Topic: Proptosis - Basics

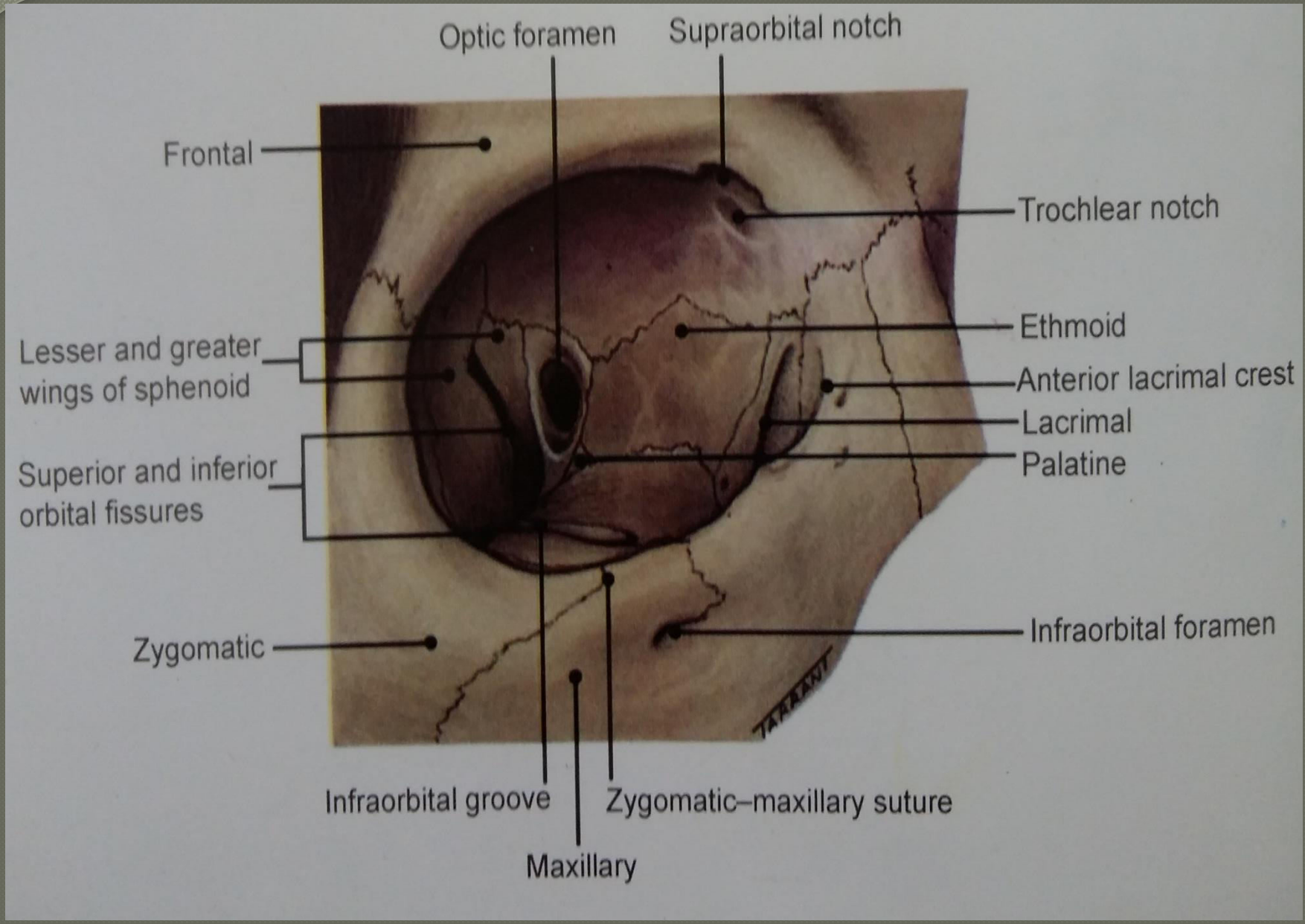
Learning objectives:

Discuss anatomy of orbit, definition enumerate differential diagnosis/causes etiology of proptosis in children and adults

Prof. Dr. Tariq Farooq Babar
Head & Incharge Eye "A" Unit
Khyber Girls Medical College,
MTI/Hayatabad Medical Complex,
Peshawar

Clinical anatomy of orbit

Orbit is a pear shaped cavity stalk of which is the optic canal



Anatomy of the orbit

Orbit has:

- Roof
- Lateral Wall
- Floor
- Medial Wall

Roof:

- Consist of two bones
  - Lesser wing of sphenoid
  - Orbital plate of frontal bone
- Defect in orbital roof – pulsatile proptosis
- Due to transmission of CSF pulsation to orbit.

Lateral wall:

- Consists of two bones
  - Greater wing of sphenoid
  - Zygomatic
- Anterior half of globe vulnerable to lateral trauma – it protrudes beyond the lateral orbital margin.

Floor:

- Consists of three bones
  - Zygomatic
  - Maxillary
  - Palatine
- Posteromedial portion of maxillary bone relatively weak – involved in a blow out fracture.

Orbital floor

- Forms roof of maxillary sinus
- Maxillary carcinoma invading orbit, displace globe upwards.

Medial wall:

Consists of four bones

- Maxillary
- Lacrimal
- Ethmoid
- Sphenoid

Lamina papyracea forms part of medial wall

Paper thin, perforated by numerous foramina for nerves and blood vessels.

Orbital cellulitis is therefore secondary to ethmoidal sinusitis.

Proptosis / Exophthalmos

Definition

- Abnormal protrusion of globe
- Displacement of globe relative to orbital rims
- Proptosis of more than 21mm or
- More than 2mm asymmetry between the two eyes is abnormal

Exopthalmos

Specially used to describe the proptosis of eyes associated with thyroid eye disease

Enophthalmos

Defined as retro displacement of eye into orbit

Conditions that mimic proptosis (Pseudo proptosis)

Ipsilateral large globe

- Megalophthalmos
- Buphthalmos
- High myopia

Ipsilateral lid retraction

- Contralateral anophthalmos

Dystopia

Displacement of globe in coronal plane

Differential diagnosis of Proptosis

Vascular

- Carotid cavernous fistula
- Cavernous sinus thrombosis
- Arteriovenous malformations such as
  - Hemangioma
  - Aneurysm
  - Varix

Trauma

- Retrobulbar hemorrhage
- Post-traumatic mucocele
- Encephalocoele (due to orbital roof fracture)

Endocrine

- Thyroid associated ophthalmopathy
- Grave's disease

Infective

- Orbital cellulitis
- Mucormycosis
- Granuloma

Inflammatory

- Orbital pseudotumor
- Myositis
- Granulomatous disease
- Sarcoidosis

Tumor

Primary like

- Schwannoma
- Lymphoma
- Optic nerve glioma

Metastases from distant sites commonly

- Leukemia
- Sarcomas

Pseudoproptosis

- Contr-lateral enophthalmos
- Contra-lateral globe rupture

Causes of proptosis in children & adults
Childhood proptosis

Congenital
- Exorbitism
- Cranio synostosis
- Skull anomalies
- Meningocele / encephalocele
- Dermoid cyst

Traumatic
- Orbital hematoma
- Traumatic hemorrhage in existing neoplasm

Inflammatory
- Orbital cellulitis
- Abscess
- Pseudo tumour
- Mucocele

Neoplastic

Ophthalmic / orbital

- Hemangioma
- Optic nerve glioma
- Rhabdomyosarcoma
- Orbital retinoblastoma
- Teratoma

Non ophthalmic

- Granulocystic sarcoma
- Metastatic neuroblastoma
- Lymphoma lymphosarcoma
- Histiocytosis x

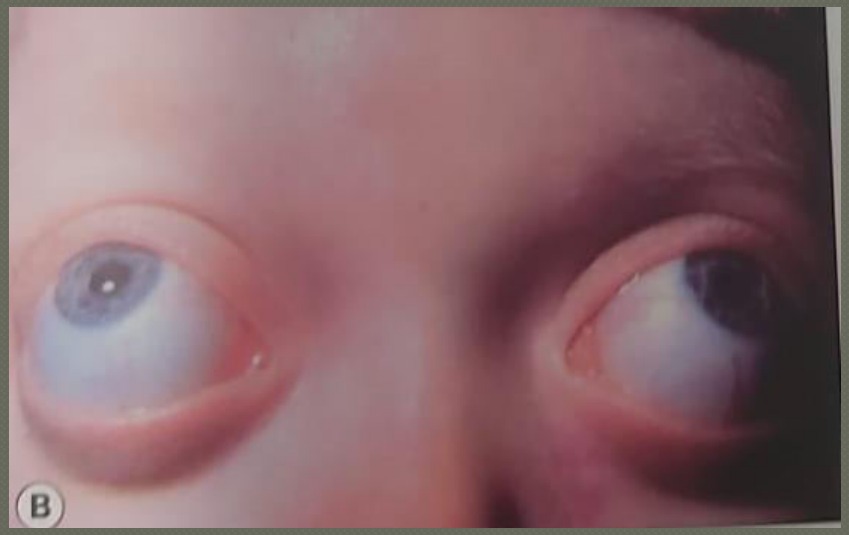
Congenital

Exorbitism

e.g Craniosynostosis

- Group of congenital conditions
- Abnormally shaped skull
- Cause being premature closure of skull sutures. e.g Crouzon syndrome and Apert syndrome and Pfeiffer syndrome


A


B

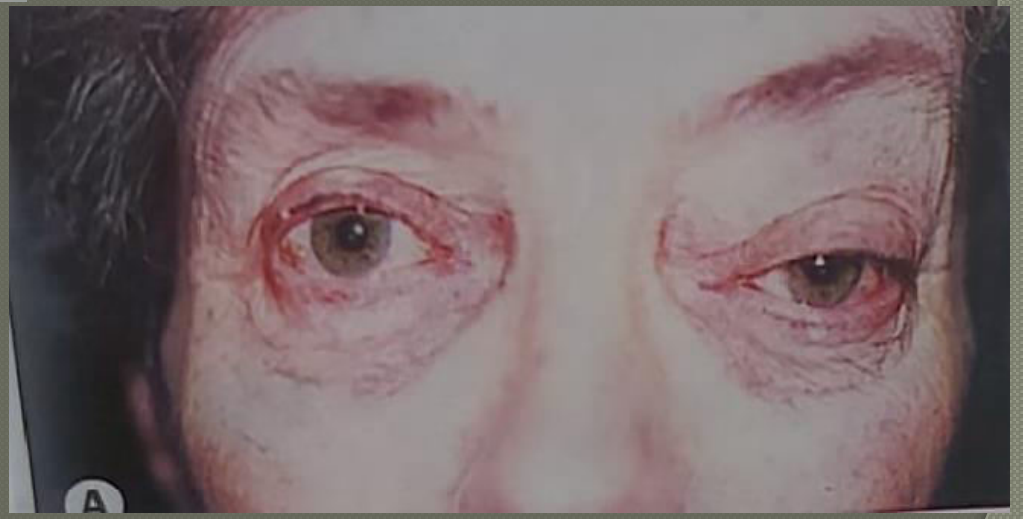
Encephalocoele

- Formed by herniation of intracranial contents through a congenital defect of the base of the skull
- Located at the front or back of the head
- A meningocoele contains only dura
- Meningo – encephalocoele also contains brain tissue.

