Surgical treatment of orbital lesions

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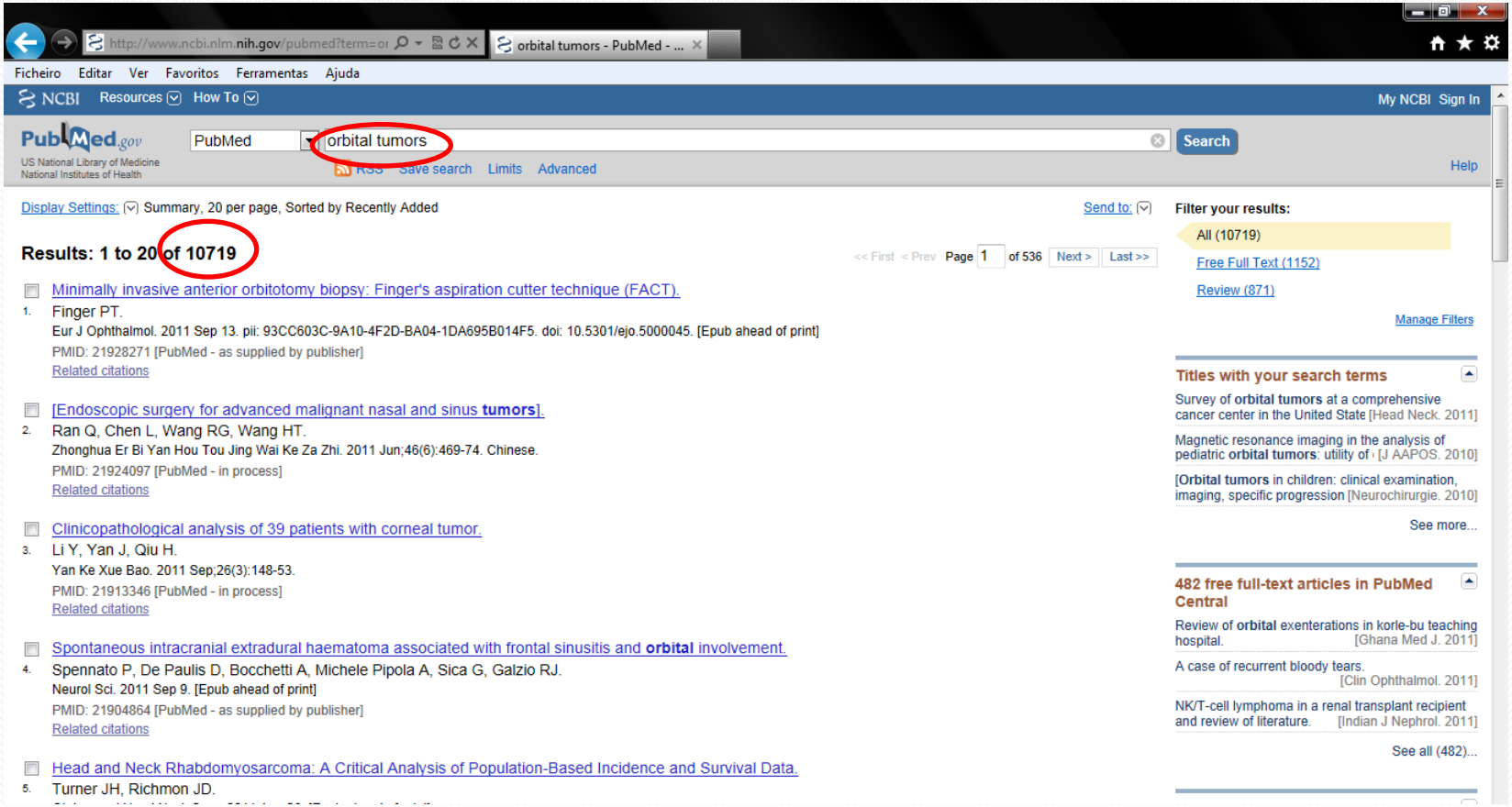
