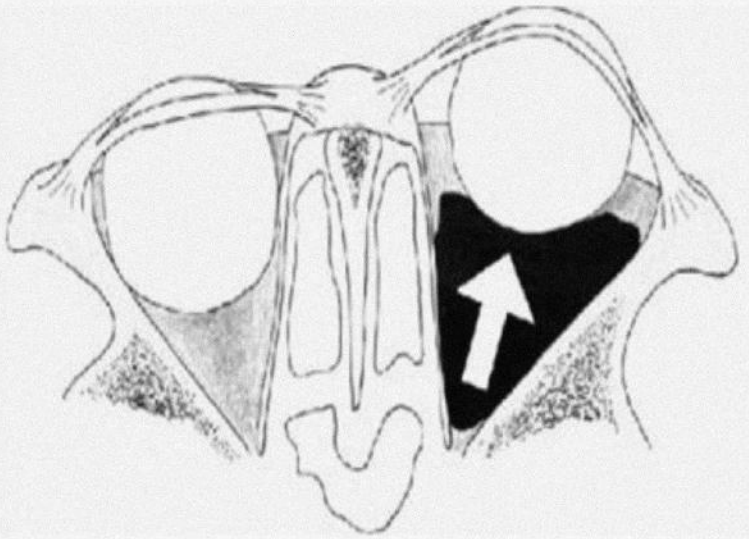
  Varies but look for s/s suggestive of severe injury, including diplopia, visual loss, ptosis, lid laceration, subconjunctival hemorrhage, periorbital ecchymosis or infraorbital anesthesia
- Ophthalmologic examination:

  visual acuity, pupil reaction, motility, sensation, globe position, lid function, integrity of globe and fundoscopy
- Imaging…

TYPES OF ORBITAL TRAUMA

- Blow out orbital floor fracture
- Blow out medial wall fracture
- Roof fracture
- Lateral wall fracture

•*Proptosis*



Without pulsation

-Orbital hemorrhage

-Emphysema

With pulsation

-High flow CCF

Orbital roof fracture with secondary herniation of anterior cranial fossa contents

Retro bulbar Hemorrhage (RBH)

- Rapidly progressive, sight-threatening emergency that results in an accumulation of blood in the retro bulbar space due to a rupture of an artery /vein
- The pressure increases and leads to the damage of the structures
- Treatment: blood evacuation, canthotomy, cantholysis

VASCULAR LESIONS

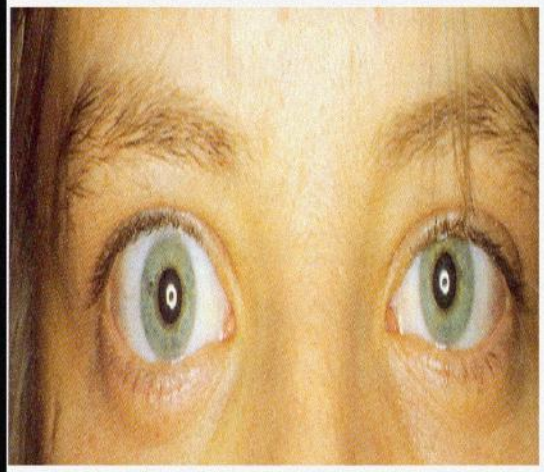
1. Cavernous Haemangioma
2. Orbital venous anomalies
3. Carotid-cavernous fistula
4. Cavernous sinus thrombosis

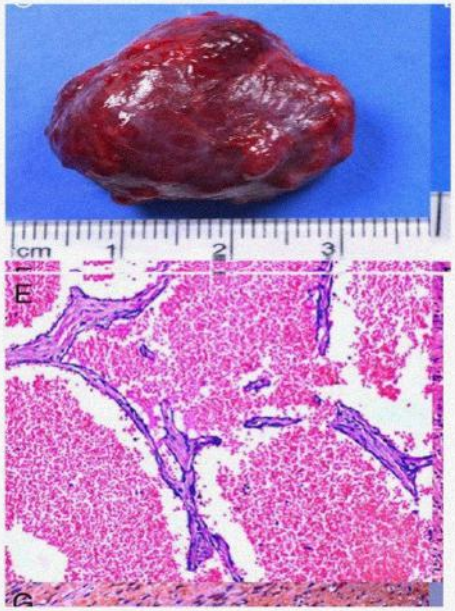
CAVERNOUS HEMANGIOMA

- It is the most common benign orbital tumor in adults
- Frequently occurs within the muscle cone
- Female preponderance is 70%
- 4^{th} to 5^{th} decades with slowly progressive unilateral proptosis
- May compress the optic nerve at orbital apex
- Gaze evoked temporary blurring of vision is an occasional feature

Treatment Surgical excision

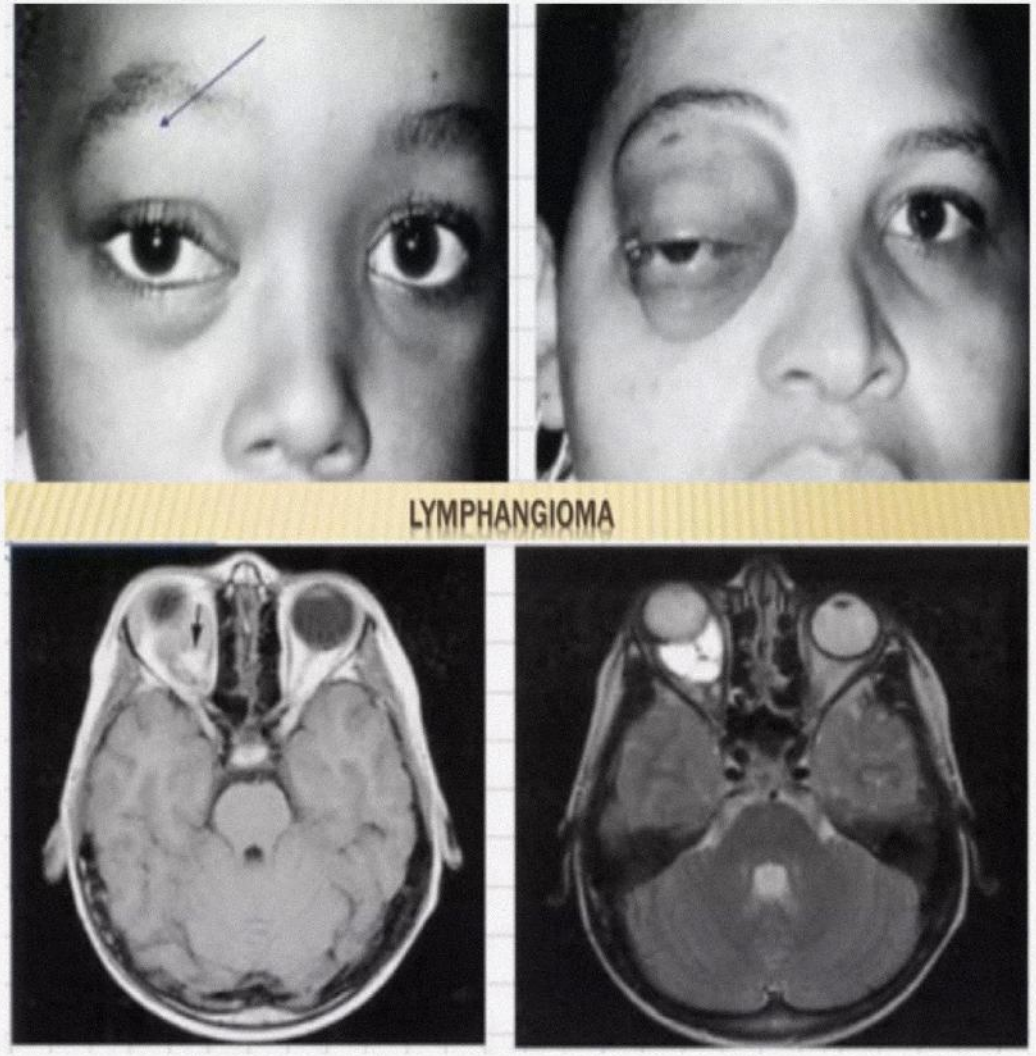
- **Benign, noninfiltrative, slowly progressive vascular neoplasm composed of endothelial-lined spaces surrounded by a well-delineated fibrous capsule.**





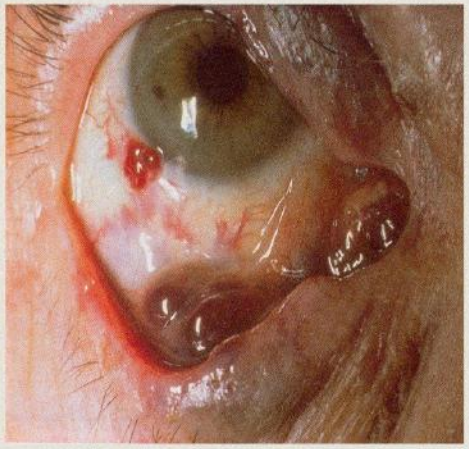
Orbital Varices/OLM

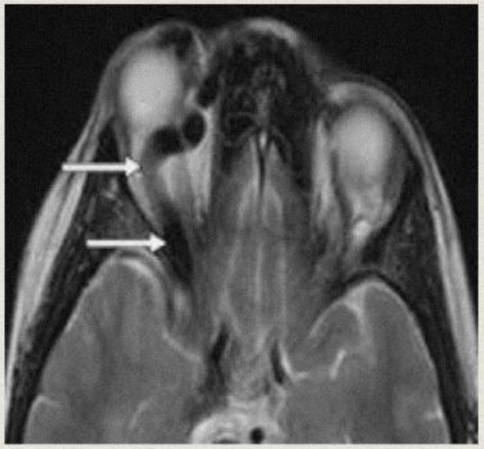
- Venous malformations of the orbit caused by vascular dysgenesis
- They consist of a plexus of thin-walled distensible low flow vein-like vessels that are commonly intrinsic to the normal circulation
- They are considered to be hamartomatous, often located in the intra conal area of orbital apex
- They cause intermittent proptosis when pressure increase due to certain maneuvers



Treatment

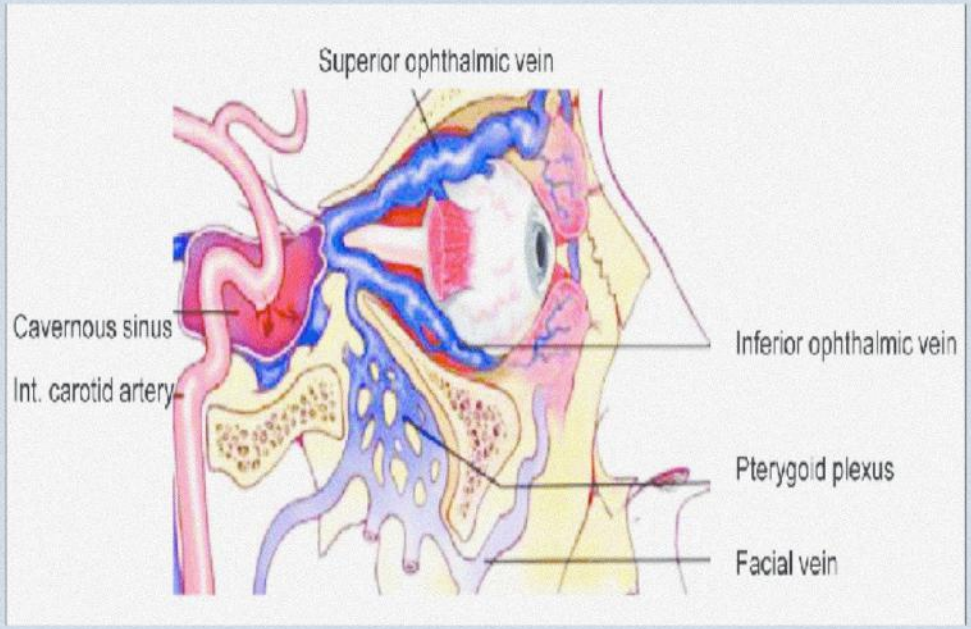
- **Excision, Aspiration of chocolate cysts, Sclerosing agents (Bleomycin)**
- ***Indications*** **for treatment are proptosis,hemorrhage,thrombosis**