Dermoid cyst

- A choristoma is a mass of histologically normal tissue in an abnormal location.
- It is derived from displacement of ectoderm to a subcutaneous location along embryonic lines of closure.
- Dermoids are lined by keratinized stratified squamous epithelium.
- They are smooth, non-tender 1-2cm in diameter


A


A

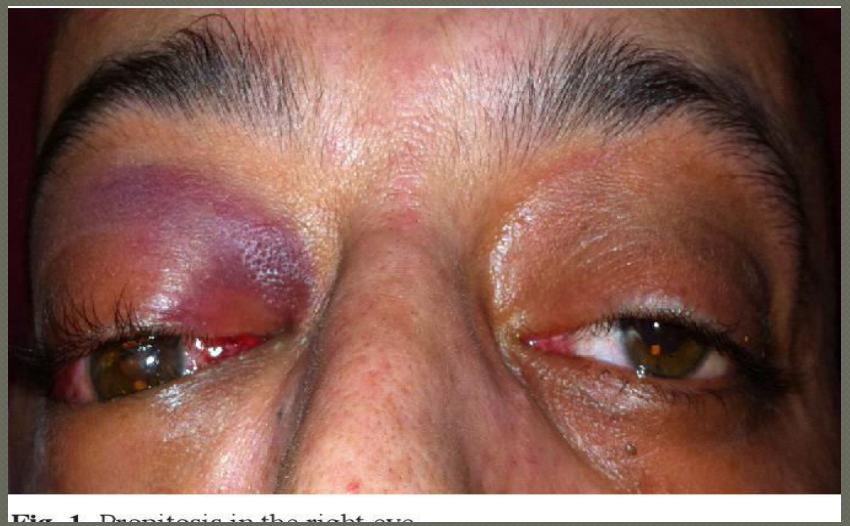
Traumatic

Orbital hematomas

- Can occur spontaneously
- Result of vascular anomalies
- Induced by trauma
- Following paranasal sinus surgery

Risk

Retrobulbar hematoma can compress optic nerve

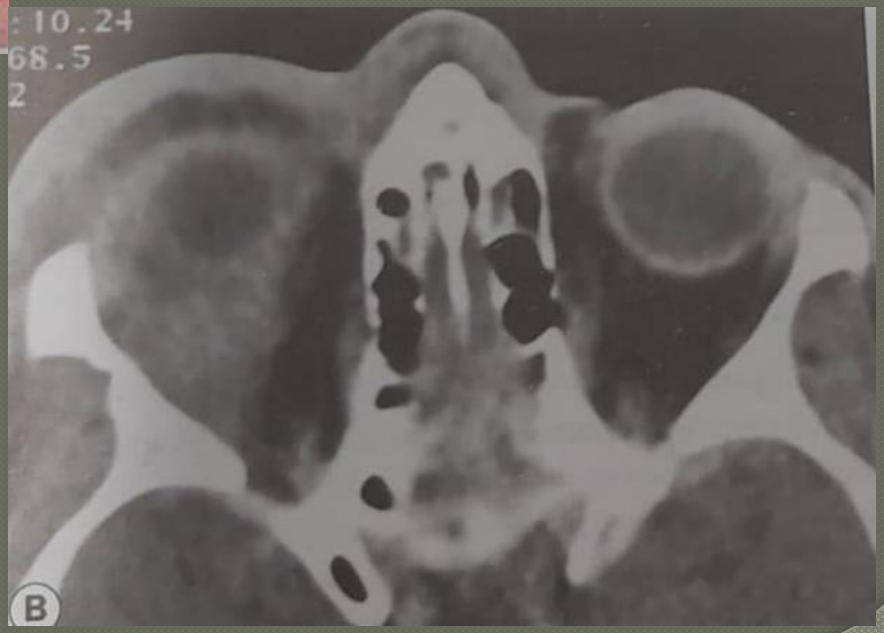


Inflamatory

Orbital cellulitis

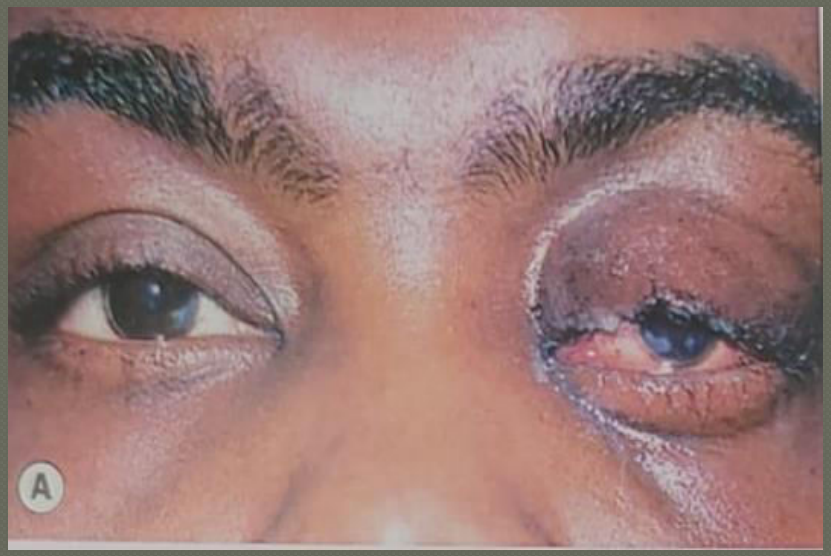
- In children usually secondary to ethmoiditis
- Child very unwell with high fever
- Rapid onset of proptosis usually down and out
- Pain, chemosis, lid oedema with restricted ocular motility
- In severe cases – there may be signs of optic nerve dysfunction

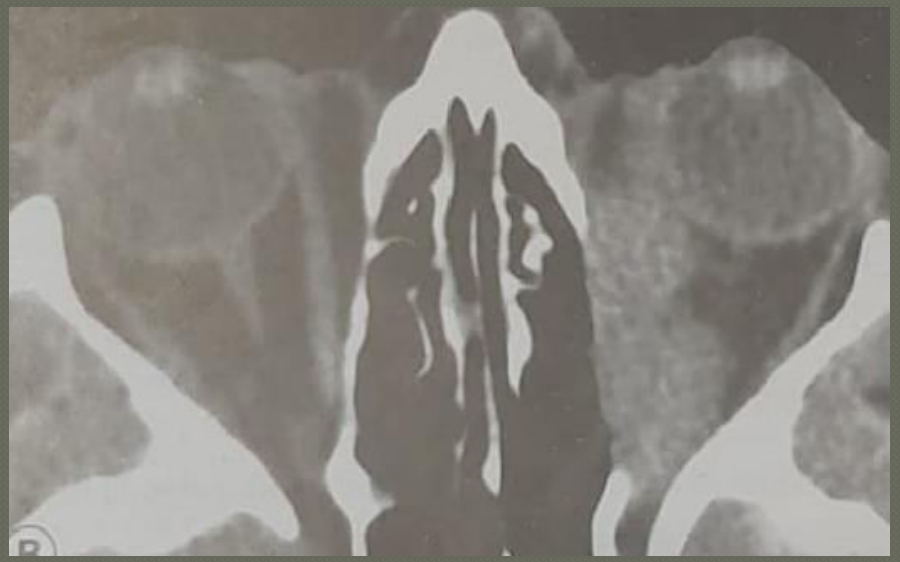

A


10.24
68.5
2
B

Pseudotumor (Idiopathic orbital inflammatory syndrome)

- Presentation between ages of 6 & 14 years
- 1/3rd of patients have bilateral involvement
- Subacute onset, axial proptosis associated with chemosis and lid oedema
- Look for Wegener granulomatosis in bilateral cases