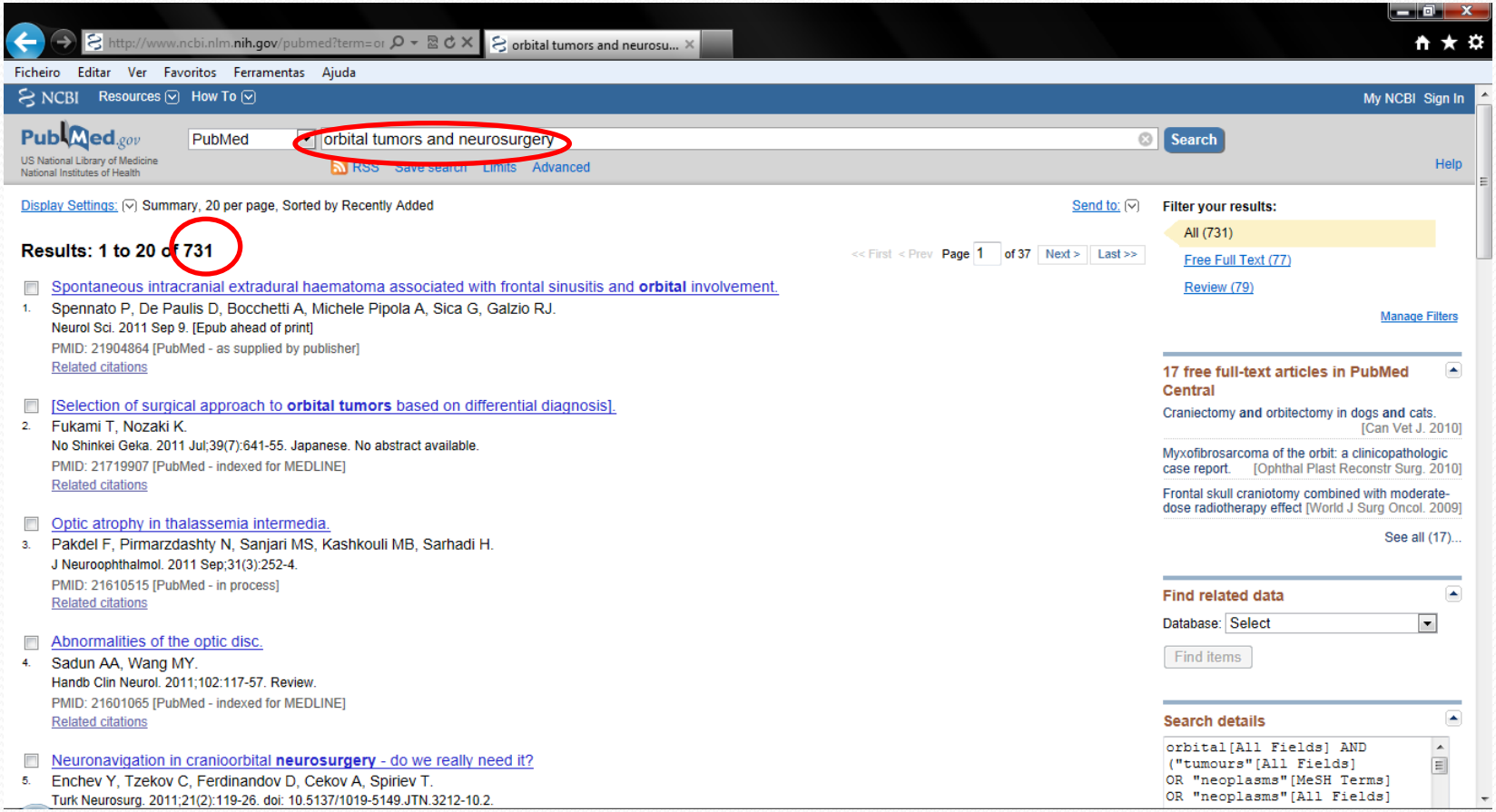
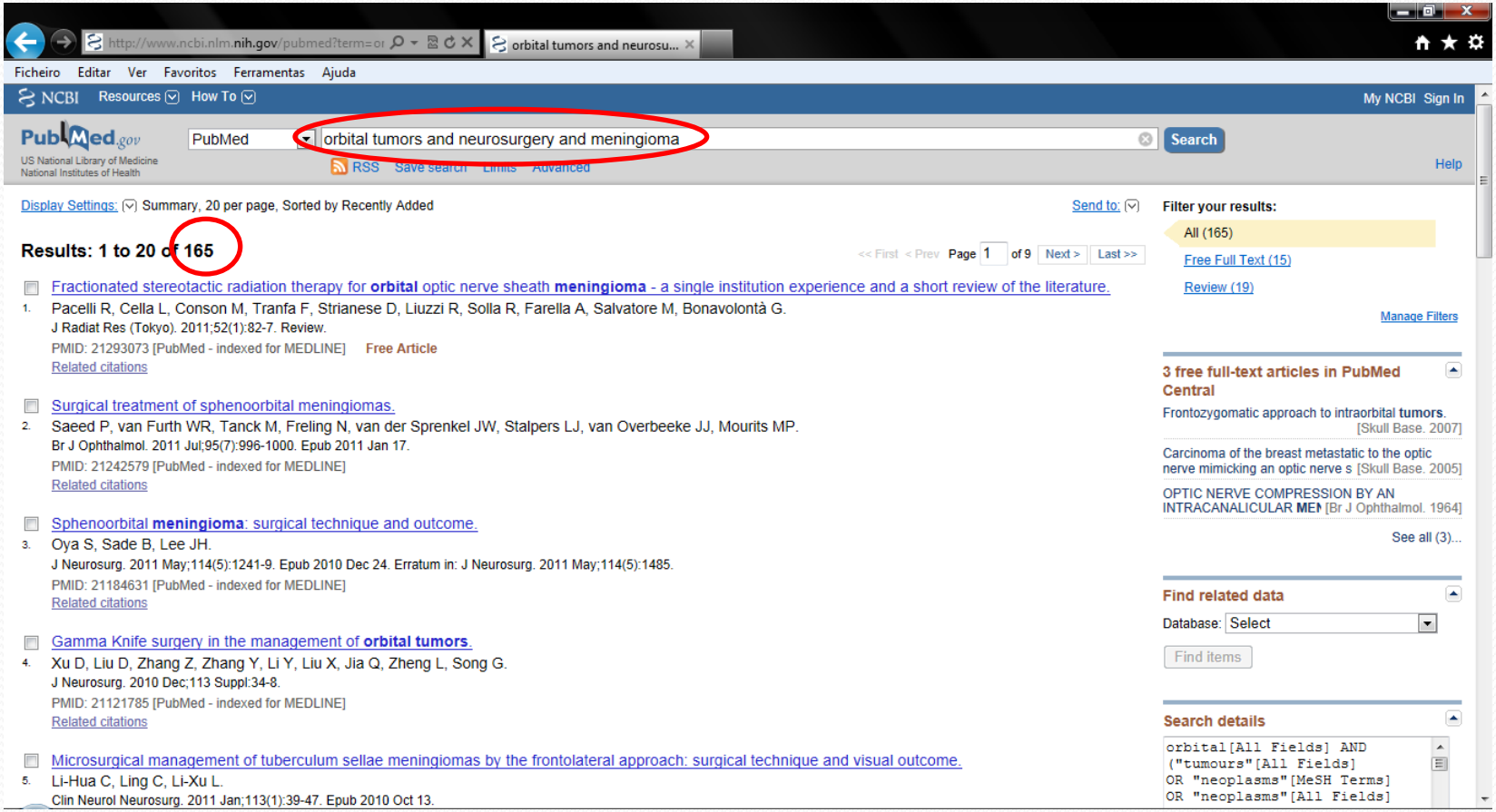
Introduction



