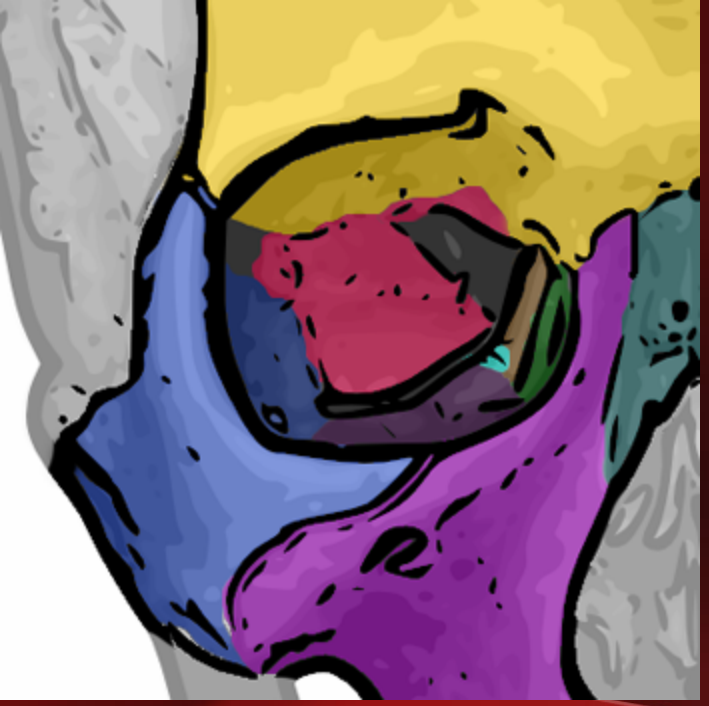


Orbital tumors

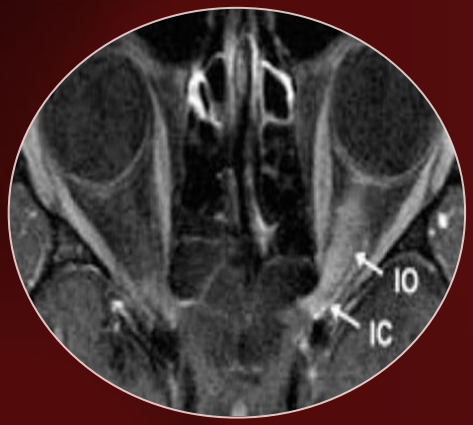
Clinical evaluation

"Six p's" of orbital lesions:

- Proptosis
- Pain
- Progression
- Pulsation
- Palpation
- Periorbital changes

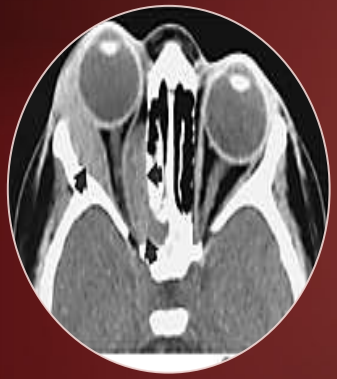
- Early Vision loss
- Papilledema
- Opticocilliary shunts
- Proptosis +/-

Intracanalicular



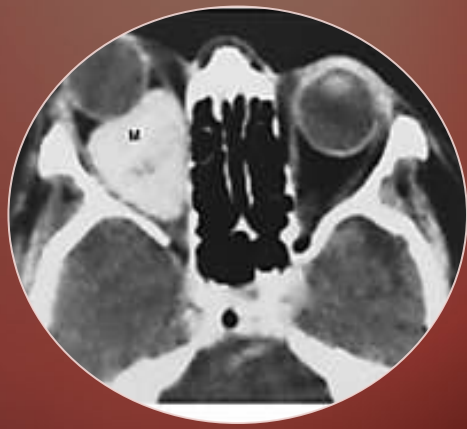
- Proptosis
- Vision loss late

Extraconal



- Vision loss
- Impairment of motility
- Axial proptosis

Intraconal



Surgical indications

- Biopsy
- Lesions affecting vision
- Lesions affecting the globe
- Compression of the optic nerve