A

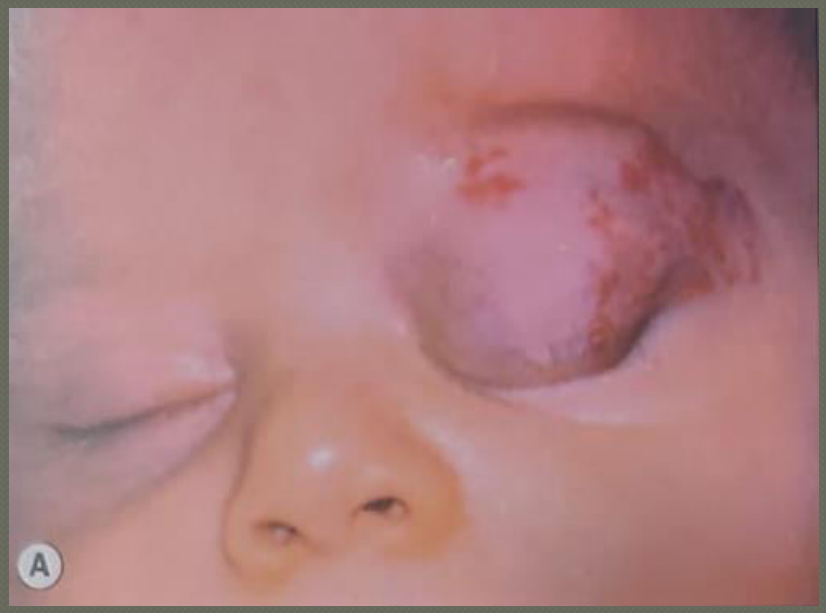


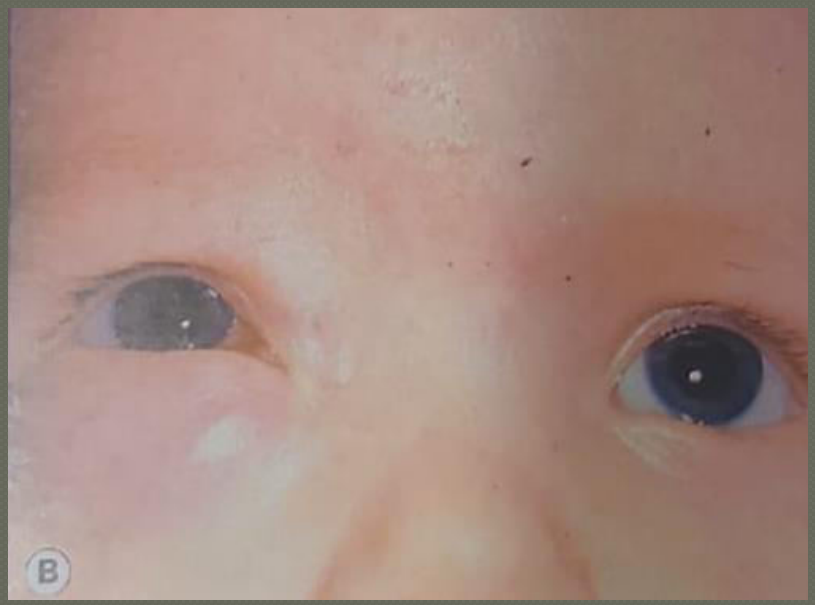

C

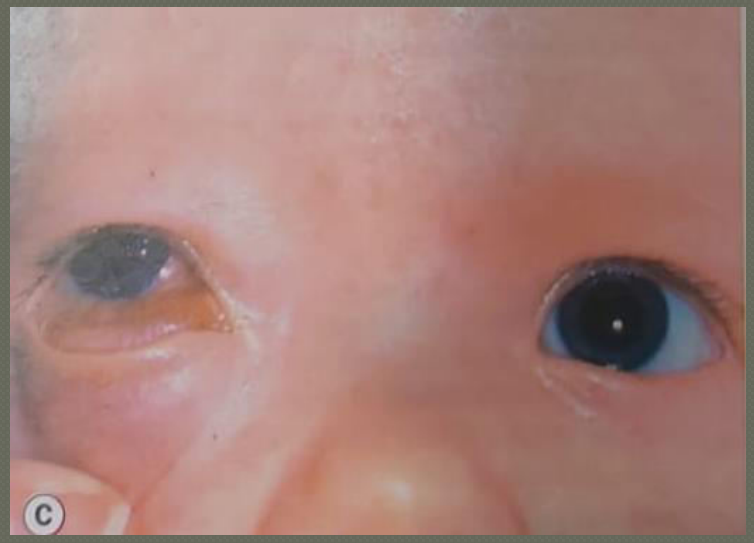
Neoplastic

Capillary hemangioma

- Presents usually at birth or early infancy
- Slowly progressive proptosis associated with an upper anterior orbital mass
- The mass becomes engorged and sligtly increases in size when the child cries
- The child may have capillary skin hemangiomas on the eyelids or else where.

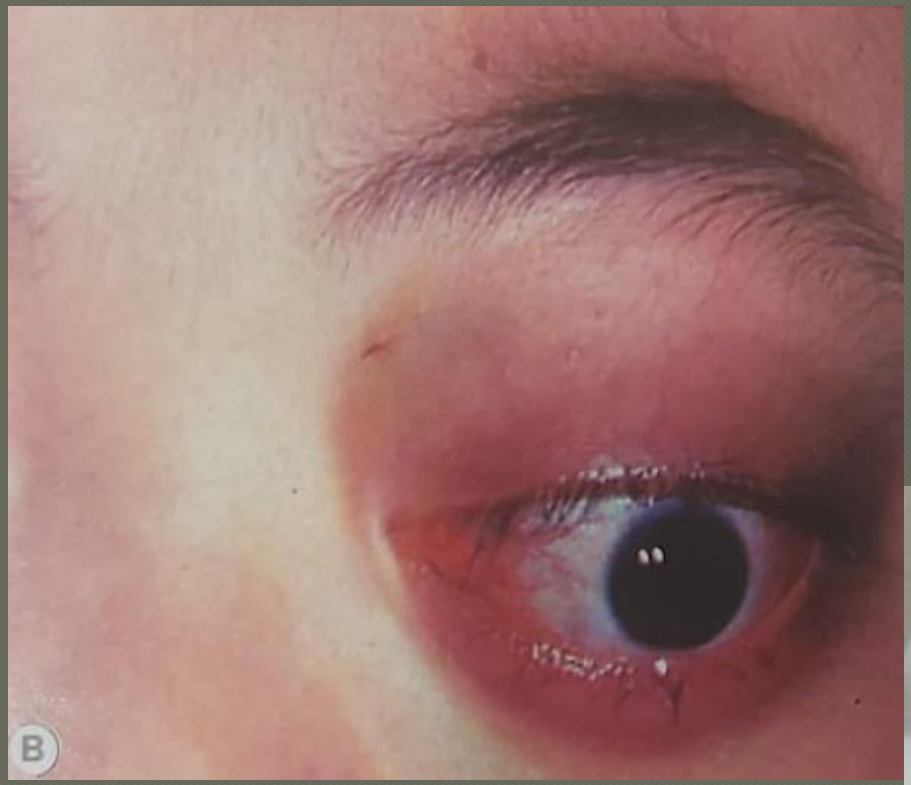

A

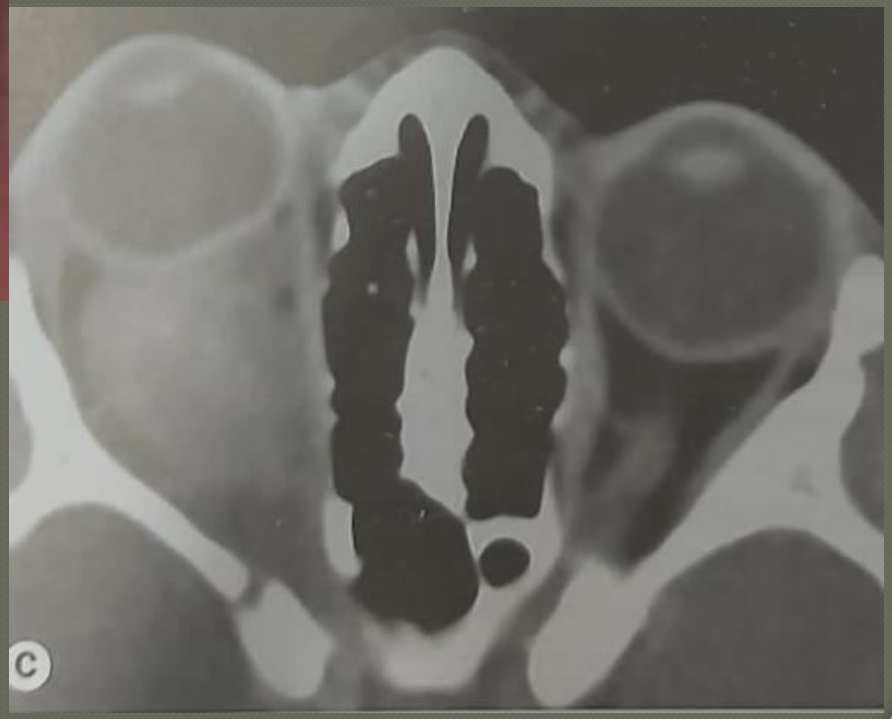

B


C

Optic nerve glioma

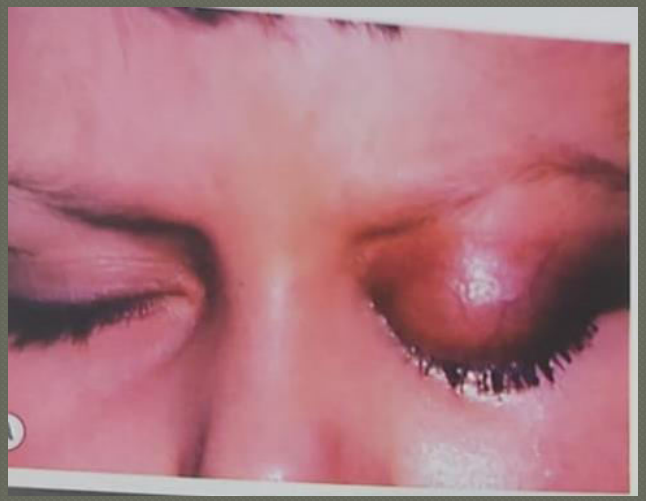
- Presentation between age of 2 and 7 years
- NF-I present in 50% of unilateral tumors and 100% in bilateral tumors
- Slowly progressive axial or non axial proptosis
- Decrease VA and an APD
- Optic disc – swollen, atrophic, optico cilairy shunts
- Disproportionate loss of VA compared to proptosis

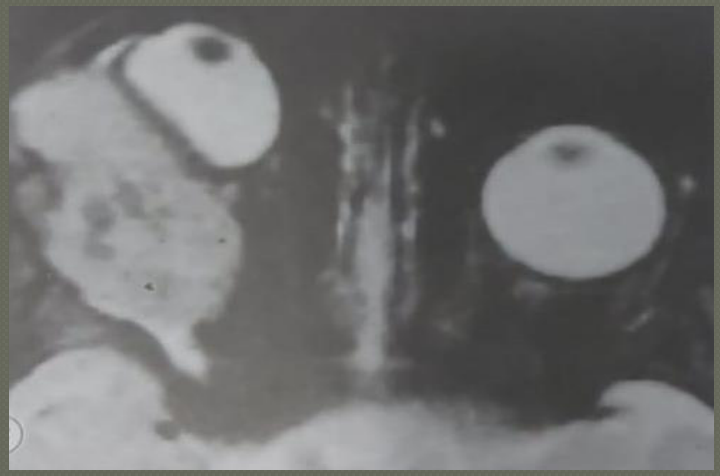

B


C

Rhabdomyosarcoma

- Presentation at age 7 years
- More common in boys than in girls
- Rapid onset of progressive painful proptosis with chemosis and lid oedema
- Location – retrobulbar followed by superior and inferior.





