Orbital tumors
“Six p’s” of orbital lesions:

- Proptosis
- Pain
- Progression
- Pulsation
- Palpation
- Periorbital changes
- Early Vision loss
- Papilledema
- Opticociliary shunts
- Proptosis +/-

Intracanalicular

- Proptosis
- Vision loss late

Extracanal

- 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 (Hemorrhage, 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\textsuperscript{st}/2\textsuperscript{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
- Dermoid cysts
- Capillary haemangioma
- Rhabdomyosarcoma

- Lymphoid tumors
- Cavernous haemangioma
- Meningioma
Cystic lesions

**Developmental cysts**
- Dermoid/Epidermoid
- Teratoma

**Acquired cysts**
- Epithelial and appendage cysts
- Lacrimal duct cyst
- Aneurysmal bone cyst

**Adjacent structure cysts**
- Mucocele
- Mucopyocele
- Dacryocele
- Cephalocele

**Cystic tumors**
- Parasitic cysts: Hydatid, Cysticercus cellulosae
- Chocolate cyst
- Cholesterol granulomatous cyst
- Orbital abscess
• 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 childhood
  - 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 / fibrillar / pilomyxoid astrocytoma

• Biopsy only if unusual clinical / imaging findings.
• 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
• Observation: newly diagnosed OPG
• Surgery:
  Single nerve with disfiguring proptosis / blindness
  Exophytic chiasm tumor with hydrocephalus / ME
• Chemotherapy: 1\textsuperscript{st} line for symptomatic OPG beyond observation
  – Packer regimen: concurrent carboplatin and vincristine
• Radiation therapy:
  progressive chiasmatic tumors in > 10 yr age, 45 – 50 Gy

\textit{Optic pathway gliomas: a review} Neurosurg Focus 23 (5):E2, 2007
• 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 on T2WI 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)

- 2nd – 5th 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