Complications

- ***PTOSIS*** : levator muscle & / or its nerve damage
- ***DIPLOPIA*** **:** EOM damage, ocular motor nerve damage, adhesions of EOM, trochlea damage
- ***VISUAL LOSS*** : CRA trauma / occlusion, globe compression, optic nerve trauma / compression (Hemorhage, edema)
- ***CSF LEAK*** : inadvertent opening of the paranasal sinuses(post ethmoid) while optic canal deroofing.
- ***EYELID MALPOSITION*** : faulty wound closure, adhesions b/w lids & orbital rim
- ***PUPIL & ACCOMODATION ABNORMALITIES*** : Posterior ciliary N & vessels damage

- ***PULSATING PROPTOSIS*** :
  Due to extensive deroofing of the orbit

- ***FRONTAL BRANCH OF FACIAL NERVE INJURY*** :
  Incision >4cm from the lateral canthal margin in lateral orbitotomy

- ***OCULAR OR FACIAL SENSORY LOSS*** :
  sensory nerve damage (nasociliary N, 1^{st}/2^{nd} division of trigeminal N)

- ***CORNEAL ULCERATION*** : direct corneal trauma, corneal dessication

Classification

- Primary / Secondary / Metastatic
- Intraconal / Extraconal / Intracanalicular
- PATHOLOGICAL
  - Cystic : dermoid / epidermoid
  - Vascular : hemangioma / lymphangioma
  - Inflammatory : pseudotumor
  - Infiltrating : lymphoid tumors / LCH
  - Mesodermal : Fibroma/lipoma
  - Neurogenic: glioma / meningioma
  - Lacrimal : adenoma / carcinoma
  - Metastatic : Neuroblastoma/ Ewings
  - Intraocular : Retinoblastoma

Pediatrics

- Dermoid cysts
- Capillary haemangioma
- Rhabdomyosarcoma

Adults

- Lymphoid tumors
- Cavernous haemangioma
- Meningioma

Cystic lesions

Developmental cysts

Dermoid/Epidermoid

Teratoma

Adjacent structure cysts

Mucocele

Mucopyocele

Dacryocele

Cephalocele

Cystic tumors

Acquired cysts

Epithelial and appendage cysts

Lacrimal duct cyst

Aneurysmal bone cyst

Parasitic cysts: Hydatid, Cysticercus cellulosae

Chocolate cyst

Cholesterol granulomatous cyst

Orbital abscess

Cystic lesions

- The most common space-occupying masses in the orbit, representing 30% - 46% of excised orbital tumors in children
- Frequently located anterior to the orbital septum along the fronto-zygomatic suture
- Small cysts: close observation
- Large cysts: excision in toto

Vascular lesions

- Approximately 15% of cases in several series
- Capillary hemangioma
  - MC vascular orbital tumor in cildhood
  - Spontaneous involution
  - Vision preservation dictates management
  - Observation/Steroids/ Co2 laser/interferon alpha
- Cavernous hemangioma
  - Adults
  - Well circumscribed
  - Surgical excision
- Lymphangioma
  - 1-3 %
  - Slowly progressive
  - Soft bluish mass superonasal quadrant
  - Bleeding – chocolate cyst
  - Steroids / surgical debulking

Neurogenic tumors

- Gliomas
- Meningioma
- Neurofibroma
- Schwannoma
- Esthesioneuroblastoma
- Paraganglioma
- Melanotic neuroectodermal tumor of infancy

Optic nerve sheath meningioma

- 2% of all orbital tumors and 1–2% of all meningiomas.
- Primary ONSM:
  - 92% intraorbital nerve sheath
  - 8% are intracanalicular in origin.
- Bilateral and multifocal presentations : NF2
- Presentation :

Triad : visual loss/optic atrophy/opticociliary shunts

Management

- Recommendations for observation without treatment should be followed only with caution
- Surgery :
  - Functional vision significantly compromised
  - Disfiguring proptosis
  - Intracranial extension
- Stereotactic fractionated radiotherapy : better visual outcome
- Chemotherapy : Unresectable / Recurrent/ Post RT
  - 5 FU, Folate, levamisole

Optic nerve Gliomas

- 3-5% of childhood brain tumors. 11-30% with NF1
- Typically occurs in the first decade of life
- Optic disc and nerve 25%, chiasm 40–75%
- Presentation : vision loss/ proptosis / strabismus / endocrinopathy
- Histologically : (LGG) pilocytic / fibrillary / pilomyxoid astrocytoma
- Biopsy only if unusual clinical / imaging findings.