Metastatic neuroblastoma

Presents during 1st 5 years of life

Primary tumor

usually develops in the abdomen

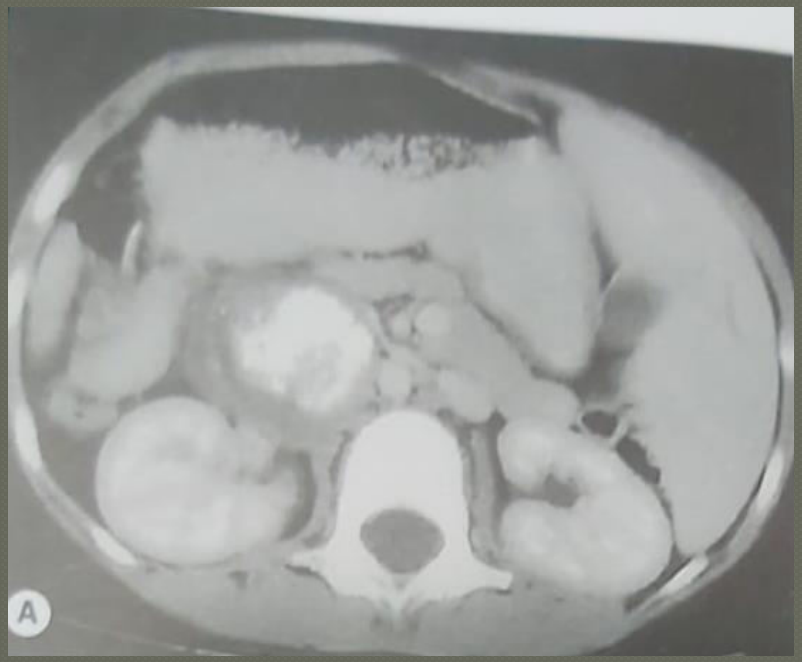
Metastatic tumor

Arising from neck or medinastinum may cause Horner syndrome

40% of metastasis involve both orbits

Sudden onset of rapidly progressive proptosis

Associations – echymosis and superolateral orbital mass



Histiocytosis x / Langerhans cell histiocytosis

A rare multisystem disorder consisting of three related and overlapping conditions

- Eosinophilic granuloma – lesions confined to bone
- Hand – Schuller – Christian disease - a triad of diabetes inspidus, proptosis and bony skull defects
- Letterer – Siwe disease – aggressive visceral involvement.

- Orbital involvement in 25% of cases with either Eosinophilic granuloma or Hand Schuller Christian disease.
- Bilateral or unilateral bony lysis and soft tissue growth
- Typically involves superolateral orbit

Proptosis in adults

Table 2: Common causes of proptosis in adults

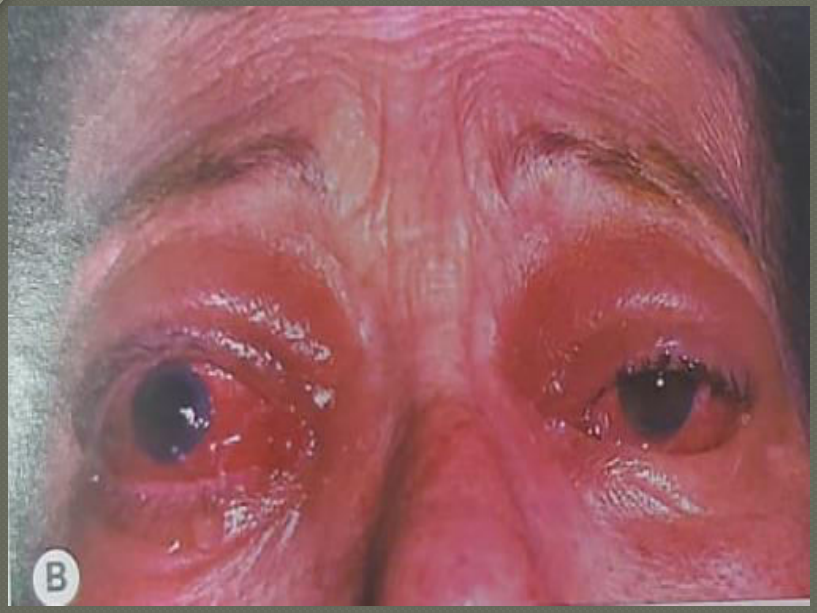
Category	Specific conditions
Endocrine	Thyroid eye disease
Trauma	Facial fracture, soft tissue swelling, retrobulbar haemorrhage
Vascular	Carotid-cavernous fistula, cavernous sinus thrombosis, cavernous haemangioma
Inflammatory	Inflammatory orbital pseudotumor, dacryoadenitis, orbital myositis, Tolosa–Hunt syndrome, Wegener's granulomatosis, sinus mucocele, sarcoidosis, Churg–Strauss syndrome
Infective	Orbital cellulitis, mucormycosis
Tumours	Lymphoma, schwannoma, sinonasal tumour, lacrimal gland tumour, meningioma, neurofibroma, optic nerve glioma, metastasis, myeloid sarcoma, ossifying fibroma, orbital osteoma, haemangioblastoma, neuroblastoma, neurofibroma, acute leukaemia
Other	Paget's disease, fibrous dysplasia, Langerhans cell histiocytosis, Erdheim–Chester disease

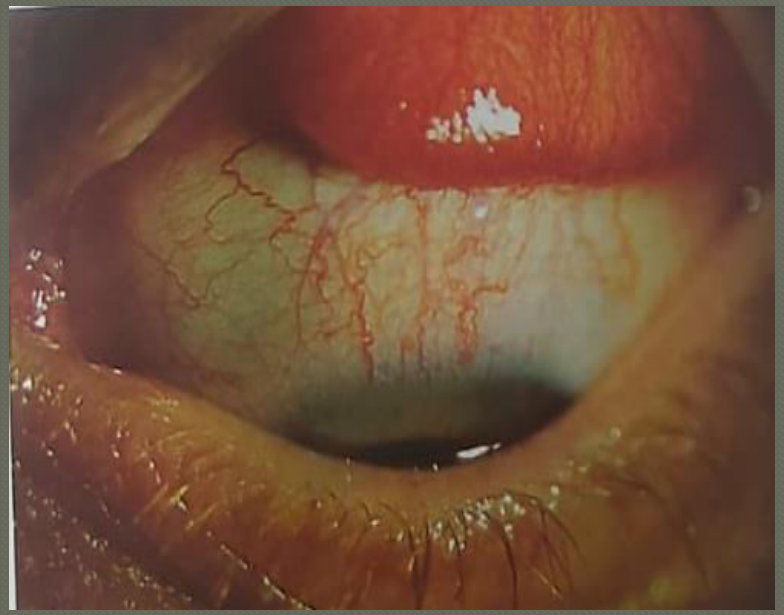
Endocrine

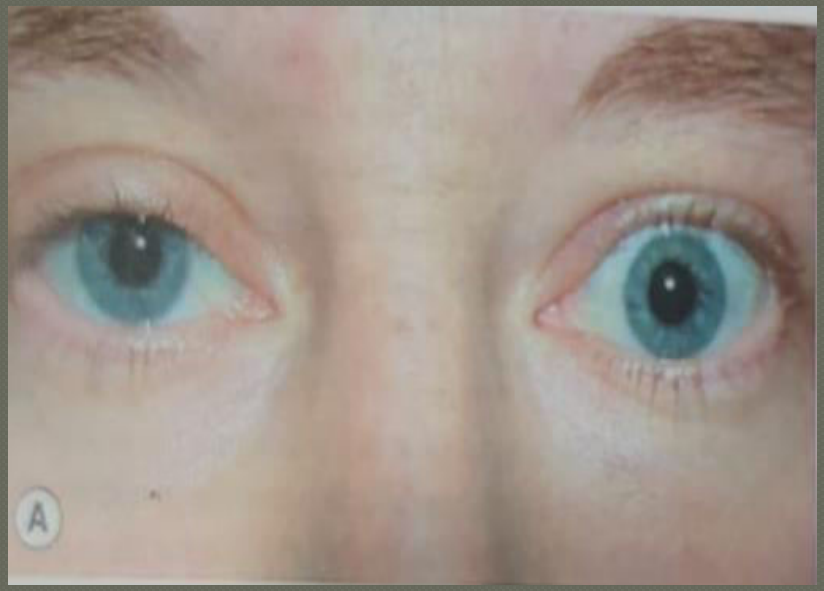
Thyroid eye disease

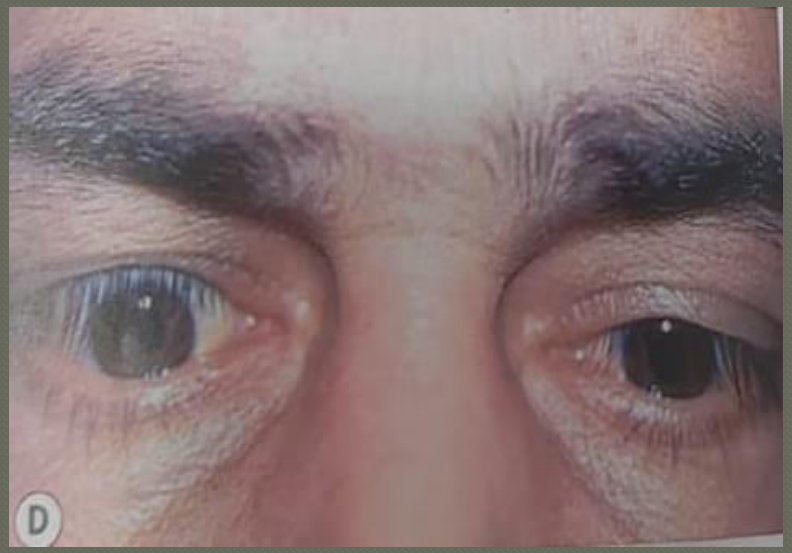
- Most common cause of proptosis in adults
- Axial proptosis
  - Unilateral
  - Bilateral
- Lid retraction and lid lag
- Injection over horizontal recti

- Superior limbic kerato conjunctivitis
- Restricted ocular motility defects
- Signs of optic nerve compression
- Chorio retinal folds


