OVERVIEW OF ORBITAL TUMORS AND APPROACH TO MANAGEMENT

Dr E M Nyenze

University of Nairobi
Work up for patient with proptosis/orbital tumor

- History
  - Age
  - Pain
  - Progression
  - Periorbital changes
  - History of other tumors
  - Past medical history
• Examination

✓ General examination of the patient, visual acuity, color vision, ?visual fields

✓ Inspection- periocular change displacement, exophthalmometry, ocular movements

✓ Palpation- ?masses- consistency, attachment to underlying structures, tenderness, fluctuation, finger insinuation, retropulstion
• Ascultation – r/ bruit in AV fistulas/malformations. Some high flow vascular tumors can have a bruit
• Examine eyeball- exposure keratopathy, dilated episcleral veins, pupils, fundoscopy disc swelling, optocilliairy shunts, choroidal folds, disc pallor
INVESTIGATIONS

• Systemic: full blood count, urea and electrolytes, ?thyroid function tests, PBF, ?
Bone marrow aspirate
IMAGING MODALITIES

• CT SCAN
• MRI
• ULTRASOUND
• PLAIN XRAY
• ANGIOGRAPHY

• CT Scan
  ✓ Modality of choice in acute setting
  ✓ Very versatile, good spatial resolution
  ✓ Shows bones and calcifications better than MRI
  ✓ Has faster image acquisition
  ❏ Radiation exposure
  ❏ Artefacts from dental fillings
- CT SCAN
- MRI
- ULTRASOUND
- PLAIN XRAY
- ANGIOGRAPHY

- MRI
  - Good resolution of soft tissues, good for orbital apex, intracanalicular part of ON and orbitocranial tumors
  - No radiation exposure
  - Poor spatial orientation
  - Contraindicated in orbital foreign body
  - Expensive
ULTRASOUND
✓ Good for intraocular lesions and orbital foreign bodys
✓ Can measure EOM thickness (thyroid orbitopathy

PLAIN XRAY
✓ Good for screening of fractures, orbital foreign bodies
✓ Not good for soft tissue
✓ Useful for assessment of sinuses

POOR PENETRATION
<table>
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<th>Table 1. Common indications for requesting an orbital CT scan.</th>
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<td>Unexplained proptosis, ophthalmoplegia or ptosis</td>
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<td>Palpable orbital mass</td>
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<td>Pre-septal cellulitis with orbital signs</td>
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<td>Orbital signs associated with para-nasal sinus disease</td>
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<td>Unexplained afferent dysfunction</td>
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<td>Ocular surface or lid tumour with suspected orbital spread</td>
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<td>Intraocular tumour with proptosis</td>
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<td>Orbital trauma</td>
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Axial scan orientation

- 4a lower axial scan
- 4b midlevel scan showing optic canal and SOF
- Upper axial scan showing SOV
Coronal scan orientation

- 5a mid level coronal scans
- 5b coronal scan behind the eye ball
- 5c apical scans
Causes of proptosis

• Classification
  - acute or chronic
  - in children or adult
  - congenital or acquired
  - According to tissue of origin; vascular, neural tumors, mesenchymal tumors, lymphoproliferative tumors, lacrimal gland tumors, secondary orbital tumors and metastatic
ACUTE PROPTOSIS IN CHILDREN

- Orbital cellulitis
- Retrobulbar haemorrhage, lymphangioma with acute haemorrhage
- Rhabdomyosarcoma
- Orbital lymphoma
- Metastatic tumors (neuroblastoma)
- Orbital retinoblastoma
- Sarcomas- ewings, osteosarcoma,,liposarcoma,fibrosarcoma
- Leukemia (chrolomas)
- Idiopathic orbital inflammatory disease
Slowly progressing proptosis in children

- Optic nerve glioma
- Lymphangioma
- Capillary haemangiomma
- Fibrous dysplasia
- Paranasal sinus Mucoceles
- Neurofibromas, schwanomas
- Histiocytic disorders (LCH)
- Orbital varices
ACUTE PROPTOSIS IN ADULTS

- Orbital cellulitis
- Retrobulbar haemorrhage
- Idiopathic orbital inflammatory disease
- Lymphoproliferative diseases
- Sarcomas – fibrosarcoma, liposarcoma, rhabdomyosarcoma
- Lacrimal malignancies especially adenoid cystic carcinoma
- Metastatic lesions especially from breast, bronchogenic carcinoma or prostate
Slowly progressing proptosis in adults

- Thyroid orbitopathy
- Pleomorphic adenoma
- Meningioma
- Cavernous haemangiomas
- Orbital varices
- Lymphoproliferative disorders
- Osteomas/ fibrous dysplasia
- Sinus Mucoceles
- Schwanomas, neurofibromas
- Lymphangiomas
- Histiocytic disorders
Capillary haemangiomas