Imaging

- An enlargement of the optic nerve without calcification, as tubular / fusiform / lobulated
- Classically a J-shaped sella
- Optic foramina views : optic foramen > 7.0 mm or a difference of more than 2.0 mm.
- T2WI demonstrate homogeneous high signal intensity of the affected nerve in contrast to the low signal of the C/L unaffected optic nerve

MANAGEMENT

- Observation: newly diagnosed OPG
- Surgery :
  - Single nerve with disfiguring proptosis / blindness
  - Exophytic chiasm tumor with hydrocephalus / ME
- Chemotherapy : 1st line for symptomatic OPG beyond observation
  - Packer regimen: concurrent carboplatin and vincristine
- Radiation therapy:
  - progressive chiasmatic tumors in > 10 yr age , 45 – 50 Gy

Optic pathway gliomas : a review Neurosurg Focus 23 (5):E2, 2007

PROGNOSIS

- Confined to optic nerve:
  - Treated : 0% tumor-related mortality.
  - Observed: 21% exhibited progression
    5% died 91% maintained stable vision.
- Chiasmatic gliomas :
  42% rate of progression / recurrence
  29 % Tumor related mortality.
- Good prognosis: NF1 and anterior location
- Poor prognosis: younger age at presentation

Peripheral nerve tumors

- Constitute 5 –15 % of the orbital tumors.

- 5 types :
  - Solitary neurofibroma
  - Diffuse neurofibroma
  - Plexiform neurofibroma
  - Schwannomas
  - Malignant peripheral nerve tumors

Solitary neurofibroma

- B/w 3rd – 4th decade
- Slowly progressive, painless proptosis with minimal or no visual dysfunction
- Typically located in the superolateral orbital quadrant
- Isointense to brain & muscle on T1WI & hyperintense to fat onT2WI with heterogenous enhancement
- Pseudocapsule : easy to dissect
- Prognosis is good
- No need for postoperative RT.

Plexiform neurofibroma

- Associated with NF
- Occur mostly in infants & children
- A palpable mass in the eyelid (usually lateral third) with subsequent ptosis & lid hypertrophy
- May spread to forehead or adjacent areas of temple

Schwannomas (Neurilemmomas, Neurinoma)

- 2^{nd} – 5^{th} decade, F>M
- Usually originate from sensory branch of the trigeminal nerve
- High incidence in patients with NF 2.
- Well encapsulated
- C/f : Proptosis, trigeminal distribution of pain & numbness
- T1WI : Iso- to hypointense signal in relation to the orbital fat with a varying degree of contrast enhancement
- Malignant transformation is rare.

Metastatic Tumors

- 8% of all orbital tumors
- Most common in women – breast
- Most common in men – prostate & lung
- Symptoms – proptosis, diplopia, pain, vision loss

- Presents in 7th decade

- FNAB for diagnosis (80%)

- Prognosis is very poor (avg. survival 10 months)

- XRT usual; Chemo and Hormonal occasional

FIBROUS DYSPLASIA

- Normal cancellous bone is replaced by immature woven bone and fibrous tissue.
- 2.5% of all bone tumors.
- Frontal, sphenoid, ethmoid, and maxillary bone complexes
- Sclerotic/ cystic (lytic)/ mixed varieties (40%)of cases.
- classic “ground-glass” appearance on CT.
- Surgery: cosmetic deformity / progressive vision loss

AIIMS Data Search

- 50 Orbital tumors
- Haemangioma: 11, Lymphangioma :2
- Meningioma 6 : ONSM 2
- Pseudotumor: 5
- Lacrimal gland tumor: 5
- Dermoids :3, mucocele: 1
- Metastatic: 3
- Glioma/ haemangiopericytoma/chondrosarcoma / fibrous dysplasia : 2 each
- GCT/ABC/Amyloidosis :1 each

From 2009 OCT– 2011 OCT

THANK YOU