B



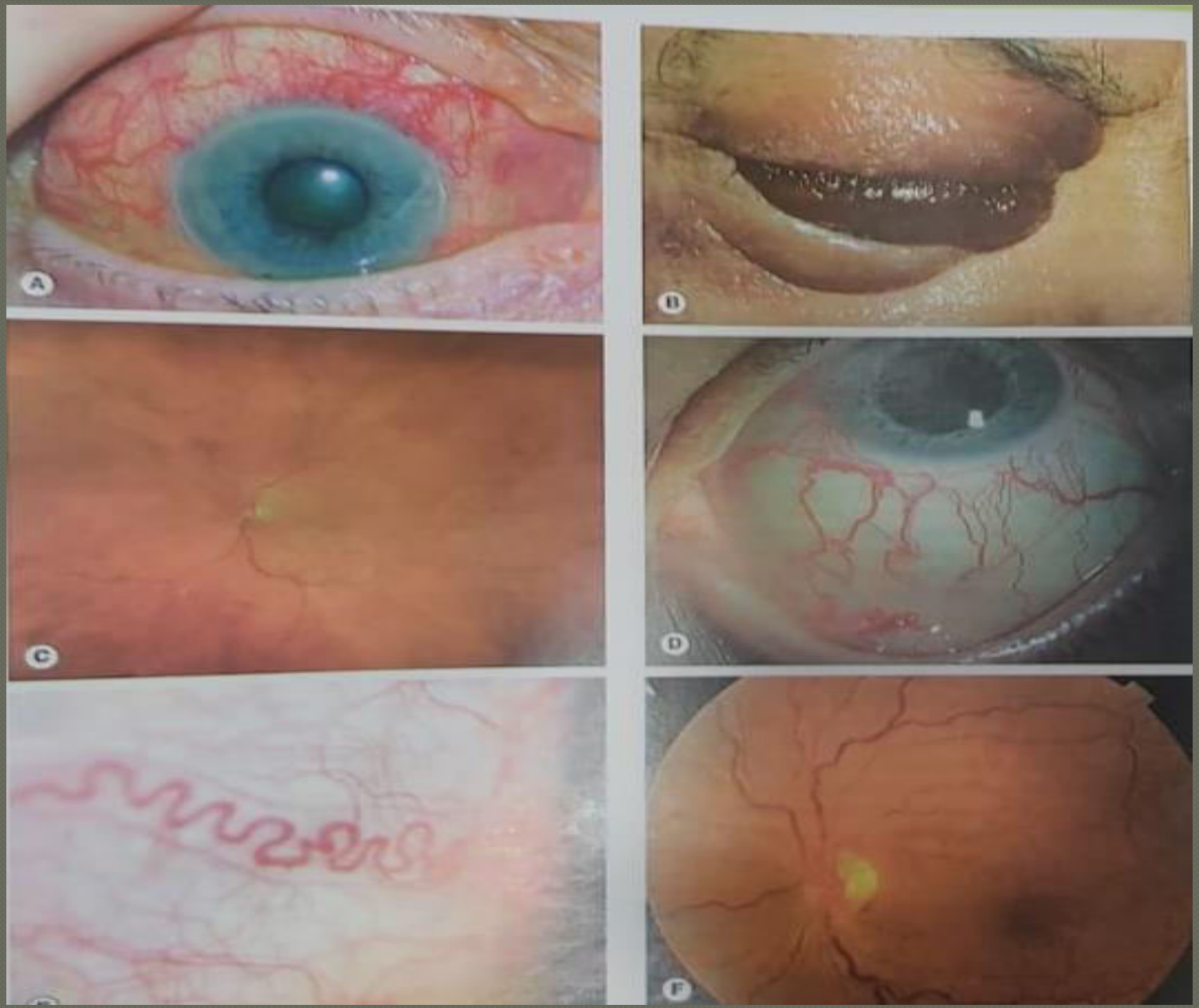

A


D

Vascular

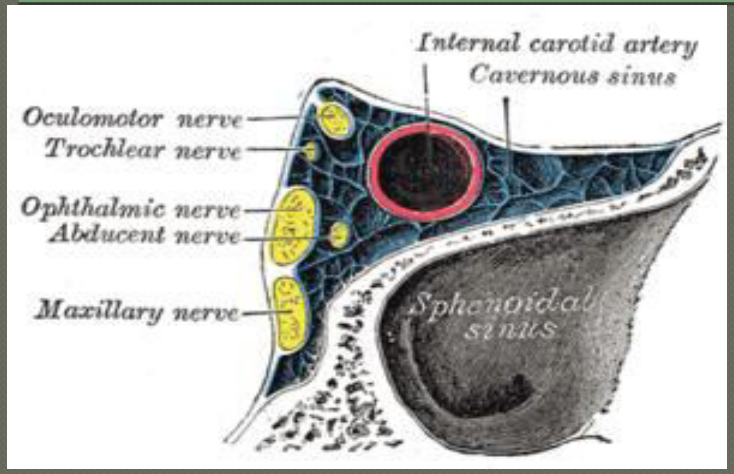
Carotid – cavernous fistula

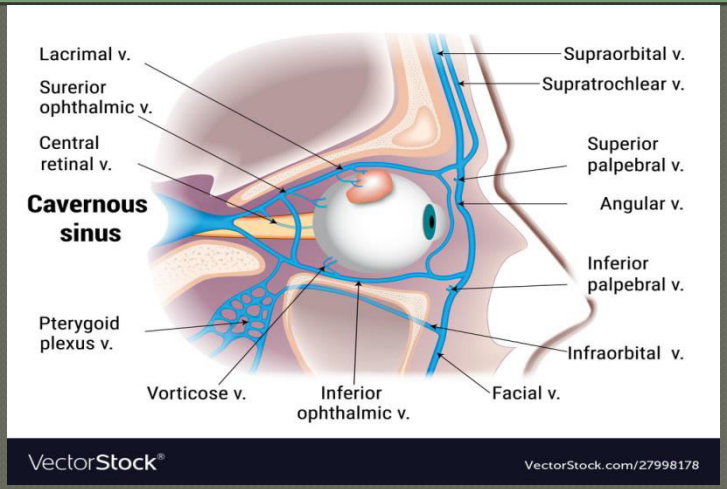
- Caused by either head trauma or a spontaneous rupture of an intracavernous aneurysm
- Unilateral painful pulsatile proptosis associated with a bruit
- Grossly dilated epibulbar vessels
- Ophthalmoplegia


A
B
C
D
F

Cavernous sinus thrombosis

- A serious condition
- Most commonly secondary to skin or paranasal sinus infection such as sinusitis, orbital or preseptal cellulitis or otitis.
- Similar to a CCF except that the patient is usually more ill because of systemic infection.
- Patient have severe headache, vomiting, unilateral or bilateral proptosis.
- Congestion of conjunctival or retinal veins, reduced vision, 3rd to 6th CN palsy.





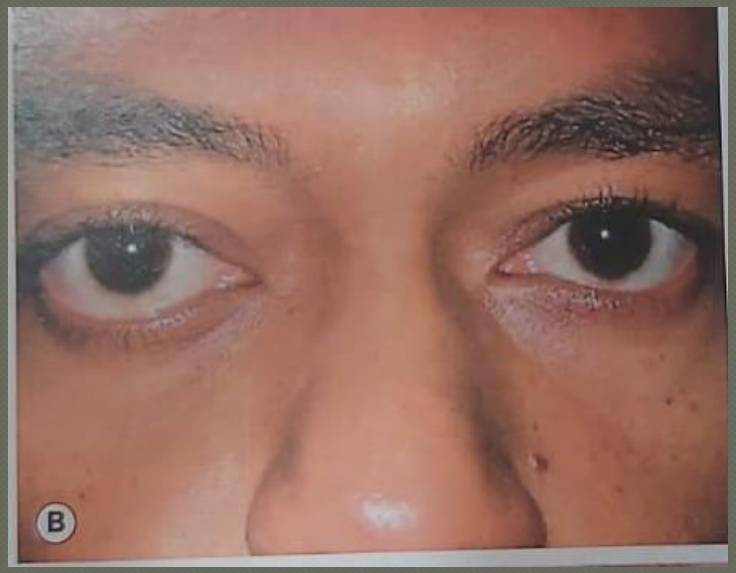
Clinical features

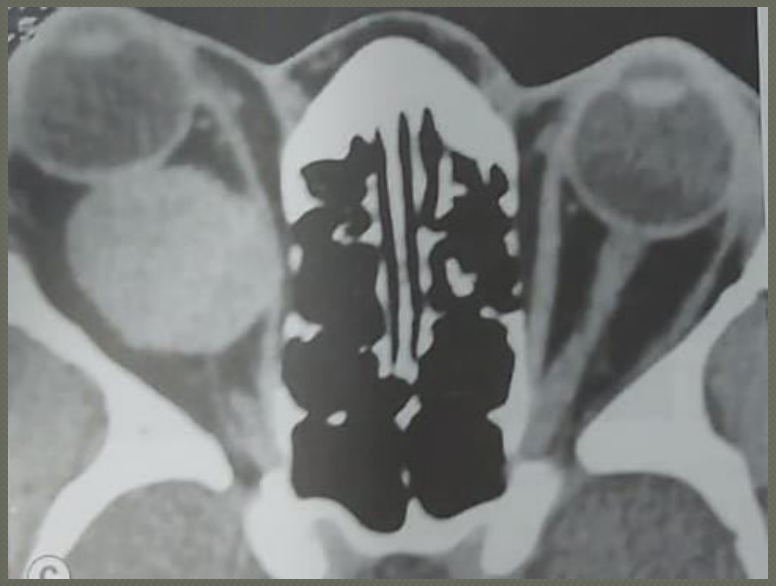
- Infections from face, orbit, sphenoid sinus can cause thrombosis.
- III,IV,V1,V2,VI can be involved resulting in various diplopias
- Rupture of ICA can cause pulsatile exophthalmos

Caput medusae in cavernous sinus thrombosis

Cavernous hemangioma

- Presents during the 4th & 5th decades of life.
- Most commongly encountered benign orbital tumor in adults.
- Unilateral, usually axial, slowly progressive proptosis.
- Look for hypermetropia and chorioretinal folds.