Carotid-cavernous fistula

- CCF is the result of an abnormal vascular connection between the internal carotid artery (ICA) or external carotid artery (ECA) and the venous channels of the cavernous sinus leading to a compromise in venous drainage from the eye
- Direct type…….Trauma/ruptured anuerysm
- Indirect type……..Hypertensive old ladies /postmenopausal/insidious



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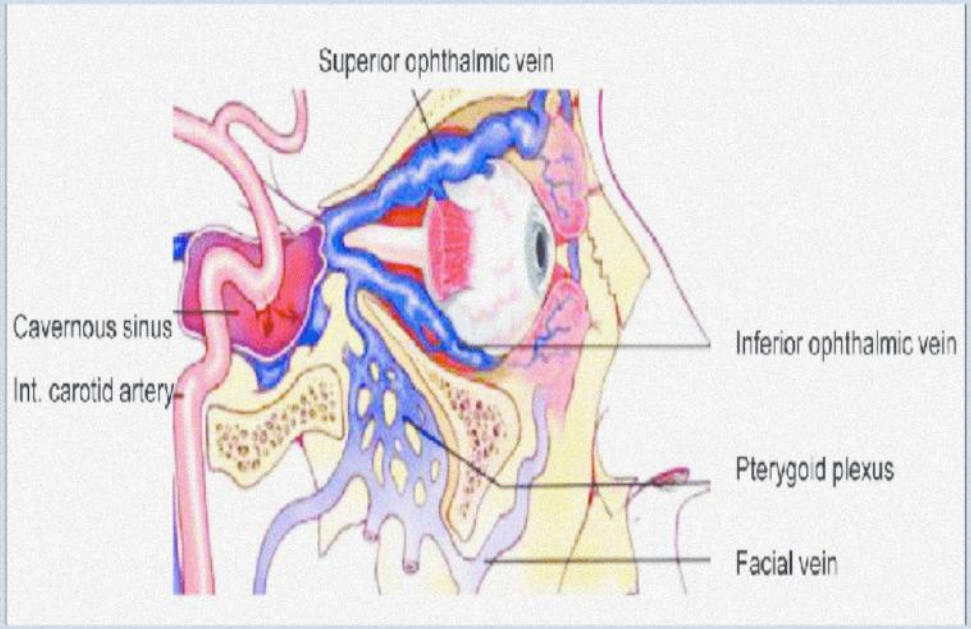
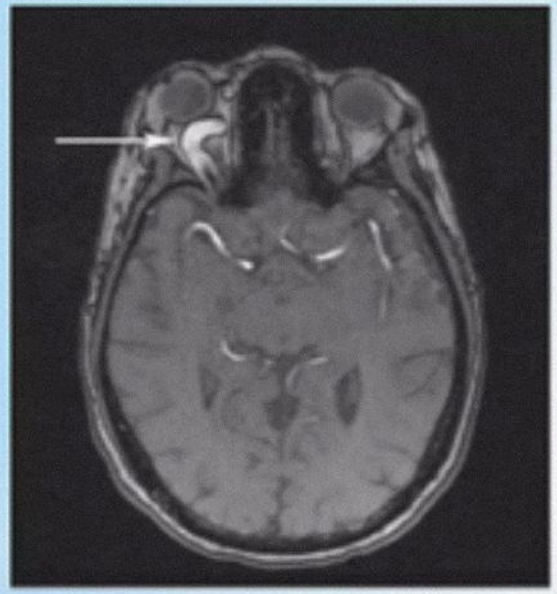


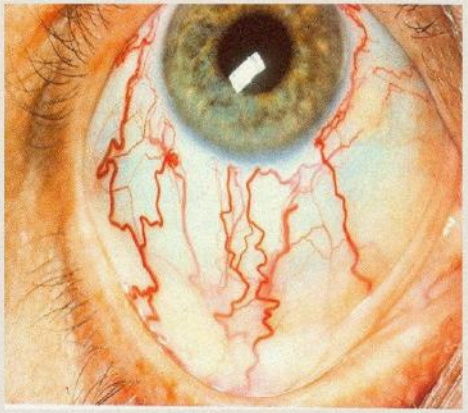
MRI



Magnetic Resonance Angiogram (MRA) image demonstrating an enlarged superior ophthalmic vein

MRA demonstrating a right carotid cavernous fistul

- Congestion, headaches, cranial nerve palsies, intermittent proptosis
- Treatment -------endovascular embolization



LOW FLOW CCF

HIGH FLOW CCF

CAVERNOUS SINUS THROMBOSIS

- **Anterior -** extends into medial end of superior orbital fissure.
- **Posterior -** upto apex of petrous temporal bone.
- **Medial –** Pitutary above and sphenoid below
- **Lateral –** temporal lobe and uncus

Causes

Septic CST

- Infectious

Aseptic CST

- Trauma
- Post surgery

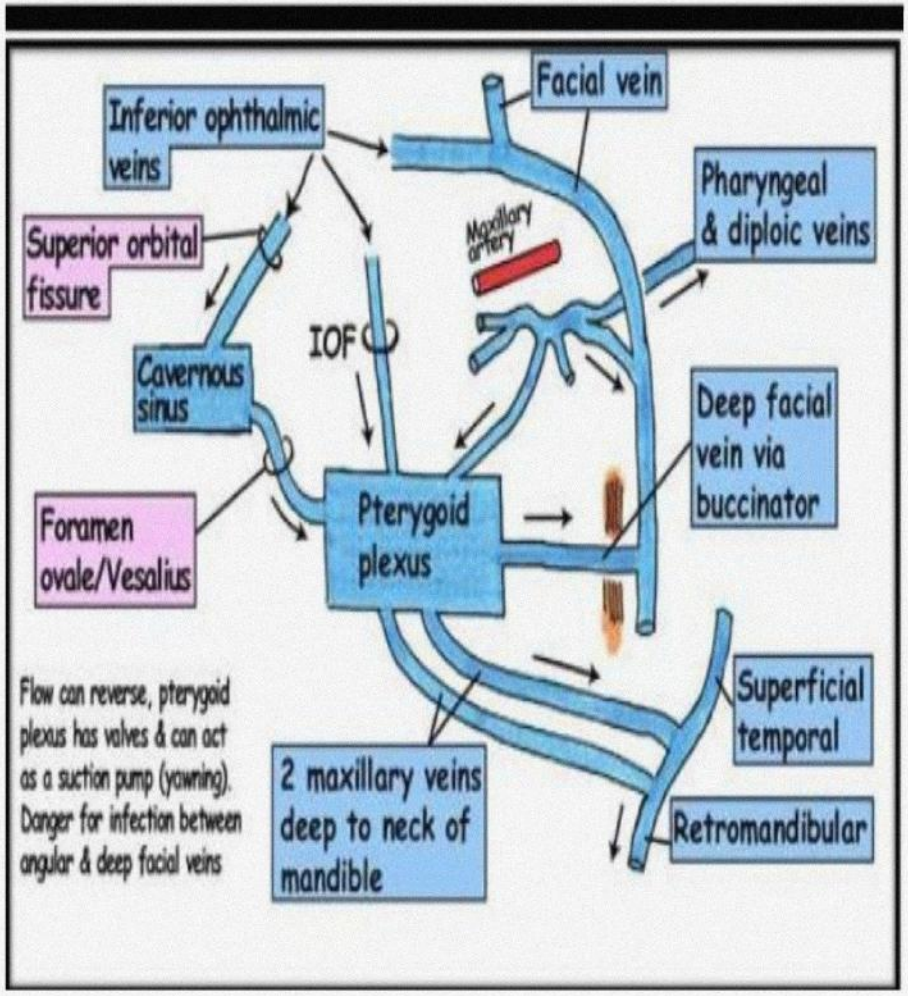
Rhinoplasty
Base of skull
Tooth extraction

- Hematologic
- Malignancy

Nasopharyngeal Ca.

- Dehydration

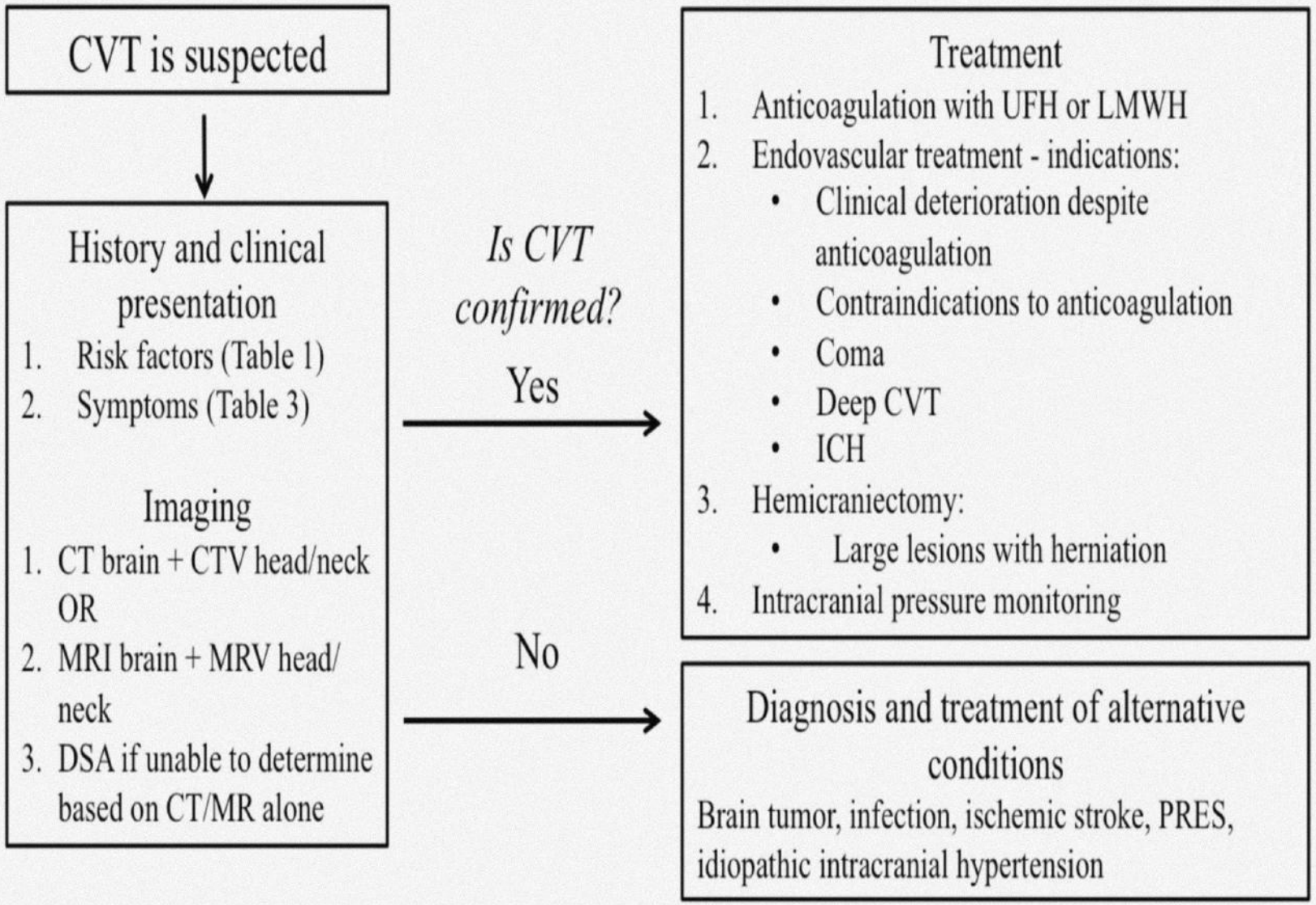
Venous connections of cavernous sinus



Venous obstruction

- Proptosis (first oedema & chemosis)
- Oedema of eyelids and bridge of nose
- Dilatation and tortuosity of retinal veins
- Retinal hemorrhages
- Involvement of the contralateral eye - (48 hours)
- When pterygoid plexus is occluded along with sinus, - oedema of the pharynx or tonsil