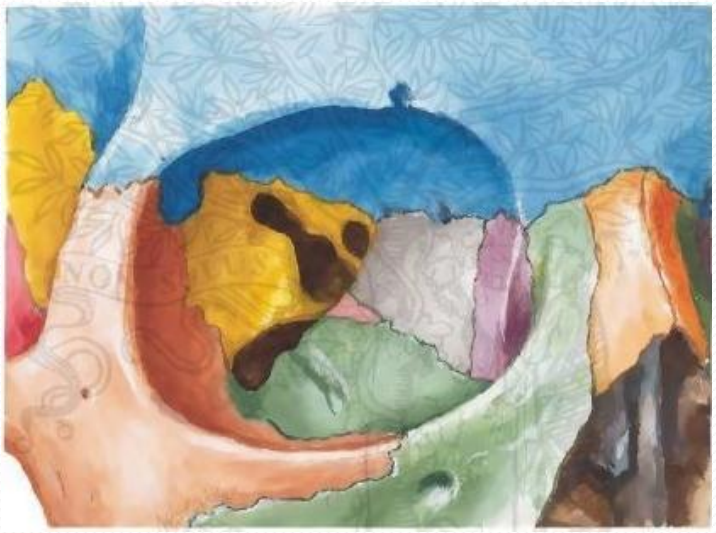
Introduction



Introduction



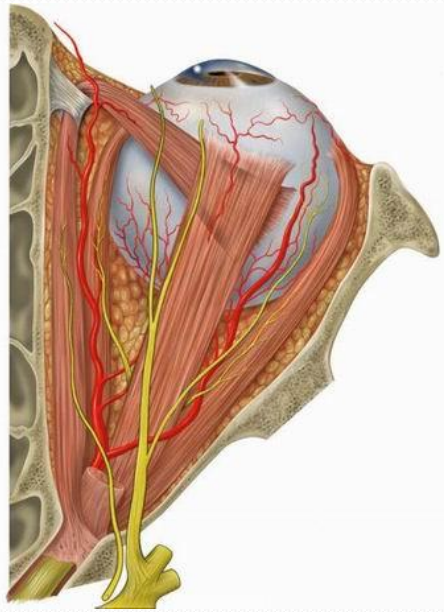
Anatomy



- Mesenchymal cells
- 7 bones

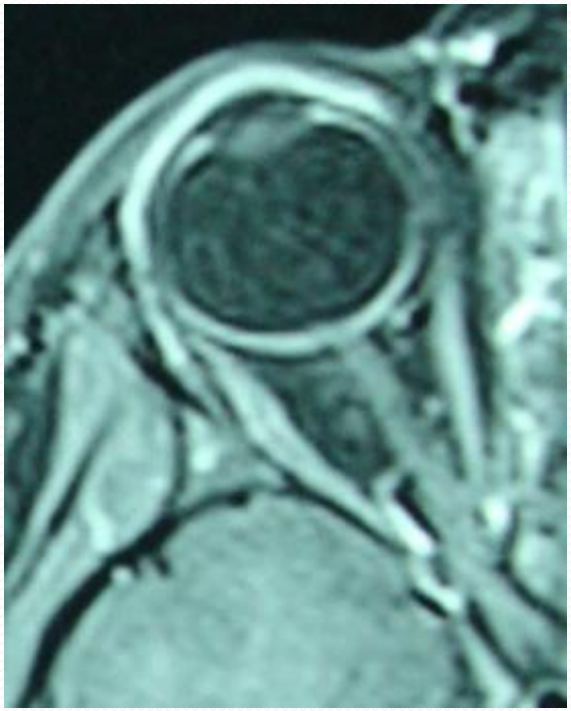
- 30 ml volume
- 45 mm medial lenght
- 40 mm width
- 35 mm height

Anatomy



- Complex structure
  - globe
  - extraocular muscles
  - fat
  - vascular, nerve, glandular, and connective tissues