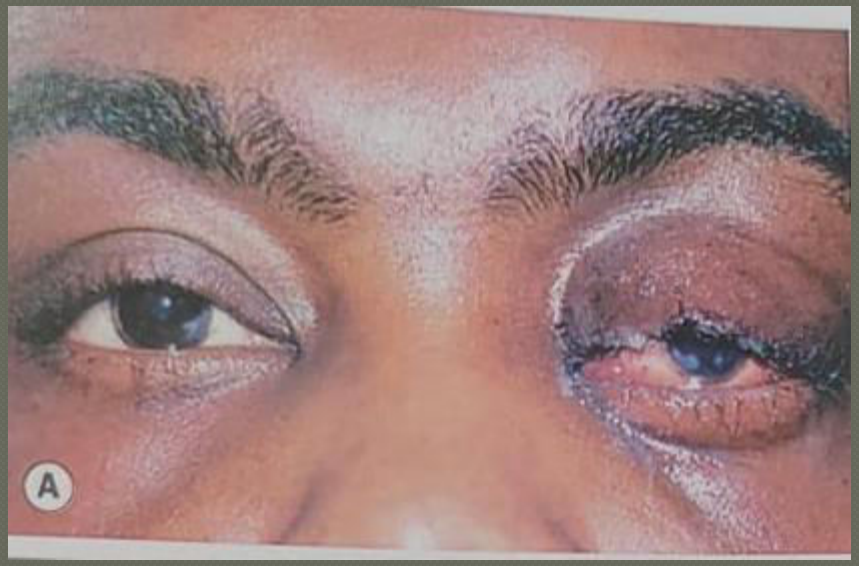


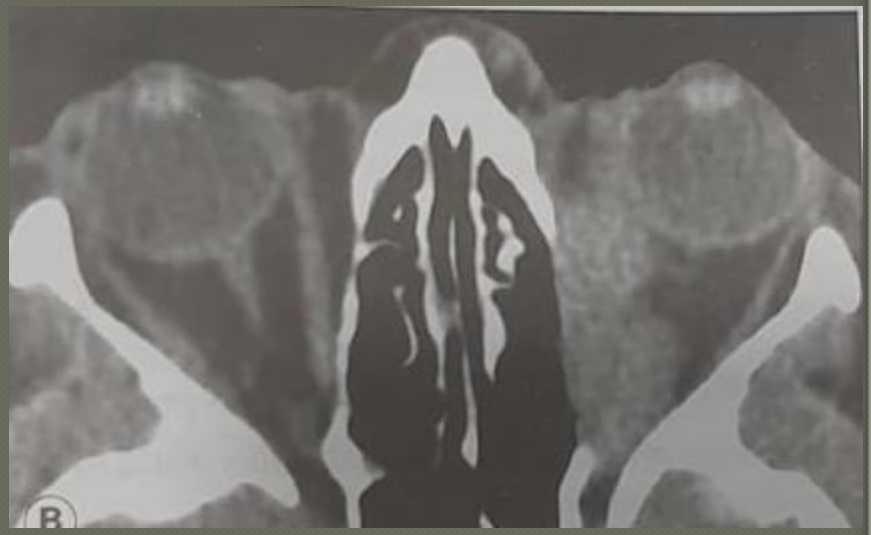


Inflammatory

Orbital Pseudo-tumor

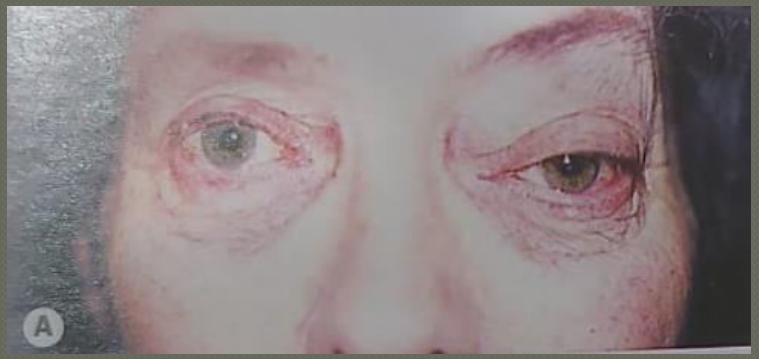
- Presents between ages of 20 & 25 years
- Acute orbital myositis is a type of pseudotumor which primarliy affects one or more of the extraocular muscles.
- Usually unilateral axial proptosis
- In patients with acute orbital myositis, pain and diplopia increases on attempted gaze into the field of the affected muscle

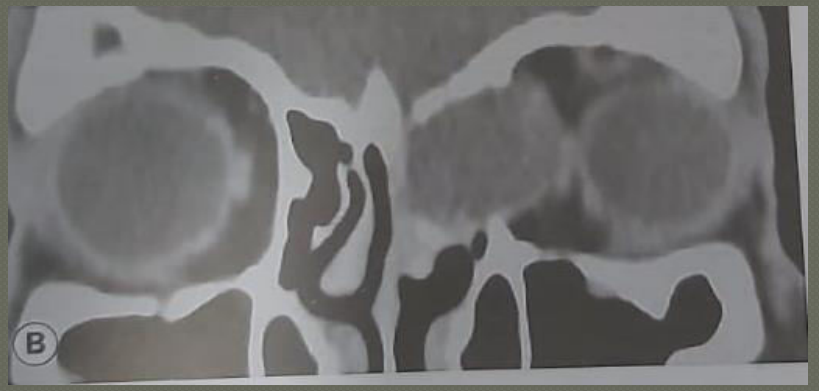

A


B

Mucoceles

- Presents with a combination of ptosis, proptosis and globe displacement.
- The proptosis may fluctuate when the walls of the mucocele become inflammed.
- Frontal mucocele displaces the globe downward.
- Ethmoidal mucocele causes lateral displacement.





Infective

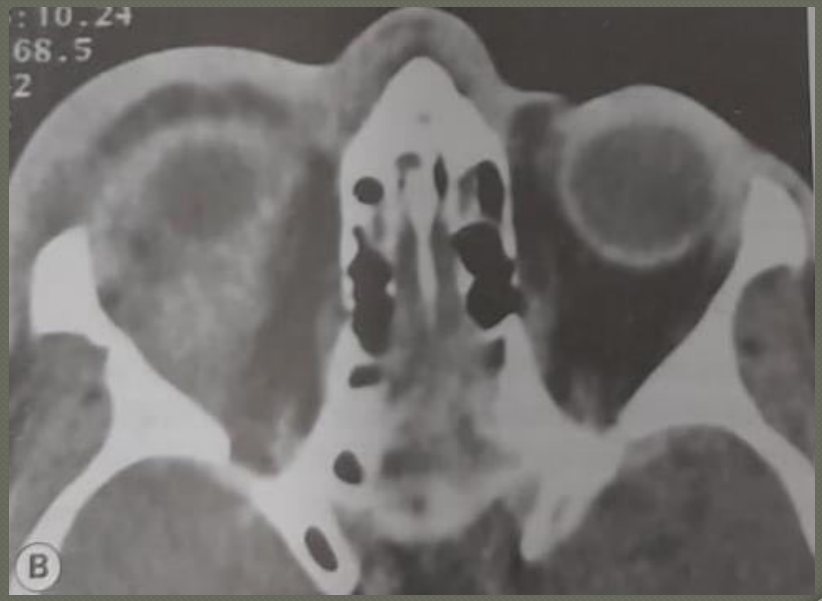
Orbital cellulitis

- There are four types of orbital cellulitis
  - Associated with sinus infection
  - From adjacent structures (e.g dacryocystitis, midfacial septum
  - Post-traumatic in injuries which penetrate the orbital septum.
  - Post-surgical (e.g retinal detachment, strabismus, lacrimal and orbital surgery).

- Unilateral painful proptosis and severe lid oedema in a very unwell patient.

Ophthalmoplegia

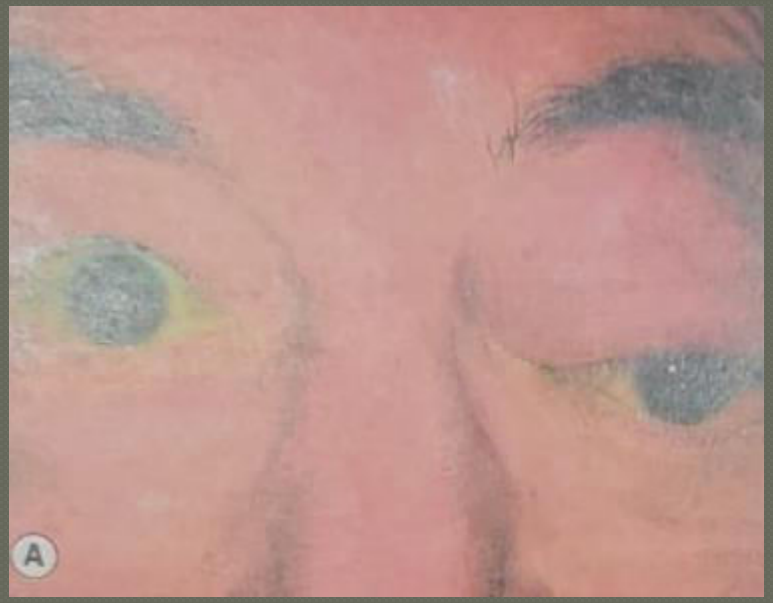
Look for signs of optic nerve compression.

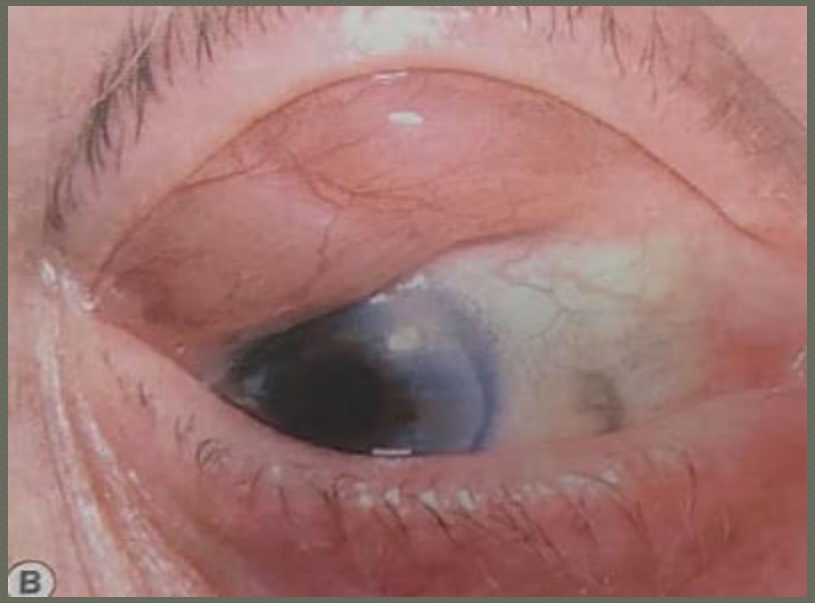


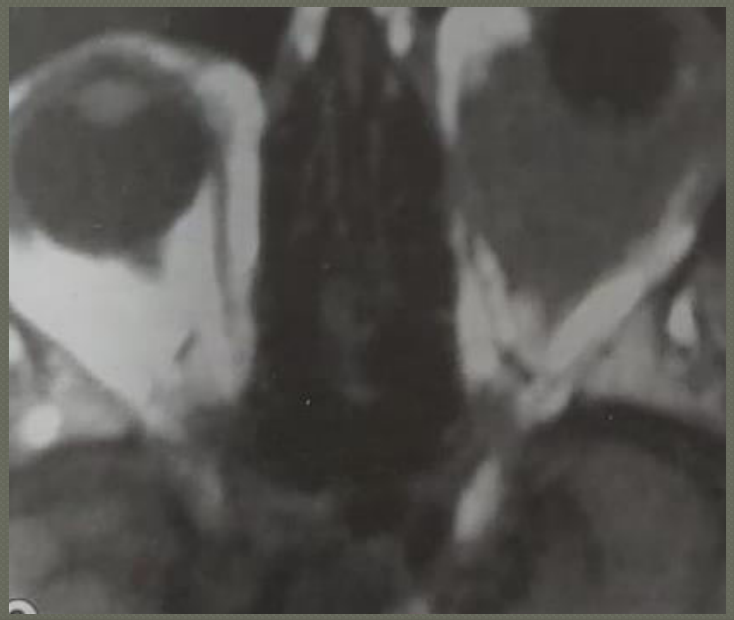
Tumours

Lymphoma

- Presentation of lymphoid tumors is usually in old age.
- Unilateral or bilateral involvement with periorbital puffiness
- Anteriorly located lesion have rubbery consistency
- Conjunctival extensions may be present.