Septic Cerebral Venous Thrombosis

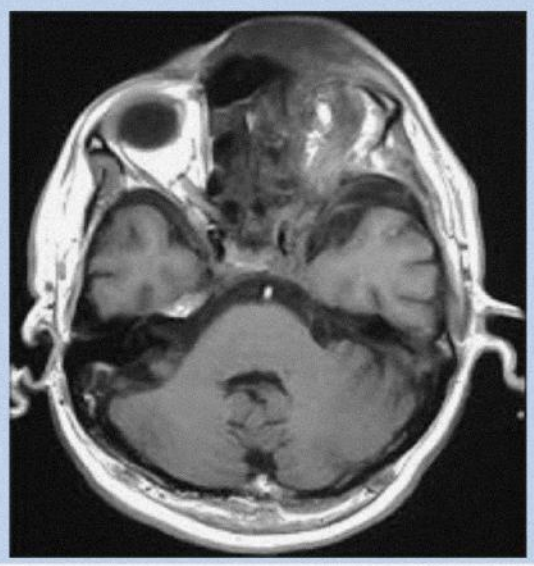

CVT is suspected
History and clinical presentation
1. Risk factors (Table 1)
2. Symptoms (Table 3)
Imaging
1. CT brain + CTV head/neck OR
2. MRI brain + MRV head/ neck
3. DSA if unable to determine based on CT/MR alone
Is CVT confirmed?
Yes
No
Treatment
1. Anticoagulation with UFH or LMWH
2. Endovascular treatment - indications:
• Clinical deterioration despite anticoagulation
• Contraindications to anticoagulation
• Coma
• Deep CVT
• ICH
3. Hemicraniectomy:
• Large lesions with herniation
4. Intracranial pressure monitoring
Diagnosis and treatment of alternative conditions
Brain tumor, infection, ischemic stroke, PRES, idiopathic intracranial hypertension

INFECTIVE

- Orbital cellulitis
- Orbital Mucormycosis

Rhino-orbital Mucormycosis

- An **aggressive invasive fungal infection** which tends to affect patients with a history of diabetes (especially with ketoacidosis), chronic steroid use, and immunosuppression
- COVID Pandemic



- Inhalation of spores → upper respiratory infection → Spread to contiguous sinuses → spread to orbit and brain.
- **PATHOGENESIS** : Invasion of blood vessels by the hyphae results in **occlusive vasculitis** with **infarction** of orbital tissues.
- **CLINICAL FEATURES** : Facial and periorbital swelling, diplopia , visual loss ,proptosis , ophthalmoplegia , **black eschar** on the palate, turbinates, nasal septum, skin, eyelids.
- **INVESTIGATIONS** : Tissue biopsy from nasopharynx/orbit. **Histology: Non-septate , large , branching hyphae** .
- **COMPLICATIONS** : Retina vascular occlusions, cranial nerve palsies, cerebrovascular occlusions.
- **TREATMENT** : Treatment of underlying conditions, wide excision of necrotic issues , intravenous **amphotericin B** (1mg/kg/day) or Liposomal amphotericin B for three weeks , Oral Posaconazole(300 mg/day) and **hyperbaric oxygen therapy**.

ORBITAL INFLAMMATORY DISEASE

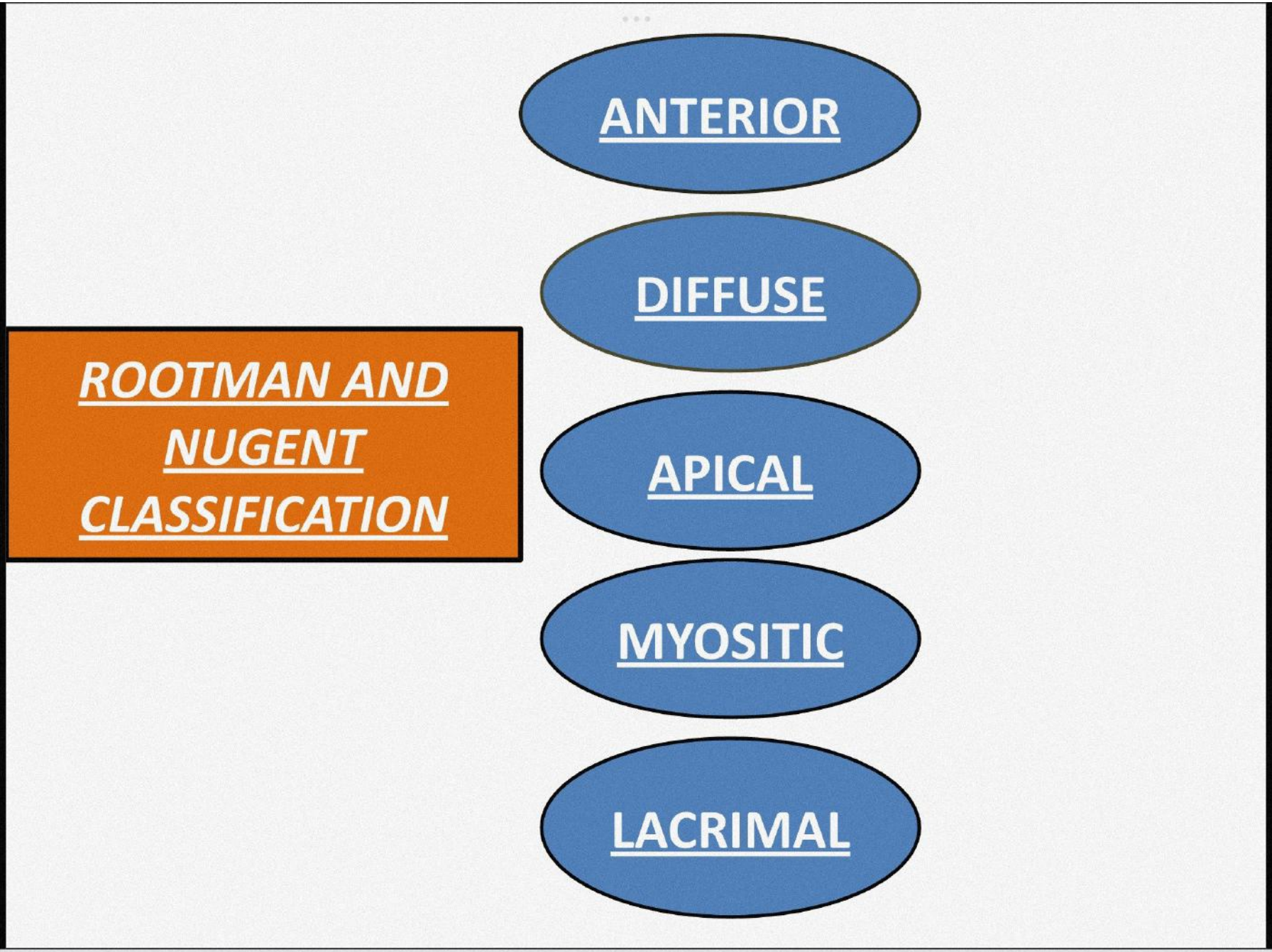
- Characterized by non-neoplastic and noninfectious space occupying orbital lesion
- Unilateral disease is the rule in adults but in children bilateral involvement occurs in 30%
- Presentation is usually b/w 20 and 50 years with abrupt painful onset

Classification of Orbital Inflammation

- **Idiopathic inflammatory disease**
- **Neoplasm**
  - Lymphoma
  - Rhabdomyosarcoma
  - Choroidal Malignant melanoma with extrascleral spread
  - Metastatic disease
- **Conenital malformation**
  - Dermoid cyst
  - Lymphangioma
- **Infectious disease**
- **Systemic inflammatory disease**
  - Thyroid associated ophthalmopathy
  - Wegener's granulomatosis
  - Churg strauss disease
  - Giant cell arterirtis
  - PAN
  - Sarcoidosis
  - Crohn's disease
  - SLE, RA, Scleroderma
  - Erdheim chester syndrome
  - Histiocytosis X
  - Idiopathic fibrosclerotic disorders

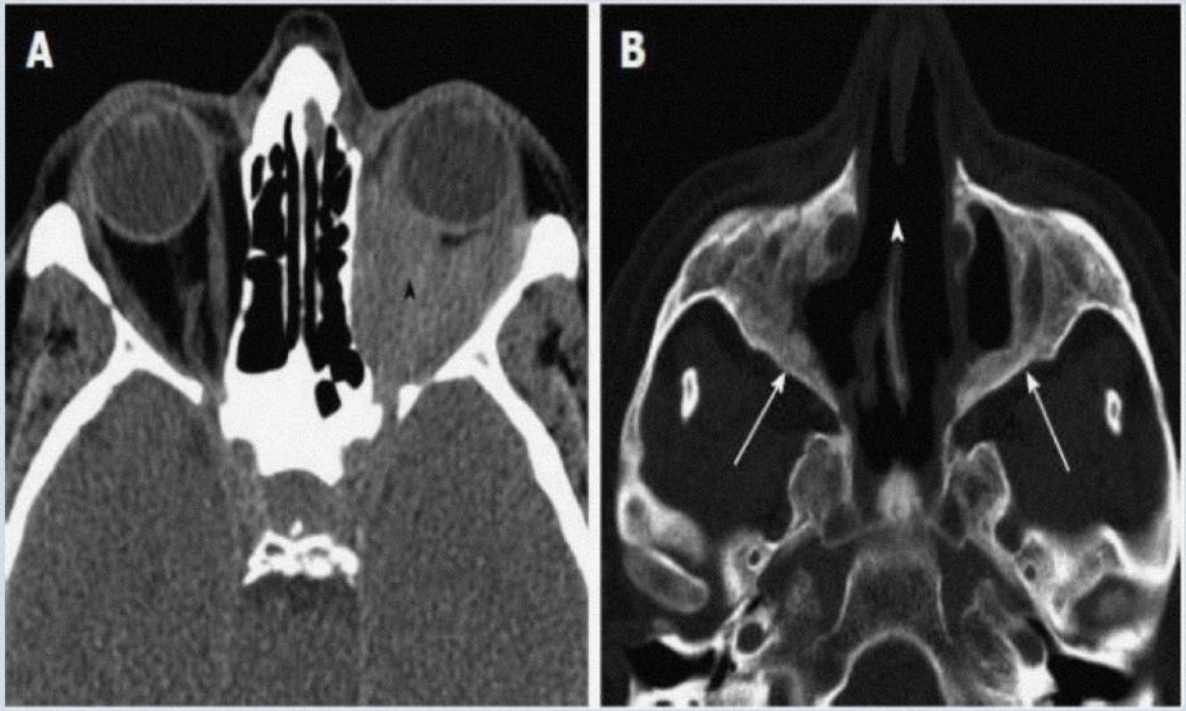
SIGNS

- CONJUNCTVAL CHEMOSIS /CONGESTION
- EYELID SWELLING
- RESTRICTED EOM
- PROPTOSIS
- IMPAIRED VISION
- ↓corneal sensitivity
- Hi IOP
- INTRAOCULAR uveitis/ RD