Anatomy

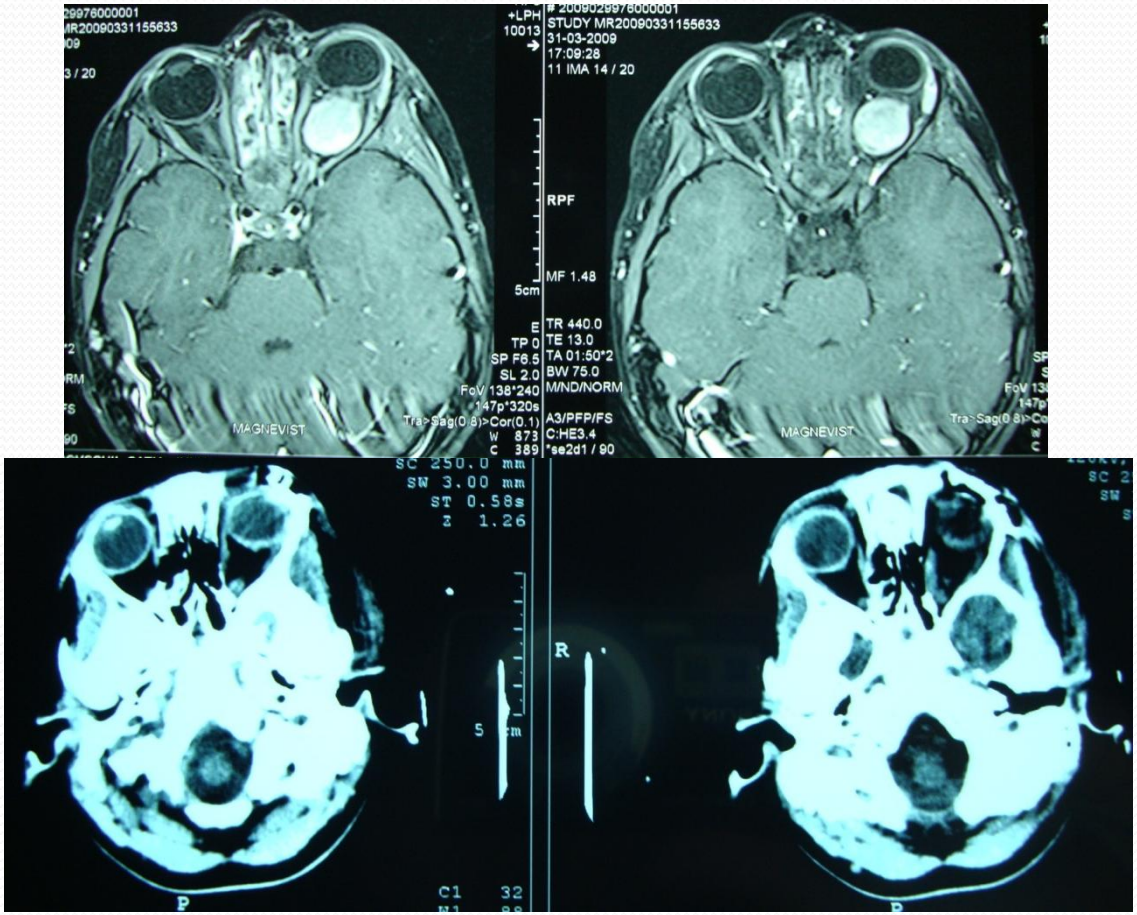


- Anterior compartment
  - lids
  - lacrimal apparatus
  - anterior soft tissues
- Posterior compartment
  - intraconal space
  - cone
  - extraconal space

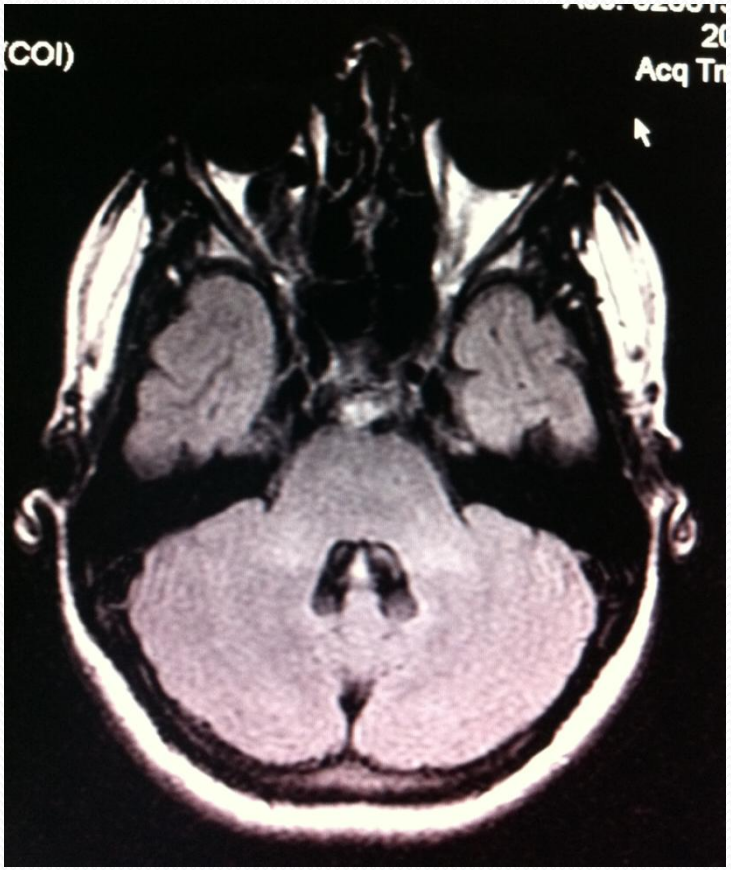
Optic nerve gliomas

- Abnormal proliferation fibrillary astroglial cells
- Most common orbital tumor NF1, 2nd children, 5th adults
- Most are pilocytic astrocytomas
- 50% diagnosed < age 5 y
- Presentation: exophthalmos, ↓ visual acuity, blindness