- Common benign tumors in children
- May be present at birth or in first few weeks of life
- Commonly superficial looking bright red soft with dimpled texture, deep look blue
- Most regress spontaneously

- Rx: observe, steroids, oral propranolol, ?timolol drops, rarely surgery
rhabdomyosarcoma

- Commonest primary orbital malignancy in children
- Average age 8-10 years
- Presents with acute proptosis
- Biopsy usually indicated. Incision biopsy if large lesion
- Cross striations seen on microscopy
- Electron microscopy and immunohistochemistry can be useful in workup

• Rx. Chemotherapy and radiotherapy. Exenteration for recurrent cases
Lympoproliferative disorders

- Mainly B cell Non hodgkin lymphomas
- May be: low grade (MALT, CLL and folicular centre )or :high grade( large cell lymphoma, Burkitt lymphoma and Lymphoblastic lymphoma)
- Usually painless mass in anterior orbit may have salmon patch appearance
- On CT scan usually mold to surrounding structures
- Incision biopsy for histology and molecular analysis for clonality done
Lympoproliferative disorders

• 20-30% of orbital lymphoma have history of systemic dse and additional 30% of developing in 5yrs
• Localised lesions treated with radiotherapy
• Systemic and aggressive lesion require chemotherapy and radiotherapy
Pleomorphic adenoma

- Commonest epithelial tumor of lacrimal gland
- Usually at 4th to 5th decades of life affecting slightly more men than women
- A firm lobular mass usually palpable supralateral orbit
- Usually well defined on CT
- Histologically benign epithelial cells with cartilagenous, mucinous or osteodysplasia

Rx complete removal. Recurrence 32% if capsule damaged with 10% risk of malignant transformation per decade
Adenoidcystic carcinoma of the lacrimal gland

- commonest malignancy of lacrimal gland
- 4-5th decades
- Can follow incomplete excision of pleomorphic adenoma
- Rapidly growing, pain early due to perineural invasion

Rx; enblock excision ± radiotherapy

CT; Poorly defined mass with bony erosions
Hist; cells growing in nests/tubules or cribriform Swiss-cheese pattern
Optic nerve glioma

- Benign lesion especially in children
- Approx 50% assoc with neurofibromatosis
- Gradual, painless unilateral proptosis assoc with visual loss and RAPD
- Optic atrophy common
- Hist; fusiform intradural lesion
- CT; fusiform enlargement of optic nerve may be kinked

Rx: observation, surgical excision, radiotherapy, chemotherapy
**meningioma**

- Invasive tumors from arachnoid villi
- May affect orbit from sphenoid wing, superior orbital fissure or optic canal
- Can also be primary optic nerve tumor
- Sphenoid wing meningioma causes hyperostosis of involved bone with temporal fullness
Primary orbital meningioma

- Usually affects ON
- Usually women in 3rd - 4th decade
- Slow proptosis with late visual loss
- ON may be normal, atrophic or swollen or may have optociliary shunts
- CT tubular enlargement of ON some times with tramtracking

- Rx observe, subtotal excision, radiotherapy,
Cavernous haemangiomas

- Most common benign orbital lesion in adults
- Affects women > men 4th to 5 decades
- Slowly progressing axial proptosis
- CT scan; well defined intraconal mass highly enhancing with contrast
- Hist: well encapsulated lesion with vascular spaces

- Rx: complete excision
lymphangioma

- Starts in first decade
- May increase in upper respiratory infections
- Acute proptosis may also occur after acute haemorrhage
- Hist; large serum filled channels lined by flat endothelial cells
- CT poorly defined heterogenous lesions

- Rx observation, excision, aspiration of haemorrhage, sclerosant injection
Orbital squamous cell carcinoma

- Usually from conjunctival SCC (OSSN), eye lid SCC or from surrounding structures especially sinuses
- Usually associated with HIV
- Can spread to surrounding structures, Lnodes
- Rx exenteration, radiotherapy, extended exenteration ± neck dissection if LN involved
Surgical spaces of the orbit

- the intraconal space (central space)
- the extraconal space (peripheral space)
- The subperiosteal space
- The episcleral space
Anterior orbitotomy approaches

- A- older stallard wright lateral
- B-eye lid crease lat
- C- canthotomy lat
- E-transcaruncular med
- F-frontoethmoidal med
- G- Vertical eyelid split
- H- Medial bulbar conj
- I- lateral canthototomy lat
- J- Subciliary inferior
- K-transconjunctival inf
- L- Lateral bulbar conj
Aproaches to deep orbital structures

• Lateral orbitotomy with osteotomy
• Transcranial orbitotomy
• Trans caruncular