Diffuse OID

- 4-11% of all OID

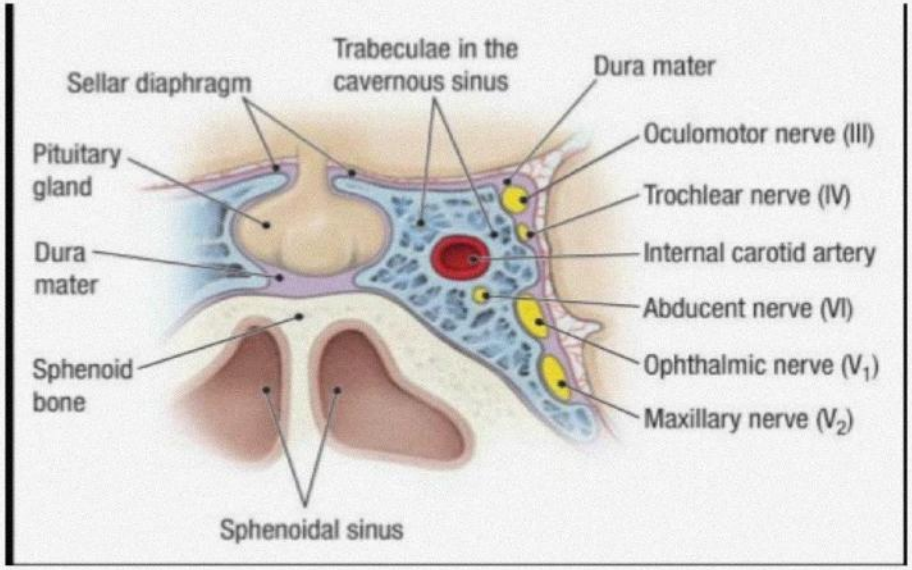


ORBITAL APICITIS

Less common and with poor outcome

Risk of invading ON and extension into Cavernous Sinus

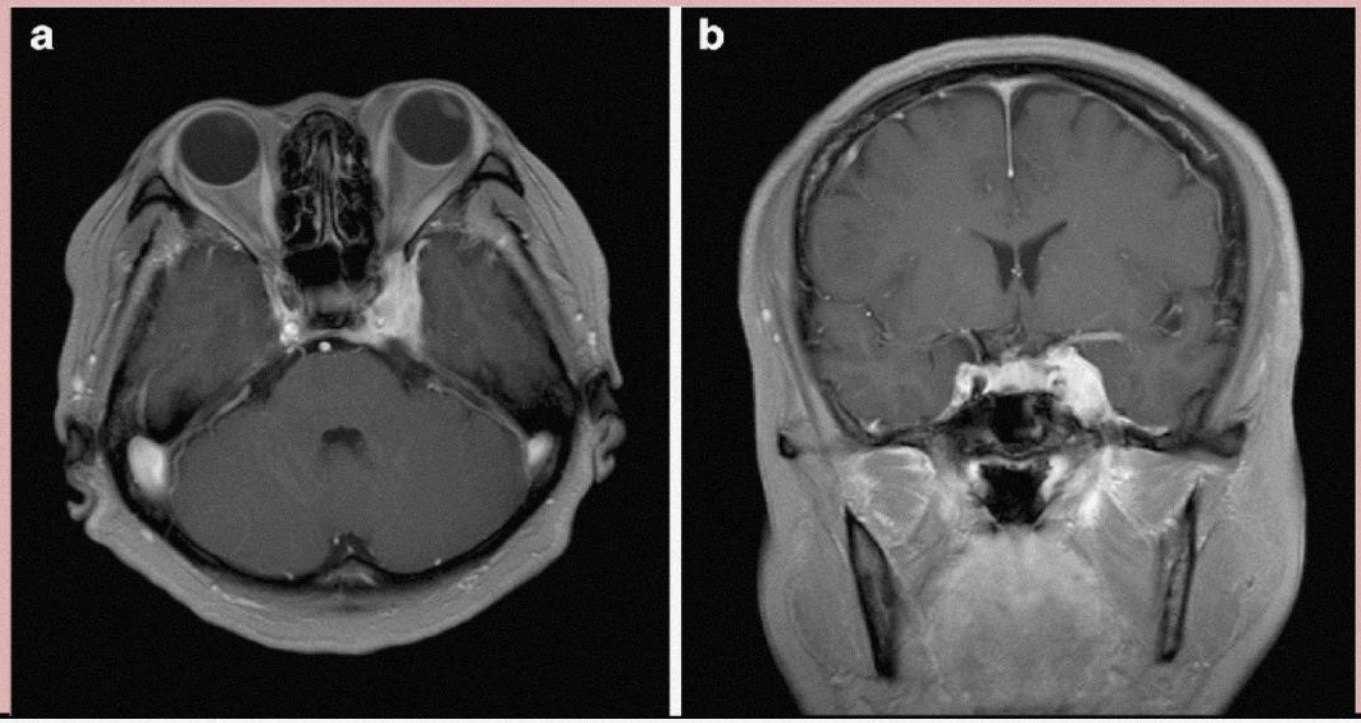
TOLOSA HUNT SYNDROME is inflammation of cavernous sinus with remitting relapsing orbital pain and invol of 3rd 4th 5th and 6th CNs



T1 MRI shows intermediate intensity as inflammatory tissue replaces the normal high intensity fat at the orbital apex

T2 MRI shows low intensity with darker signal indicating high degree of fibrosis

CNS involvement include abnormal soft tissue extension into the MCF, expansion of the ipsilateral cavernous sinus walls and post gadolinium enhancement of the meninges and dura