A

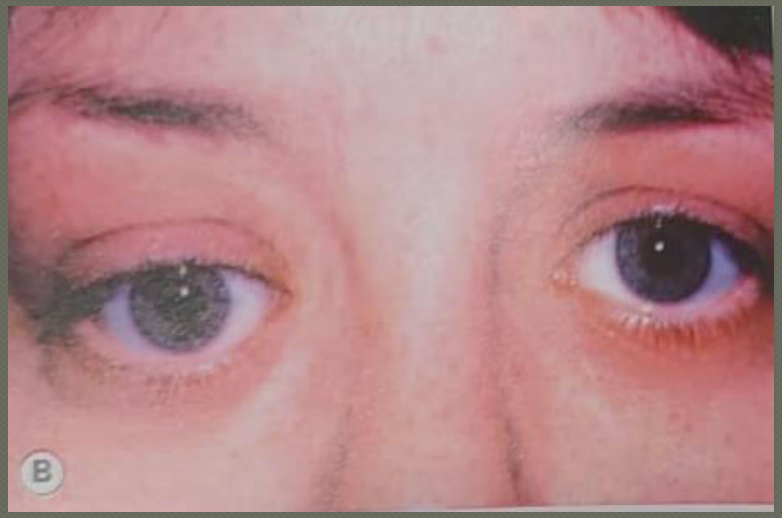

B

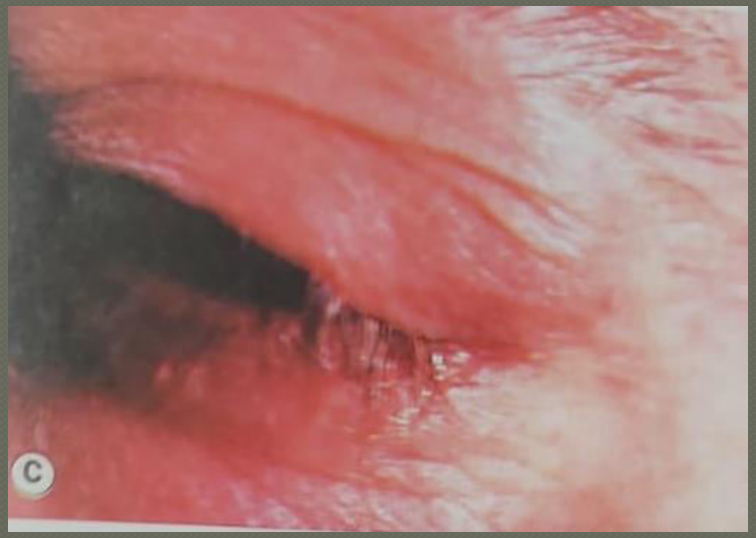


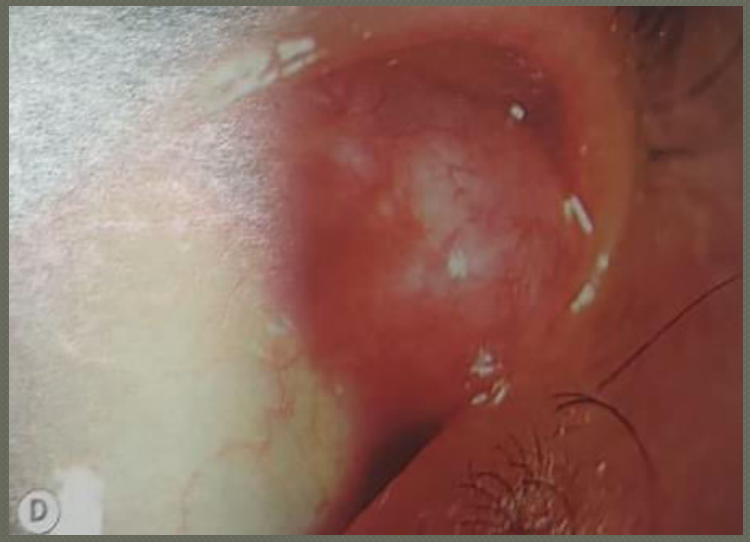
Lacrimal galnd tumors

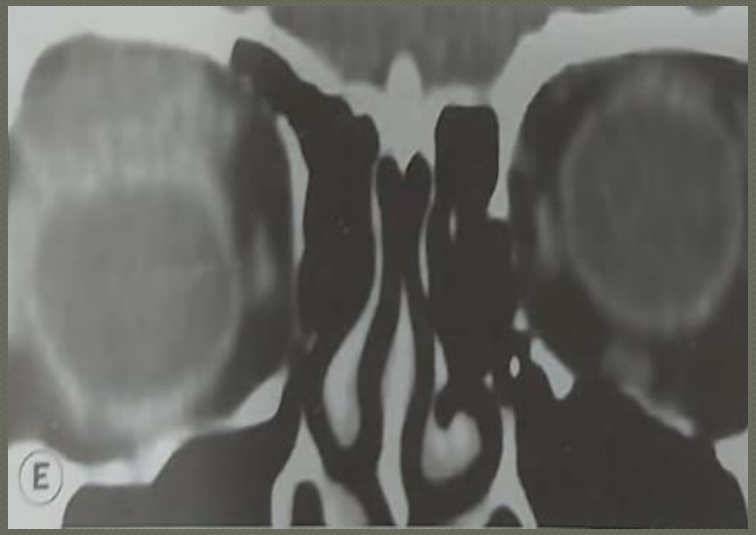
Pleomorphic adenoma

- Presentation is usually in middle age
- Chronic, painless fullness of the eyelid
- Displacement of the globe downward and inward.
- Diplopia when gaze is directed towards the lesion i.e upward and outward.


B

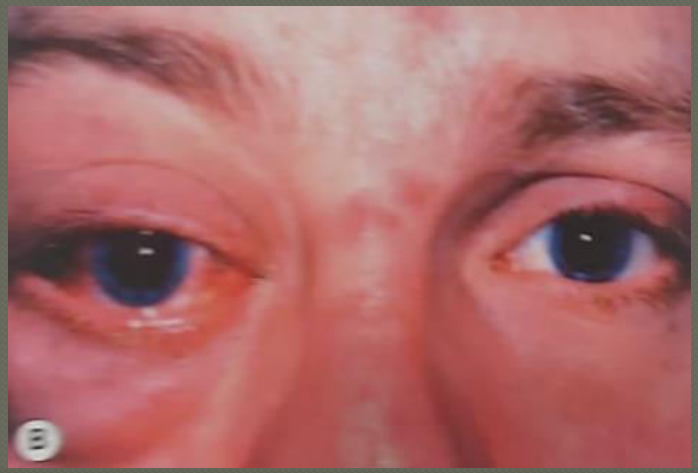

C

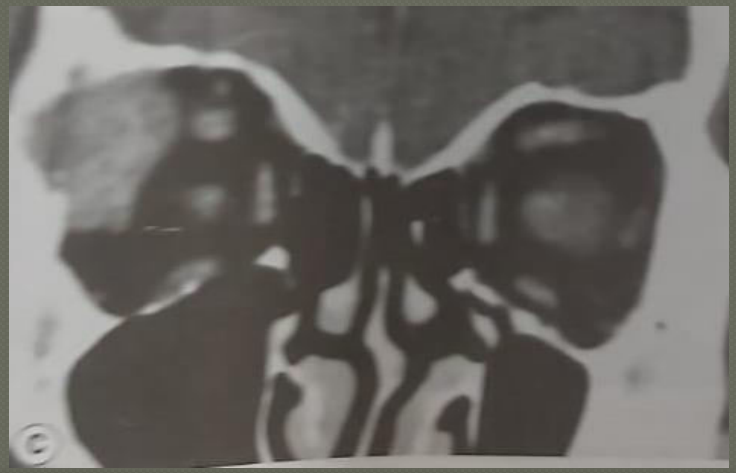

D


E

Malignant lacrimal gland tumors

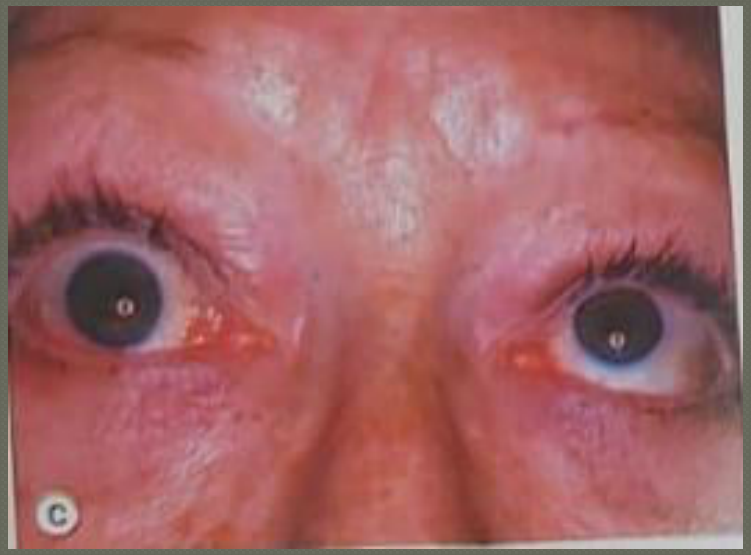
- Have shorter history than benign tumors
- Associated with pain and diplopia
- Pleomorphic adenocarcinoma occur in old age
- Adenoid cystic carcinoma occur around 40 years.