II CN pilocytic astrocytoma



II CN ganglioglioma

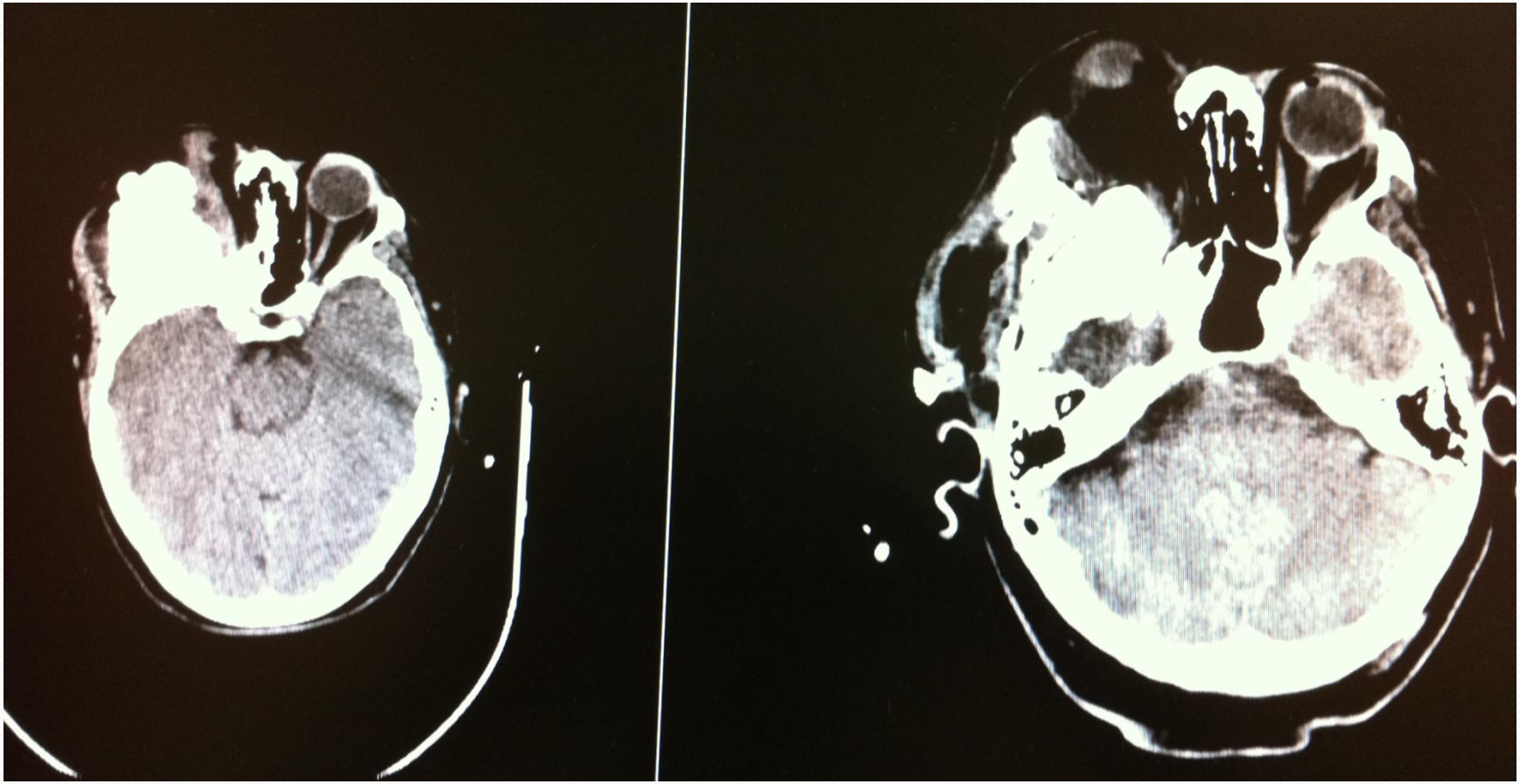


Meningiomas

- Primary
- Secondary (medial sphenoid wing, tuberculum sellae, eg)

- Comprise 10% intraorbital tumours
- : = 3:1
- Presentation: exophthalmos, ↓ visual acuity, pallor optic disc, visual field deficits. Orbital pain or headache (uncommon)

Meningioma



Meningiomas

- Optic nerve sheath meningiomas
  - rare
  - 2% intraorbital tumours
  - 1-2% meningiomas
  - slow growing
  - untreated: progressive visual decline → color blindness → complete vision loss
  - good vision: FSR (improving/stabilizing vision)
  - surgery: intracranial tumour (prevent contralateral extension)

Tumours arising from neuronal structures and their coverings within the orbit

- Nerve sheaths tumours (15%)
  - neurofibromas
  - schwannomas
  - malignant peripheral nerve sheath tumours

- Do not involve II CN (lack schwann cells)
  - develop from peripheral motor nerves (extraocular muscles)
    - V1, V2, sympathetic/parasympathetic fibers

Tumours arising from neuronal structures and their coverings within the orbit

- Neurofibromas
  - Solitary
    - single, encapsulated, not associated NF
    - goal: complete resection
  - Diffuse
    - NF, multiple, may involve orbital tissues
  - Plexiform
    - pathognomonic NF1, involve nerves, total resection difficult
    - subtotal resection (cosmetic deformity)
    - may coexist with II CN gliomas, meningiomas

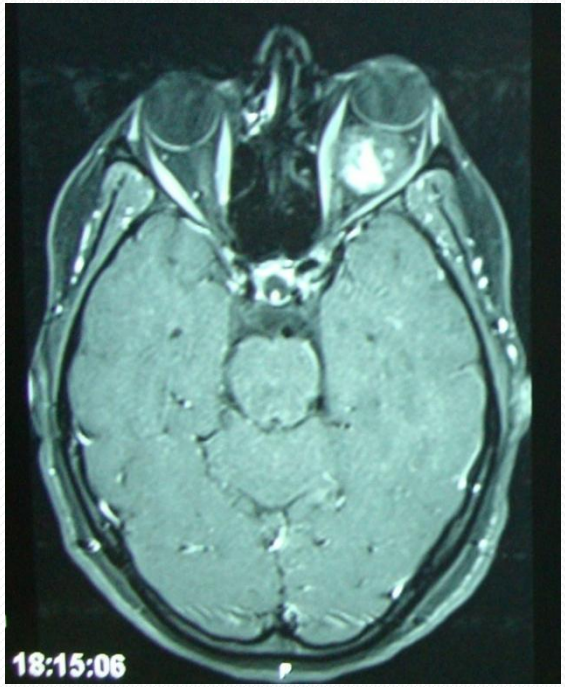
Vascular lesions

- 12-15% orbital tumours
  - Capillary hemangiomas
  - Cavernous hemangiomas
  - Lymphangiomas
  - Hemangiopericytomas
  - AVMs