Corticosteroids

First-line therapy

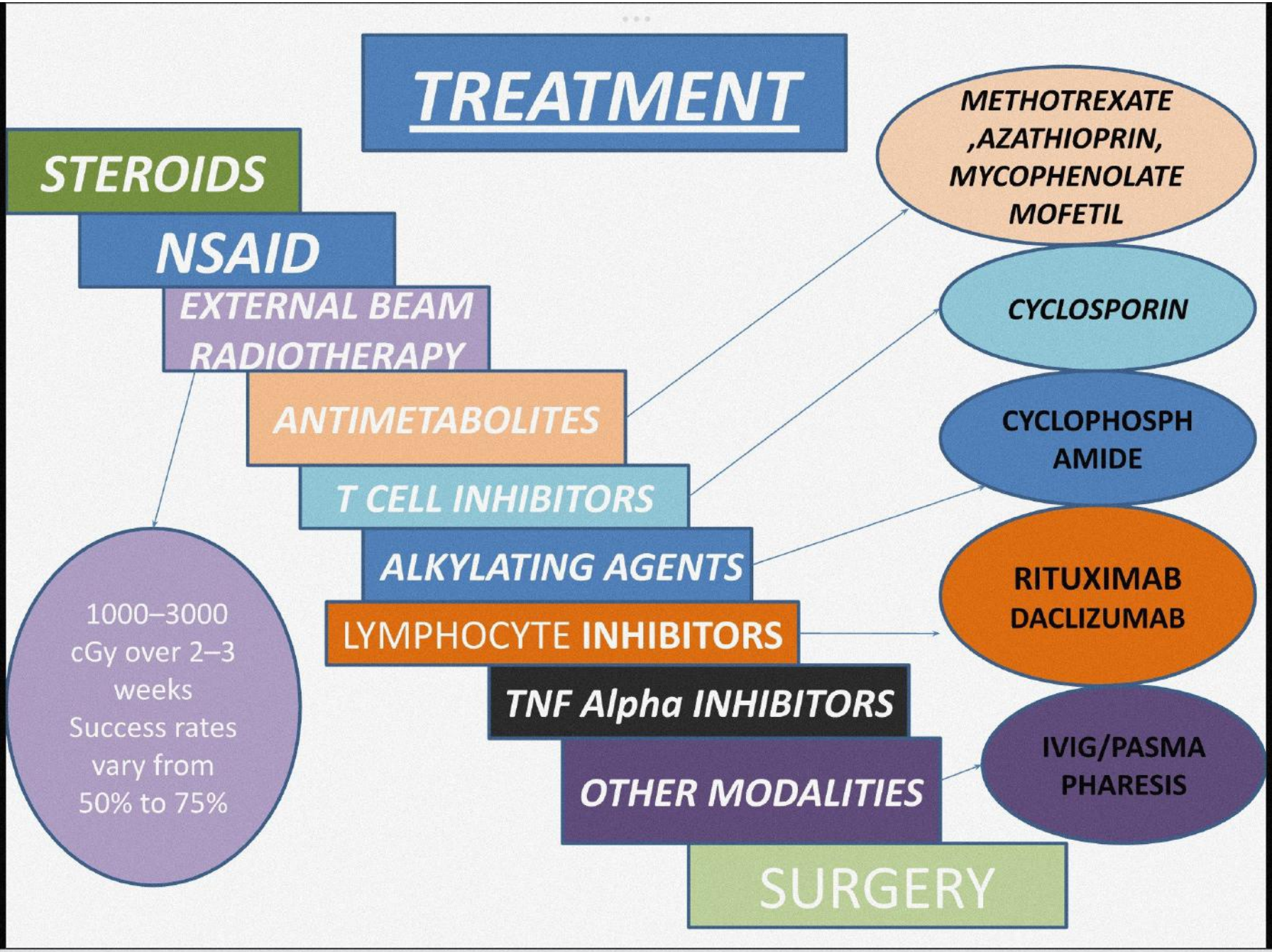
Anti-inflammatory

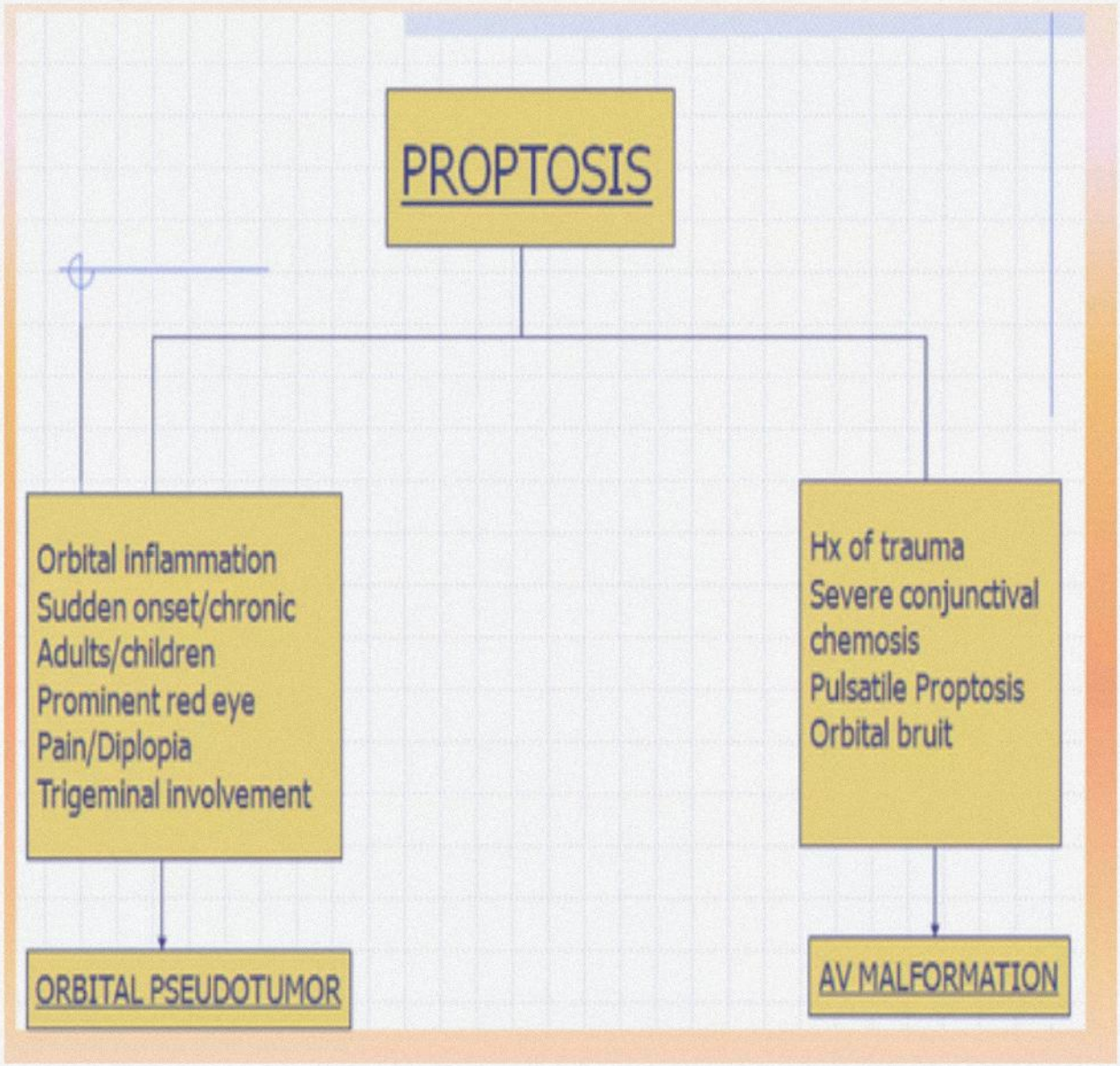
(inhibition of Phospholipase A2 and Cyclooxygenase pathways)

Immunosuppressive **(inhibition of IL and IFN synthesis, inhibition of major histocompatibility antigen expression, and cytotoxic effect on T lymphocytes)**

Over 75% of patients show dramatic improvement

Starting dose of 1 mg/kg/day of Prednisone
with a slow taper over 6–8 weeks




PROPTOSIS
Orbital inflammation
Sudden onset/chronic
Adults/children
Prominent red eye
Pain/Diplopia
Trigeminal involvement
Hx of trauma
Severe conjunctival chemosis
Pulsatile Proptosis
Orbital bruit
ORBITAL PSEUDOTUMOR
AV MALFORMATION

Neoplastic Disorders

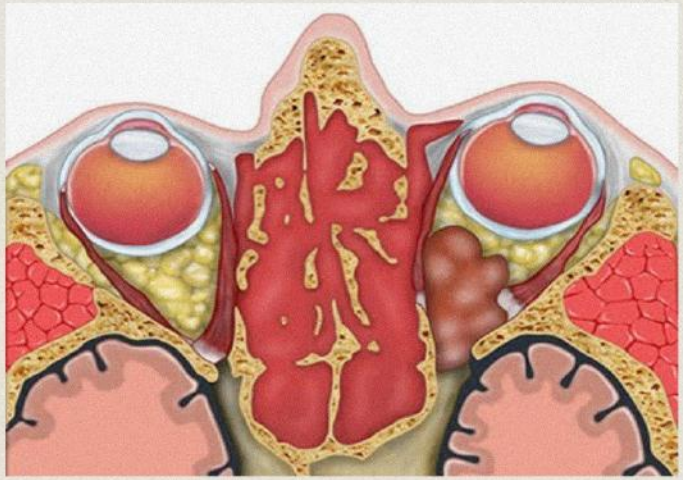
- Orbital lymphoma
- Lacrimal gland tumors
- Neural tumors
- Metastasis

Orbital lymphoma

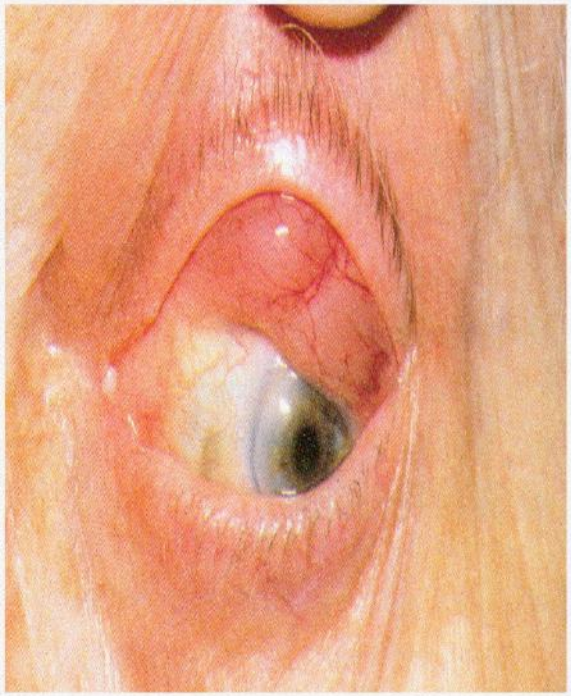
- **lymphoma occurring in the conjunctiva, lacrimal gland, eyelid and ocular musculature**
- Primary non-Hodgkin's lymphoma (NHL) of the orbit is a rare presentation with an indolent course

ETIOLOGY

- **Increasing age, immunosuppressive drugs, or autoimmune disorders such as rheumatoid arthritis, lupus, anemia, and HI**
- **highly curable** with RT



ANTERIRO ORBITAL LYMPHOMA



BILATERAL ORBITAL LYMPHOMA

Lacrimal Gland Tumors

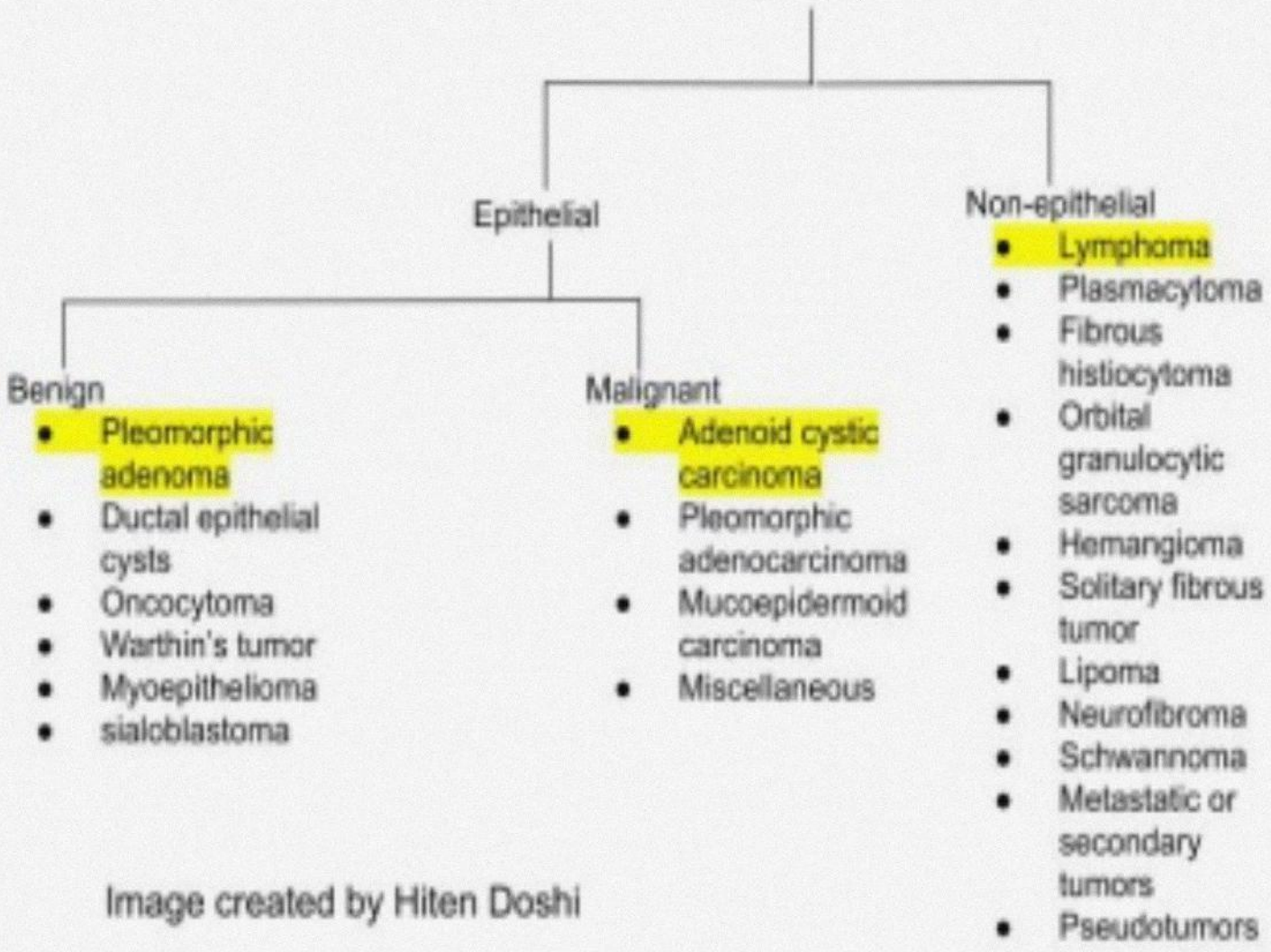
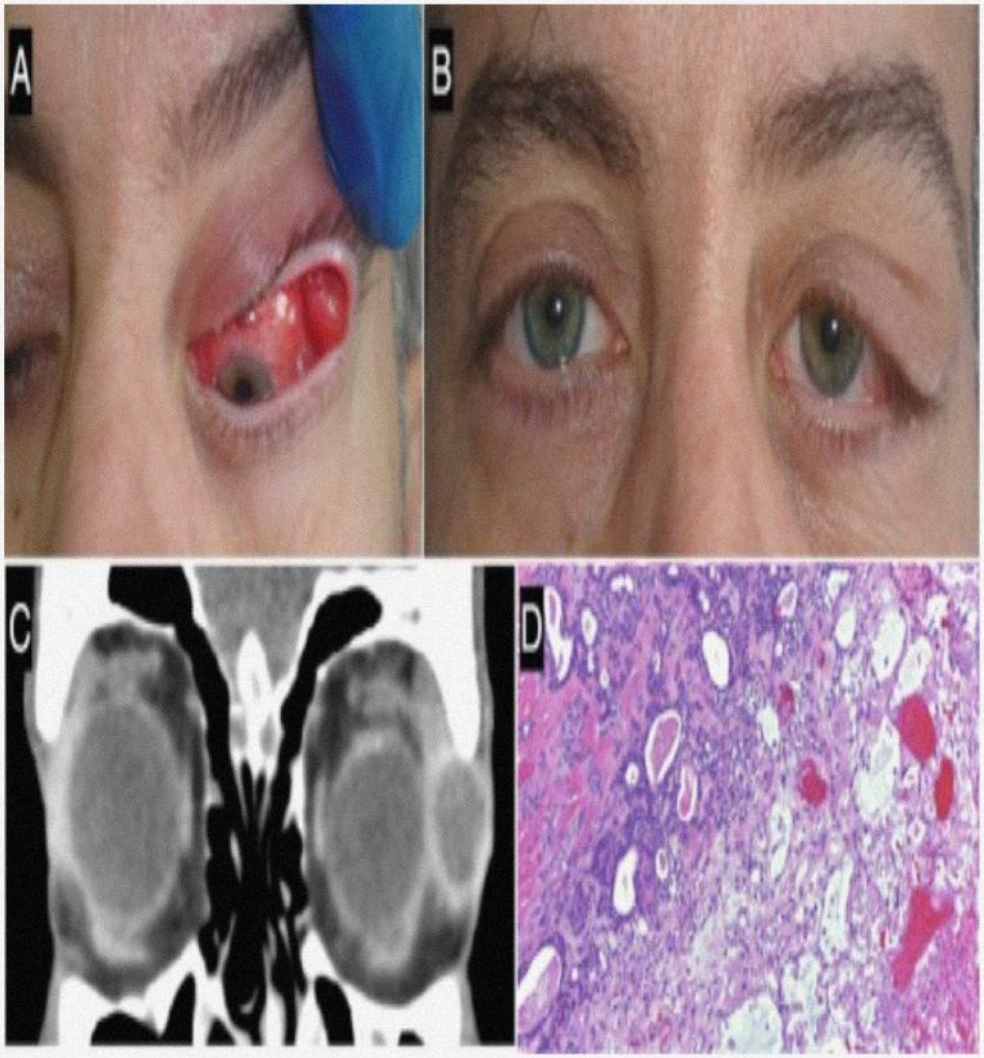