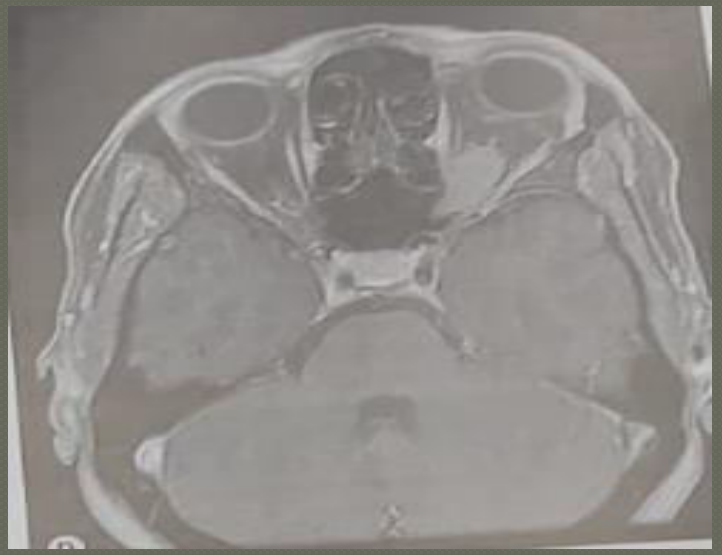


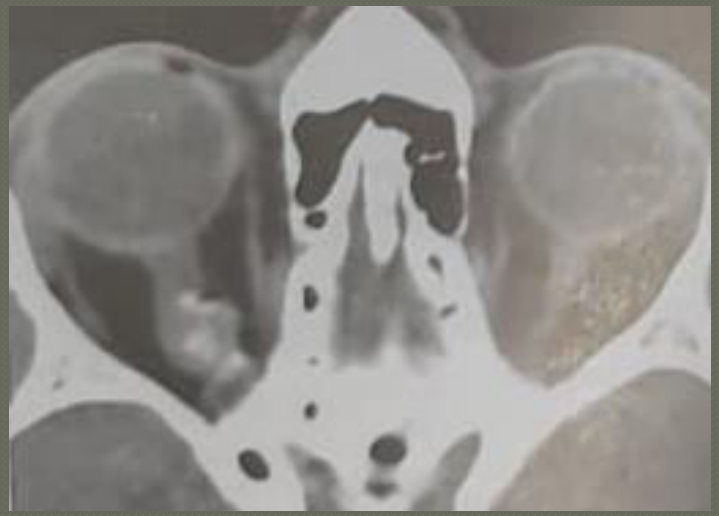


Optic nerve sheath meningioma

- Presents usually in the middle age
- Female to male ratio is 3:1
- Unilateral, slowly progressive axial proptosis
- Early decrease in VA
- There may be optic atrophy and opticociliary shunts

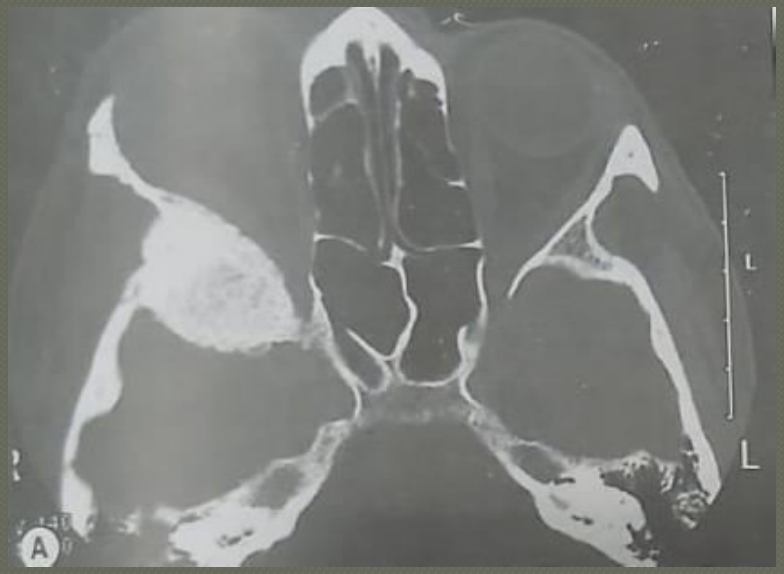






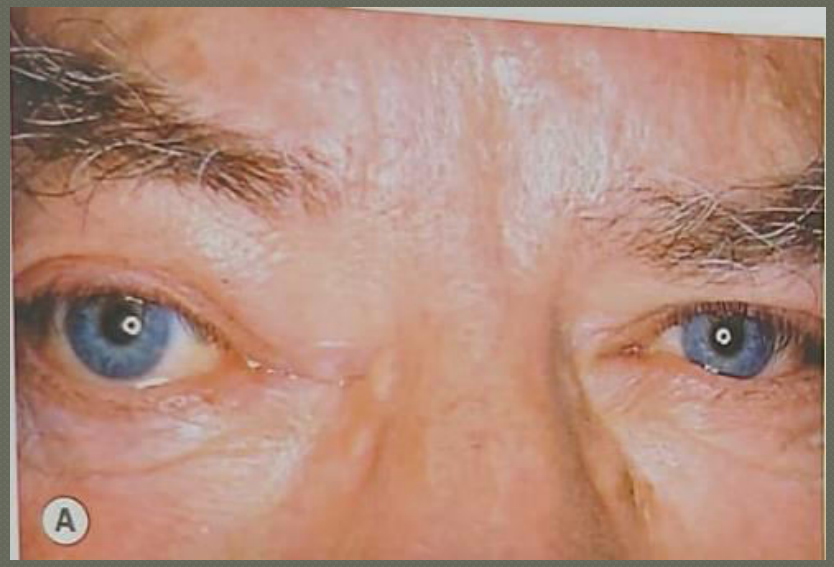
Sphenoidal ridge meningioma

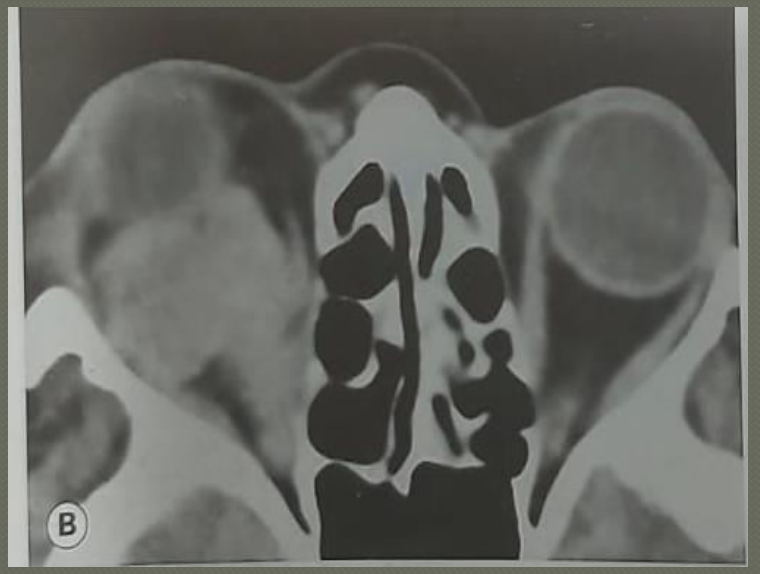
- Presents in middle age with proptosis and reactive hyperostosis
- Slowly progressive painless, downward and outward proptosis
- Fullness of temporal fossa
- Optic nerve dysfunction


L
L
A

Metastatic tumors

- Presentation is usually with a rapid onset of diplopia, lid edema and pain
- Most metastases from tumors cause proptosis while secondaries form scirrhous breast carcinoma give rise to enophthalmos.


A


B

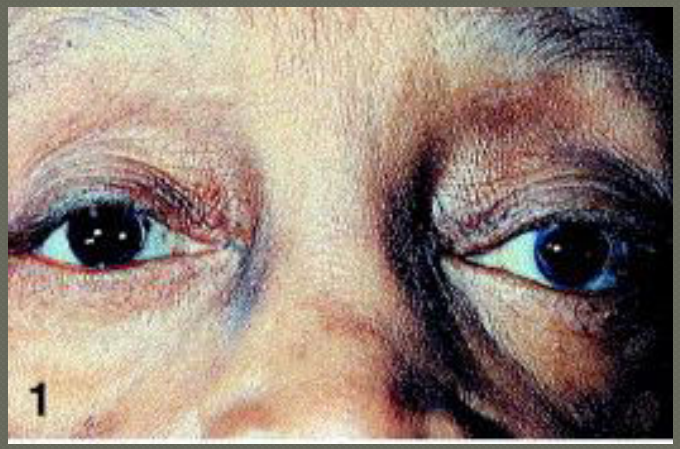
Isolated neurofibromatosis

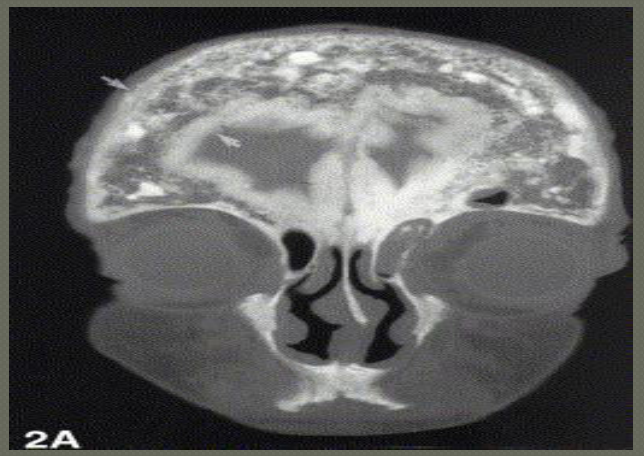
- 10% of patients have NF1
- Presentation is in the 3rd to 4th decade
- Insidious mild painful proptosis
- Usually not associated with visual impairement or ocular motility dysfunction.

Other

a) Paget disease of the skill

- May cause slowly progressive unilateral or bilateral proptosis.
- Ocassionaly patients develop osteosarcoma





Thanks