Capillary hemangiomas

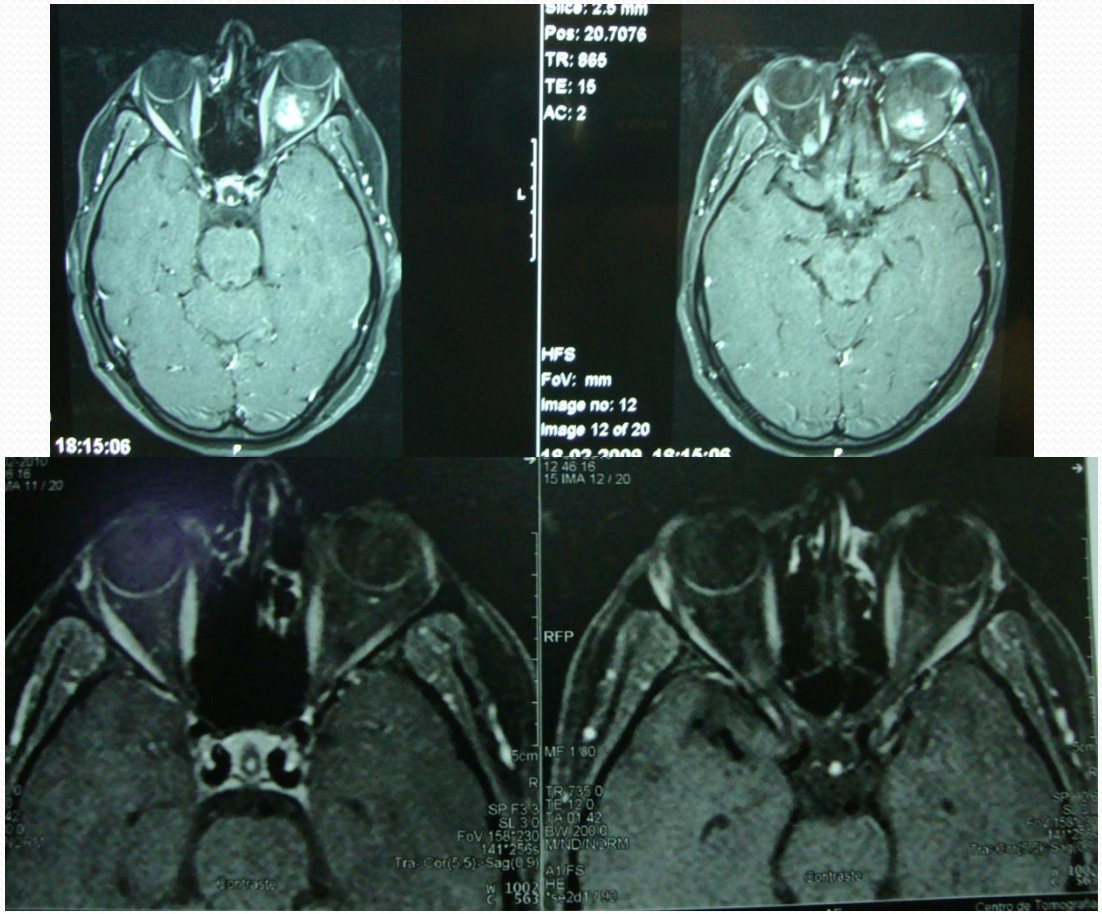
- Benign
- Infiltrative
- Associated cutaneous manifestations (strawberry nevi)
- 6 months
- Spontaneous resolution over 3-5 y (cosmetic deformity, amblyopia)
- Tx: argon laser therapy or steroids (involution)

Cavernous hemangiomas



- Most common benign orbital tumour
- Young/middle-aged adults
- Low-flow, circumscribed, behind the globe, intraconic
- Tx: surgery

Cavernous hemangiomas



Lymphangiomas

- Slow-growing
- Children, young adults
- Slowly progressive exophthalmos
- Haemorrhage
- Difficult to manage surgically (involve critical structures)
- Laser therapy as adjuvant

Hemangiopericytomas

- Malignant
- Arise from undifferentiated mesenchimal cells with pericytic differentiation
- Young/middle-aged adults
- Invasive, mestastasize (rare from orbit)
- Goal: total removal (prevent recurrence)

Tumours of mesenchimal origin

- Rhabdomyosarcoma
  - most common malignant tumour children
  - rapidly progressive
  - good response RT and CTH

- Fibrous histiocytomas
  - most common orbital tumour adults
  - insidious, locally infiltrating
  - benign (high recurrence → resected wide margins)
  - malignant fibrous histiocytomas: mestastasize, death

Tumours arising from bone and cartilaginous structures

- Osteomas
- Ossifying fibromas
- Fibrous dysplasia
  - ↓ visual acuity, cosmetic deformity
- Aneurysmal bone cysts

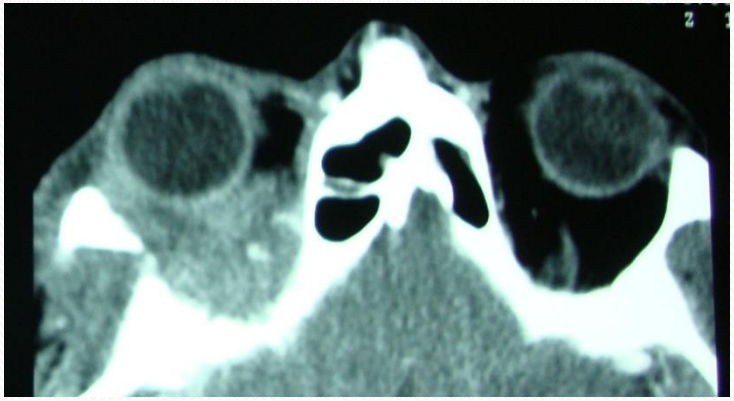
Other tumours

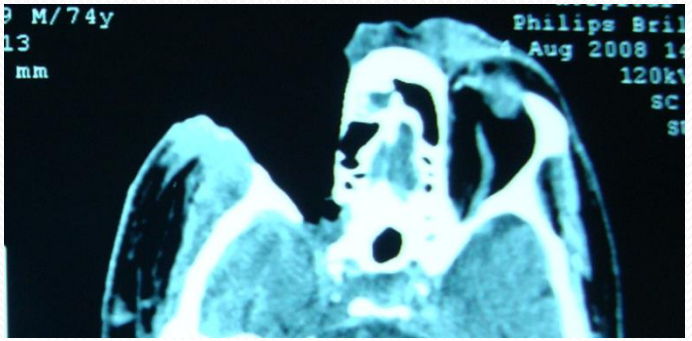
- Dermoid and epidermoid
  - benign cystic lesions
  - dermoid: located anteriorly, childhood
  - frontozygomatic suture (+ frequent)