Image created by Hiten Doshi

Pleomorphic adenoma

- **Most common epithelial tumor of the lacrimal gland from ducts, stroma and myoepithelial cells**
- **5th decade of life**
- **Painless slow growing lesion in the upper outer quadrant for ≥ 1 year**

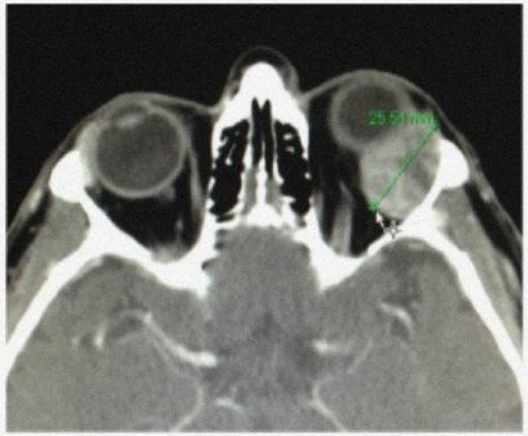


- Smooth,firm,non tender mass in the lacrimal gland fossa
- Tends to extend backwards may cause proptosis opthalmoplegia and choroidal folds

CT scan –round or oval smooth outline with excavation of the lacrimal gland fossa without destruction

Treatment .....complete excision without capsule disruption

Incomplete excision .....recurrence and malignant transformation

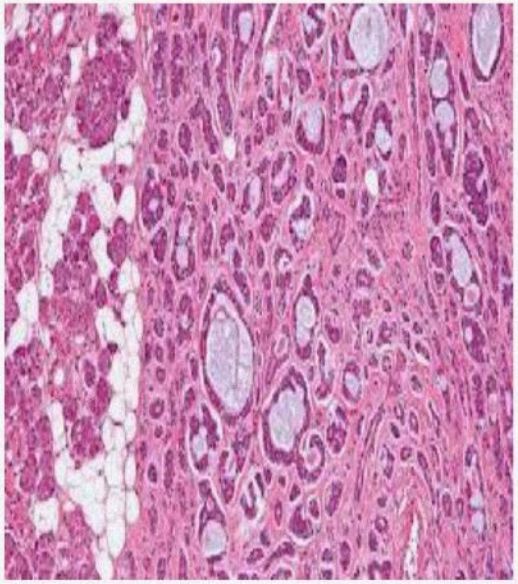


Lacrimal gland carcinoma

- Rare ,high morbidity and mortality
- 4^{th} and 6^{th} decade of life with shorter history

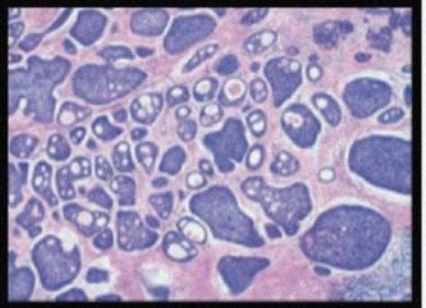
Histologic types

- **Adenoid cyst carcinoma**
- **Pleomorphic adenocarcinoma adenocarcinoma**
- **Mucoepidermoid carcinoma**
- **Squamous cell carcinoma**



ADENOID CYSTIC CARCINOMA OF THE LACRIMAL GLAND

- Most common malignant epithelial tumour of lacrimal gland.
- Presents in young or middle-aged adults , slight predilection in women.
- SYMPTOMS – Proptosis , pain due to perineural invasion is characteristic.
- SIGNS - Inferior and nasal displacement of globe, hypoaesthesia in the region supplied by lacrimal nerve, optic disc swelling and choroidal folds. Extension of the tumour posteriorly with involvement of the superior orbital fissure may cause epibulbar congestion, proptosis, periorbital oedema and ophthalmoplegia.
- INVESTIGATIONS - CT ORBIT - Ovoid soft tissue mass in the superotemporal quadrant of orbit with irregular margins and bony erosion . Focal calcifications may also be present.
- HPE – The histological patterns seen on biopsy include - cribriform , basaloid, sclerosing, comedocarcinoma and tubular. The cribriform pattern is most common and consists of lobules of tightly packed basaloid cells with intervening circular pools of mucin – swiss-cheese pattern.
- TREATMENT – Surgical resection of tumour with postoperative radiotherapy. For advanced tumors with high risk of recurrence - neoadjuvant intraarterial chemotherapy with cisplatin and doxorubicin used with exenteration and irradiation.
- Mortality is due to intracranial spread as a result of perineural invasion and pulmonary metastasis.



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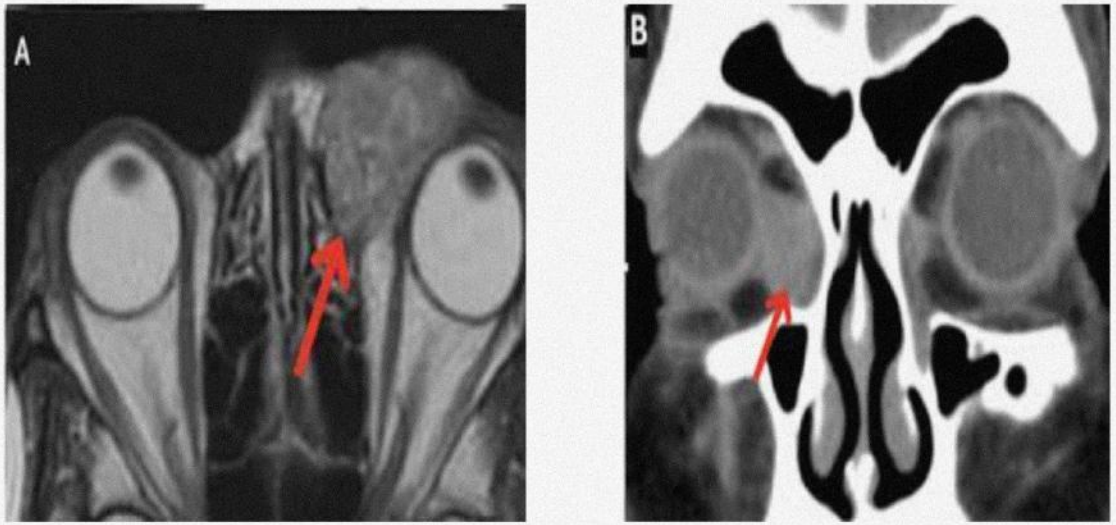
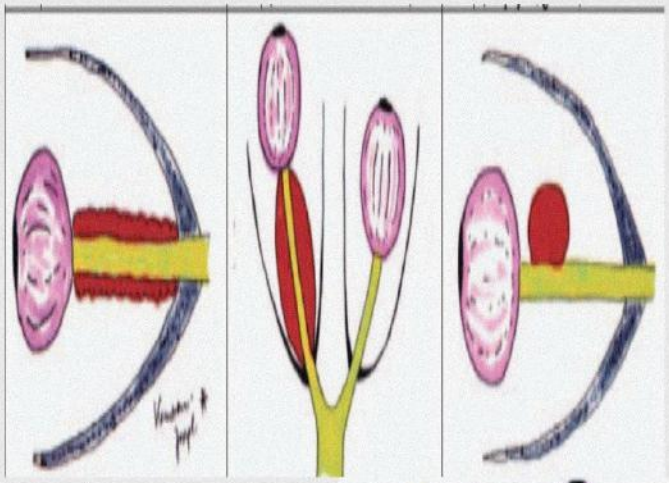
LACRIMAL SAC TUMORS



Classification

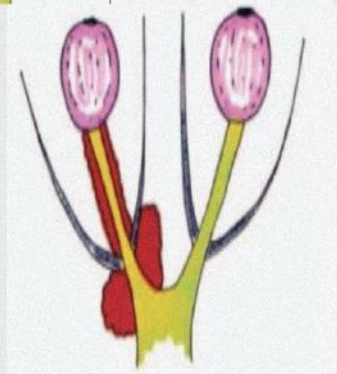
- **Primary Neural Tumors**

  Optic nerve
  Peripheral nerves



- **Secondary Neural Tumors**

Metastatic and infiltrative neoplasms
Breast cancer, prostate cancer, melanoma, lung cancer



ON may be affected by peripheral nerve tumors in the orbit

ON sheath Meningioma

- Benign tumors
- Meningothelial cells of the arachnoid layer within the optic nerve sheath
- Mean age - 41 to 48 years
- Higher incidence in females (61%)
- Unilateral (95%)
- Neurofibromatosis type 2 (NF-2) has an incidence of 9%, present at younger age, may be bilateral

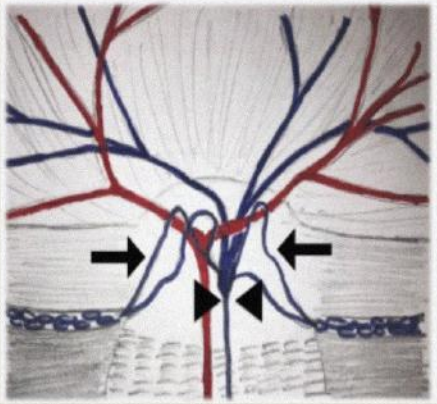
- Intracranial Meningiomas of sphenoid wing invade the orbit

CLINICAL FEATURES

- **Visual loss**

  gradual, painless, and usually 1 to 5 years before presentation
- **Proptosis**

  rarely the initial symptom, (59%) upto 2 to 5 mm
- **Chronic optic disc swelling**
- Longstanding CRVC - optociliary shunt vessels
- Optic atrophy



classic triad of gradual visual loss, optic atrophy, and optociliary shunt vessels is consistent for optic nerve sheath meningioma

OPTIC NERVE SHEATH MENINGIOMA

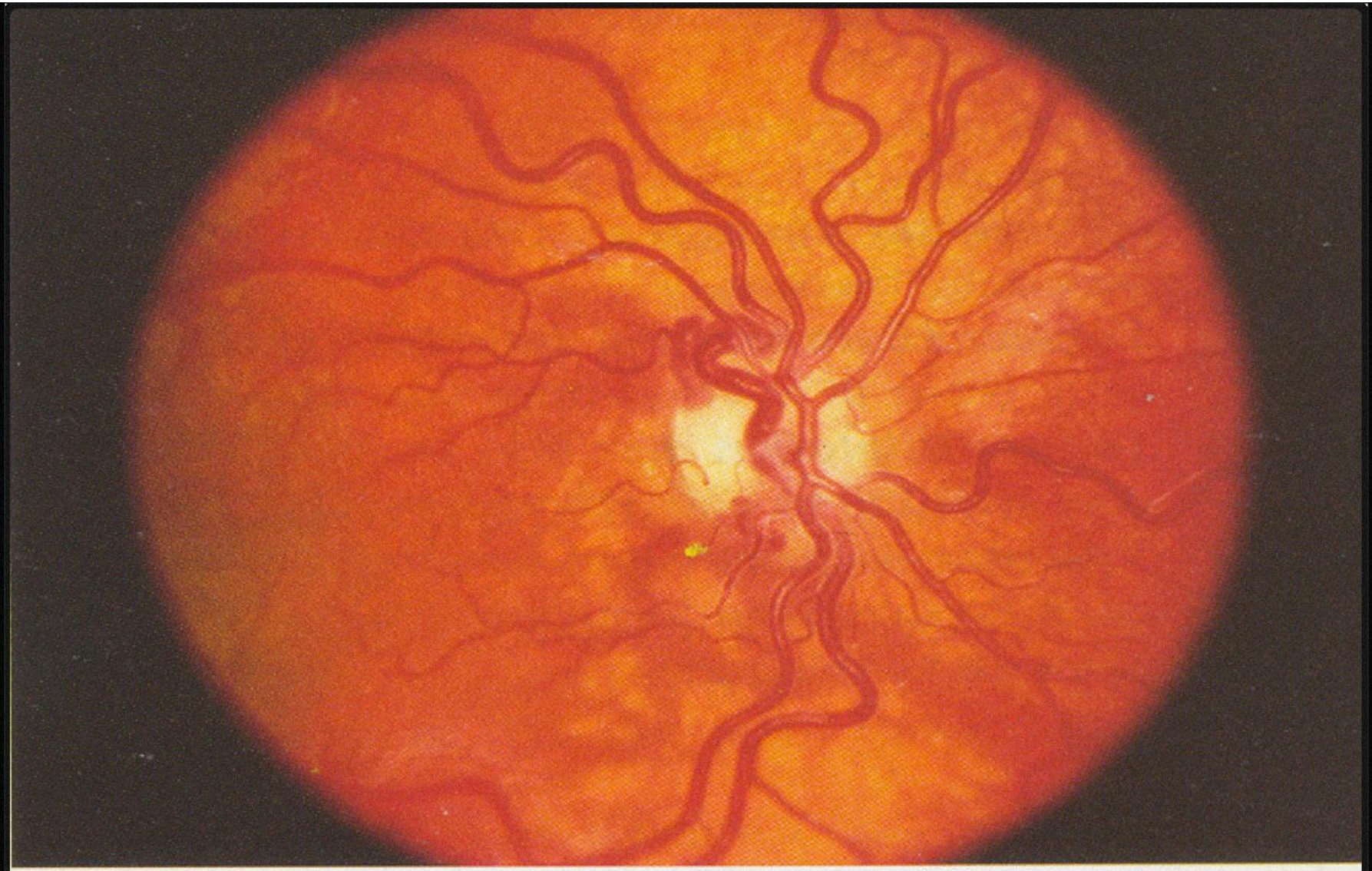


Figure 14.10 Optociliary shunt vessels

Diagnostic Approaches

CT SCAN

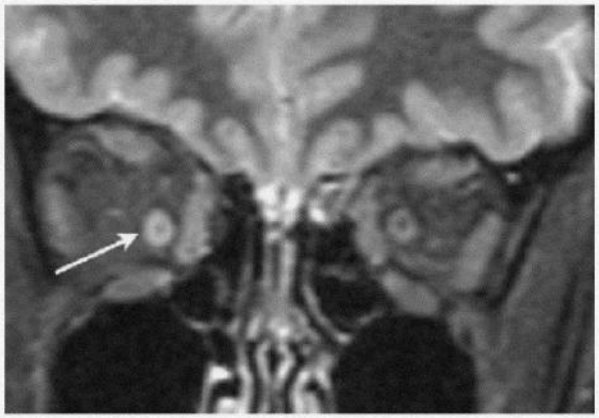
- ***Tram-track" signal*** An enlarged optic nerve with increased peripheral intensity and decreased central intensity"
- calcifications within the optic nerve sheath

MRI most sensitive and specific imaging modality

- T1-weighted MRI with contrast and fat suppression outlines the true anatomic borders of the optic nerve
- Homogenous enhancement of the lesion with gadolinium, though the optic nerve itself does not enhance

Biopsy

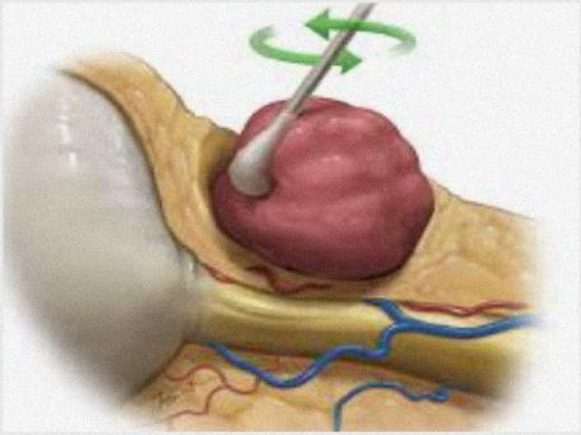
- often not needed except, if the patient has atypical sudden or rapidly progressive visual loss



TRAM-TRACK Signal

SURGICAL EXCISION

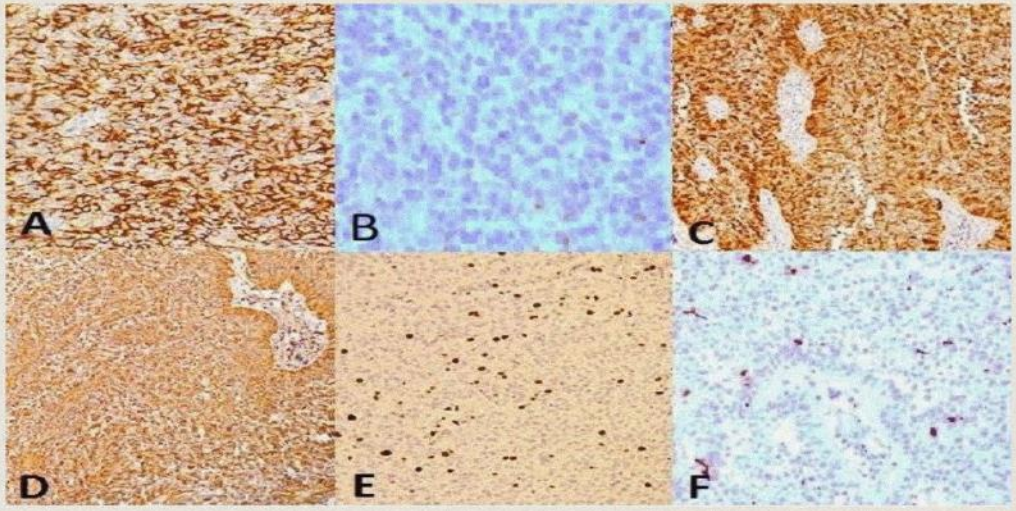
- **Progressive visual loss**
- **Disfiguring proptosis**
- **To relieve pain**
- **in children with more aggressive tumors**



- likelihood of blindness / damage to the pial vessels shared by the ON and the meningioma

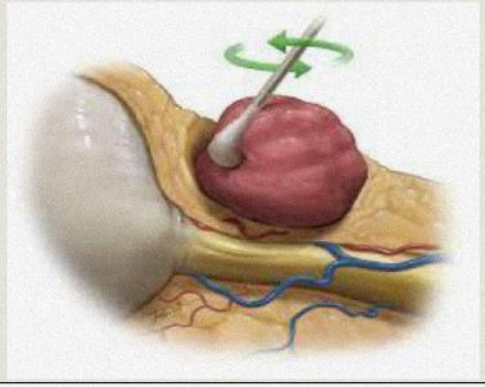
Cellular morphology
Immunohistochemistry

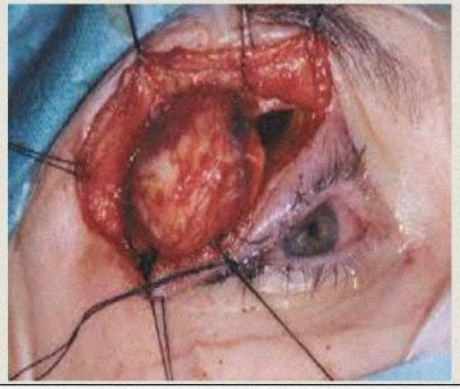
- Meningiomas are positive for EMA and S-100
- Gliomas are positive for GFAP and negative for S-100 and EMA
- Peripheral nerve sheath tumors contain Schwann cells that stain positively for S-100
- Perineural cells may also be positive for EMA
- Granular cell tumors stain positively for S-100 and may be positive for CD68 and leu-7