- Orbital pseudotumour
  - large sepctrum non-specific idiopathic inflammations
  - common cause proptosis 2nd to 7th decade life
  - multifocal involvement
  - dull orbital pain worse with eye movement (++), proptosis (+++)
  - tx: steroids, surgery, RT, immunosuppressive agents

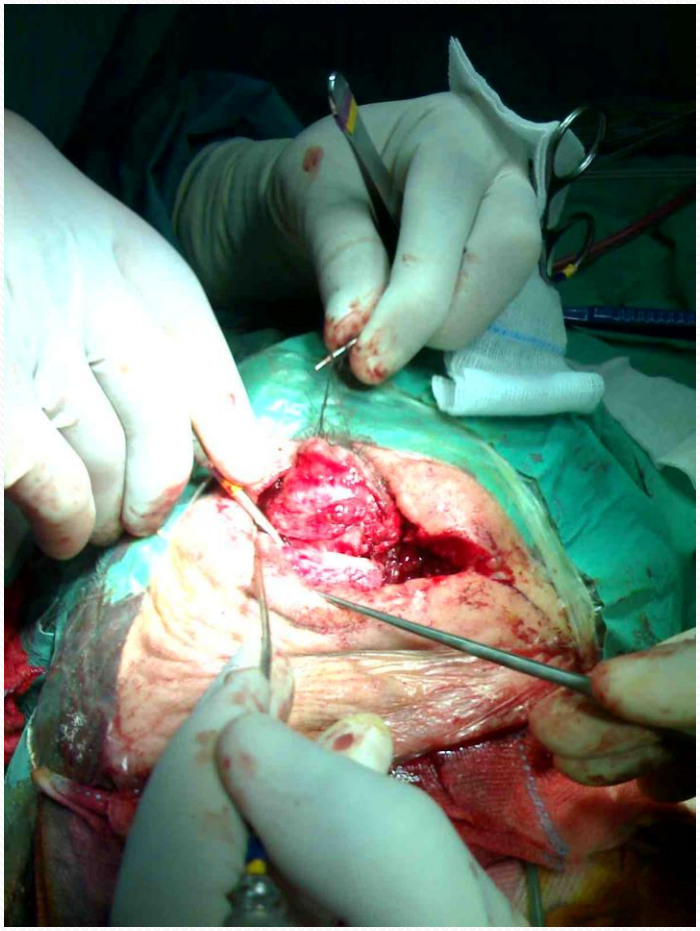
Lacrimal gland tumours

- 10% orbital tumours
- Superolateral, anterior orbit
- ½ malignant
- Radical surgery (skull bone and dura removal)