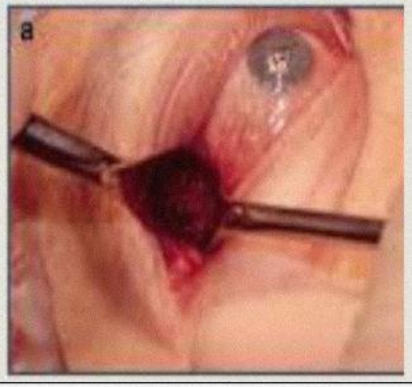


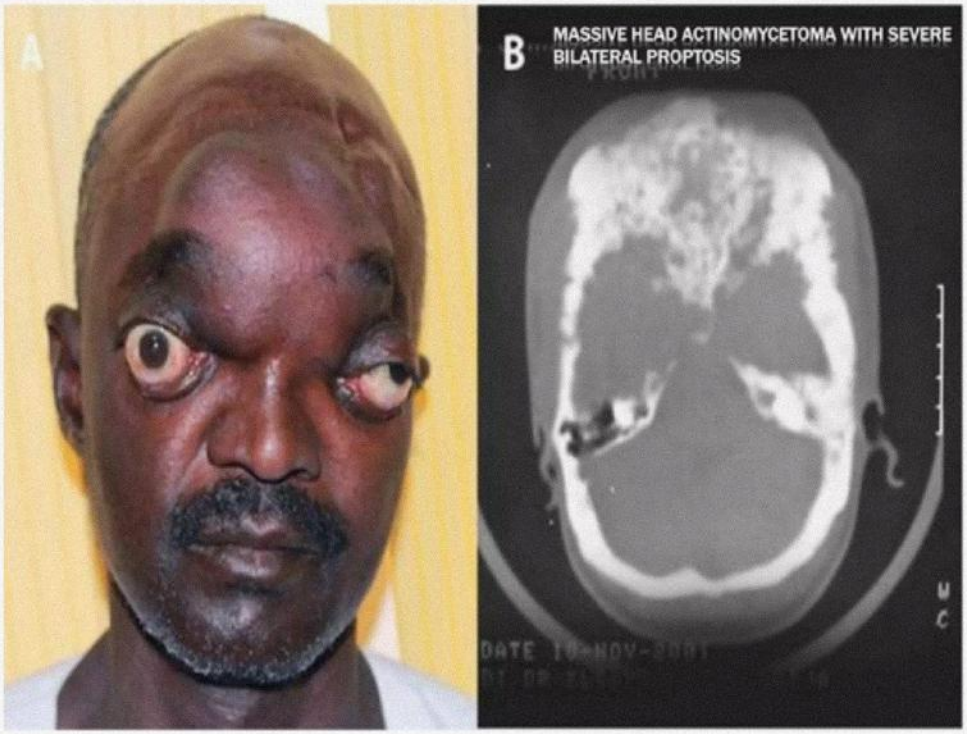
Treatment Options

- Observation
- Radiotherapy
- Chemotherapy
- Surgical excision








A
B
MASSIVE HEAD ACTINOMYCETOMA WITH SEVERE BILATERAL PROPTOSIS

Congenital malformations

- ***CRANIOFACIAL SYNOSTOSIS***

Mikulicz syndrome

- **Chronic autoimmune disease in which the glandular tissue of the head and neck are excessively enlarged, usually bilaterally**. Mostly, the salivary (parotid) and lacrimal (tear-duct) glands are affected

AXIAL PROPTOSIS

- ***Lesions of intra conal space arising from the optic nerve and central space***

Optic nerve Glioma

Optic nerve sheath Meningioma

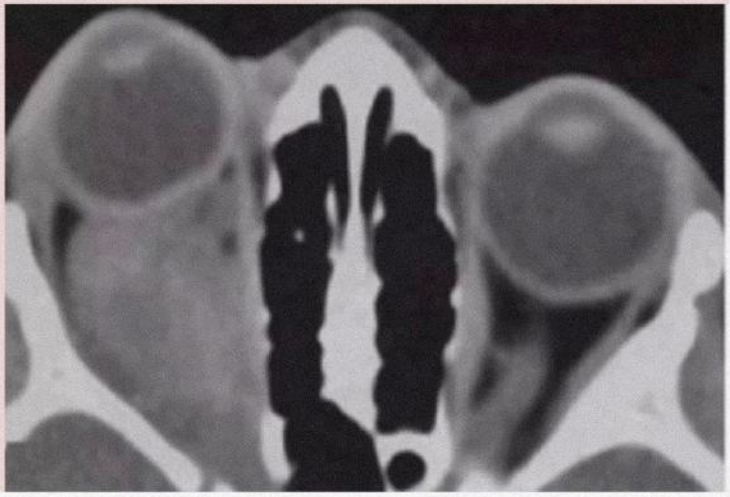
Cavernous hemangioms

Schwannoma

Neurofibroma

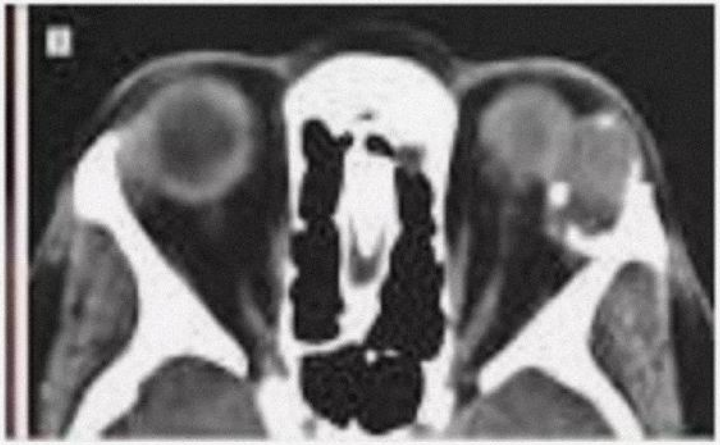
Orbital varices

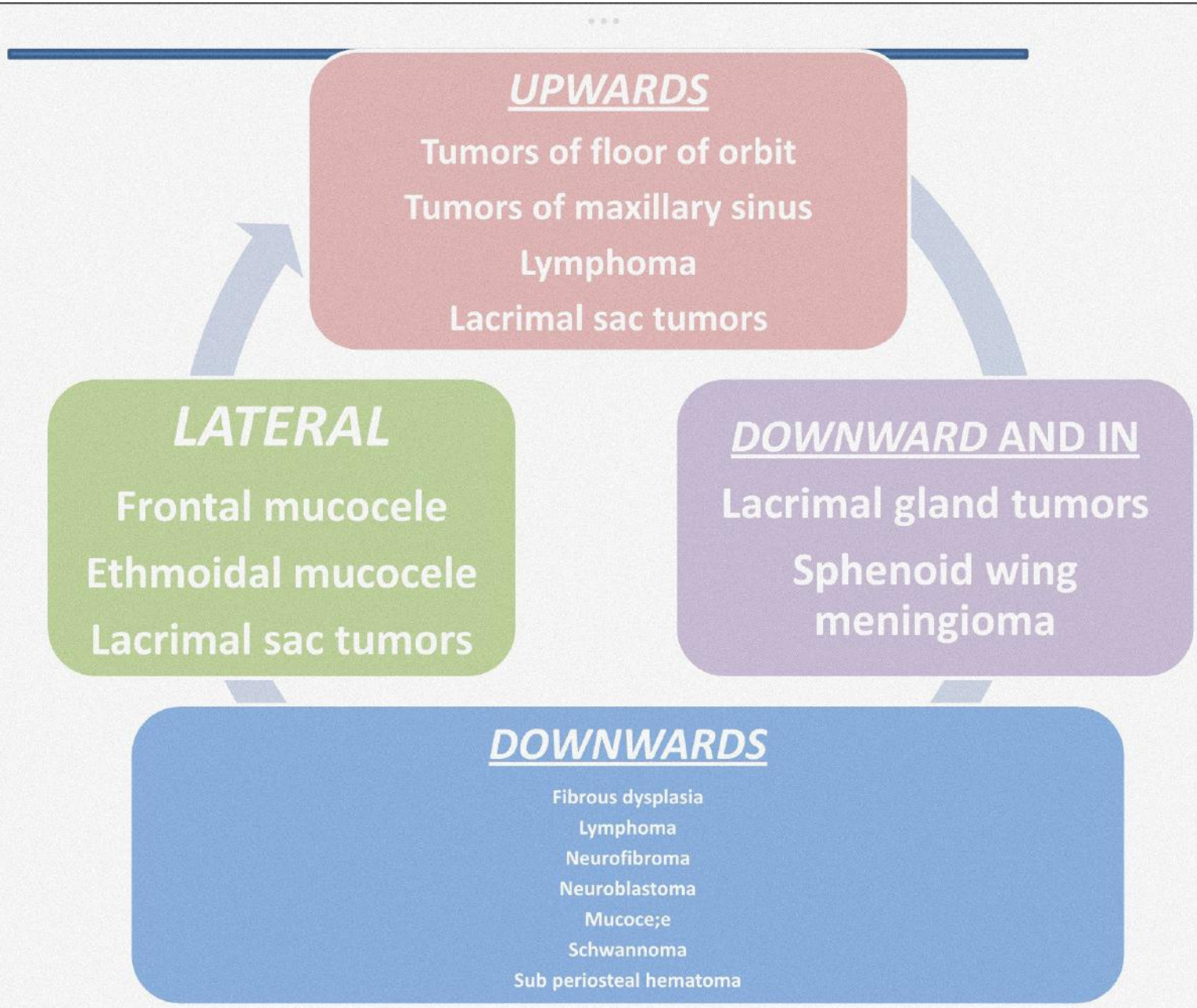
Hydatid cyst



NON AXIAL PROPTOSIS

Proptosis caused by any extraconal lesion or fracture displacement of orbital bones protruding inwardly.

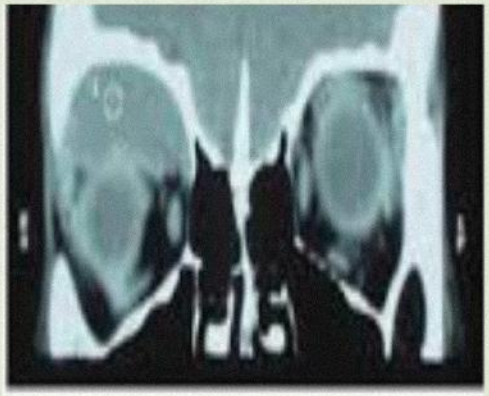




NON AXIAL PROPTOSIS

- ***Downward Displacement***

Fibrous dysplasia
Frontal mucocele
Subperiosteal hematoma
lymphoma
Neurofibroma
Neuroblastoma
Schwannoma
Thyroid Orbitopathy



- ***Upward displacement of the Globe***

  lacrimal sac tumor
  lymphoma
  Metastatic tumor
  Maxillary sinus tumor

- ***Lateral displacement of the Globe***

  lacrimal sac tumor
  Rhabdomyosarcoma
  Nasopharyngeal tumors
  Ethmoid mucocele
  Metastatic tumor

COMMON ORBITAL SPACE OCCUPYING LESIONS

ORIGIN	CHILDREN	ADULTS
CONGENITAL	Dermoid cyst Teratoma	
VASCULAR	Capillary haemangioms Lymphangioma	Cavernous haemangioma Orbital varices Haemangiopericytoma
NEURAL	Optic nerve Glioma Plexiform neurofibroma	Optic nerve meningioma Schwanoma Neurofibroma
MESENCHYMAL	AML Rhabdomyosarcoma	Fibrous Histiocytoma
HAEMPOIETIC	Histiocytosis Neuroblastoma	Lymphomas
METASTATIC	Neuroblastoma	Breast/Lung

- Proptosis with white reflex – retinoblastoma
- U/L fast progressing proptosis , fever , toxic child , pain – orbital cellulitis
- U/L axial proptosis with early vision loss – optic nerve glioma
- B/L proptosis , fever and toxemia – cavernous sinus thrombosis
- Pale child , bleeding from gums , U/L or B/L proptosis – leukemia
- U/L proptosis ,pain ,fever,hazy cornea and loss of vision - panophthalmitis

LATERALITY ON INSPECTION:

UNILATERAL PROPTOSIS:

- dermoid cysts,orbital teratoma,congenital cystic eyeball,
- Orbital hemorrhage,traumatic aneurysm,
- Cellulitis/abscess, cavernous sinus thrombosis
- TED,pseudotumour
- Varices,tumors,cysts.

BILATERAL PROPTOSIS:

- craniofacial synostosis,osteitis deformans,rickets
- TED,mickulicz syndrome
- Histiocytosis,amyloidosis,wegener's granulomatosis,
- Tumors.

TAKE HOME MESSAGES

- *The most common cause of bilateral/unilateral proptosis is GRAVES DISEASE*
- *Acute unilateral Proptosis suggests infection or vascular disorder (hemorrhage/CCF/CST)*
- *Chronic unilateral proptosis suggest tumor*
- *Role of imaging*
- *Care of exposed cornea*