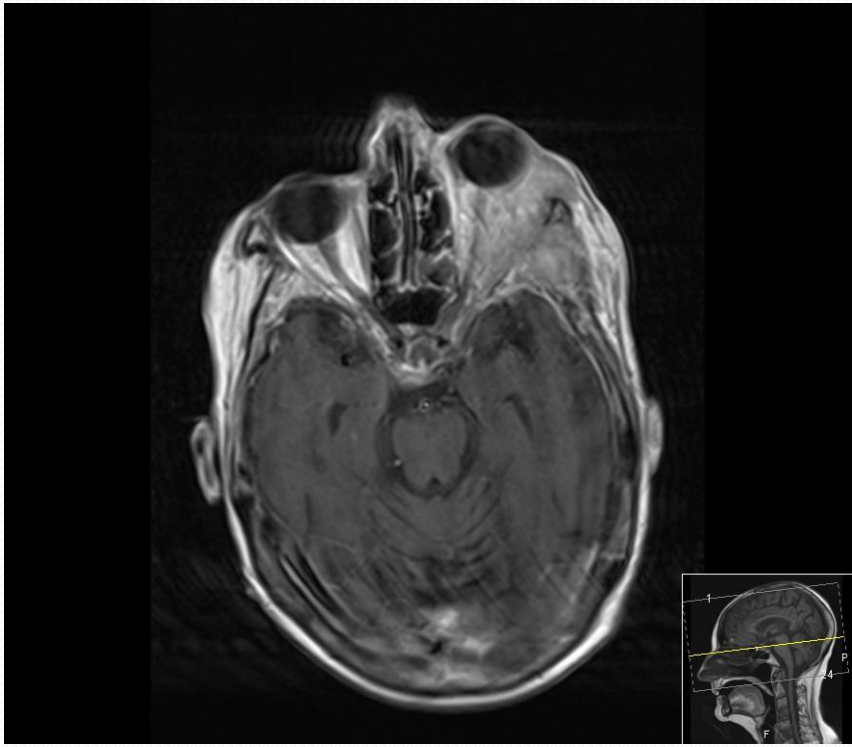


Adenoid cystic carcinoma



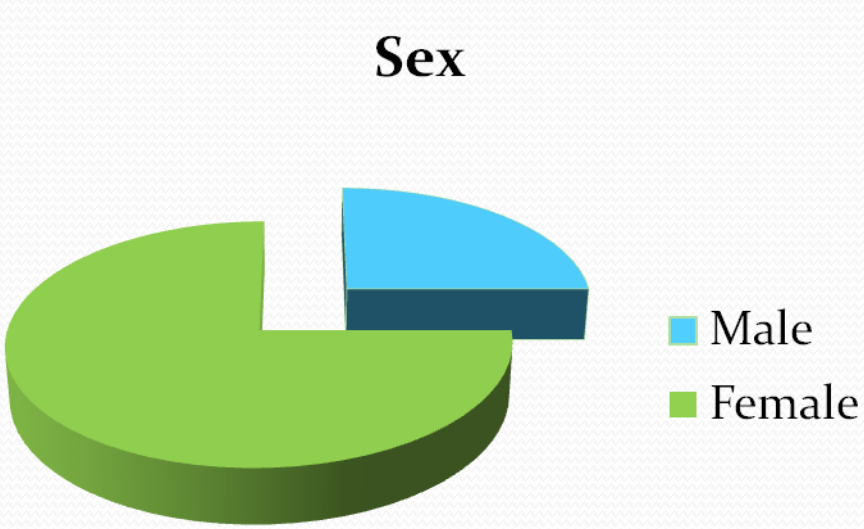
Metastatic lesions

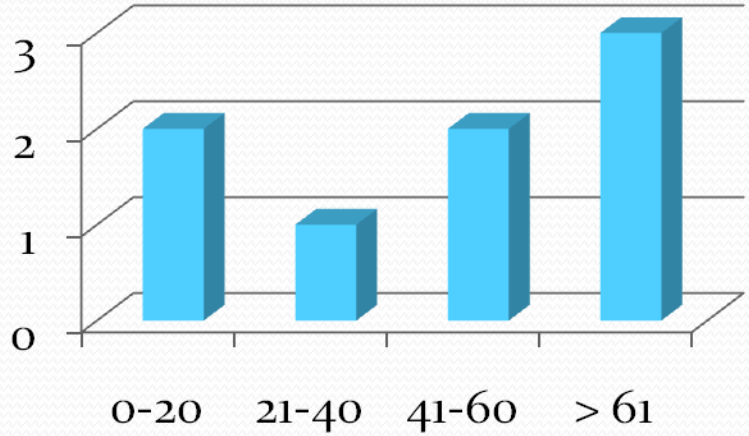
- Children
  - neuroblastoma, Ewing tumour and Langerhans cell histiocytosis
- Adults
  - brest < lung < prostate < melanoma < G-I tract < kidney



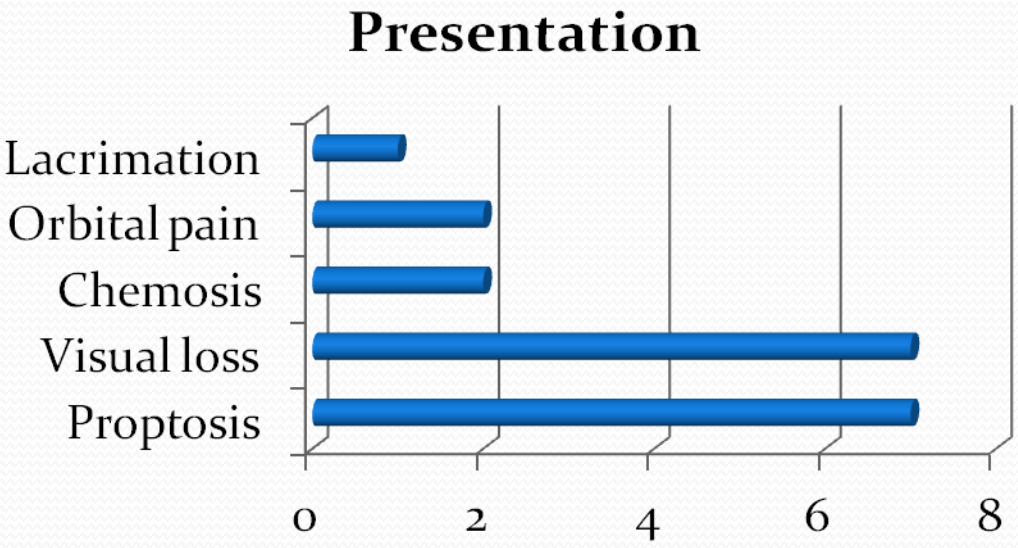
Results

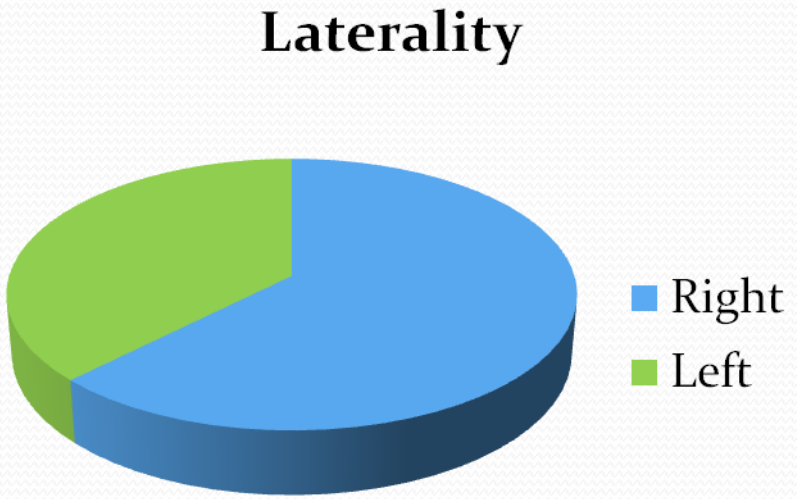
- April 2006 – January 2011 (n=8)
- 5-78 y (46,5)





Results


Presentation
Lacrimation
Orbital pain
Chemosis
Visual loss
Proptosis
0
2
4
6
8


Laterality
Right
Left

Results

Location	
Extraconal	4
Intraconal	3
Intra and extraconal	1

Resection	
Total	3
Parcial	5
Biopsy	1

Approach	
Lateral orbitotomy	4
Lateral supraorbital	2
Pterional (+ exenteration)	2
Fronto-orbital	1

Histology	
Meningioma	3
Pilocytic astrocytoma	1
Ganglioglioma	1
Melanoma metastasis	1
Cavernous hemangioma	1
Adenoid cystic carcinoma	1

Results

Complications	
CSF leak	2
Superior rectus palsy	1

Visual acuity	
Improved	3
Stable	3
Worse	2

- Proptosis improved in all patients
- 2 FSR (meningiomas)
- 1 RT (adenoid cystic carcinoma)
- 2 deaths (melanoma metastasis, adenoid cystic carcinoma)

Hospital Braga

Conclusions

- Varied pathology
- Detailed anatomical knowlegde
- Technically demanding
- Multidisciplinary approach