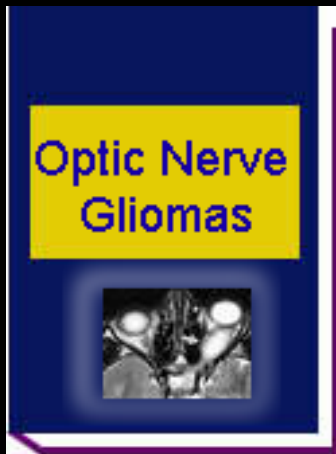




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Optic Nerve Gliomas

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Optic nerve gliomas

- 4% of orbital tumours
- 4% intracranial gliomas
- 66% of primary optic nerve tumours
- Gliomas 4:1 times more common than meningiomas
- 2-8 yrs of age
- 3:2 girl :boy dominance
- If bilateral –diagnostic of NF1

Anatomy of the optic nerve

- Four parts: total length 45-50-mm
- Intraocular 1mm
- Intraorbital 2.5 mm
- Intracanalicular 5mm
- Intracranial 10mm
- Normal diameter 4-6mm
- ON is an outpouching of the brain it has three covering layers of the brain-dura arachnoid and pia
- These coverings enclose the SAS.

Anatomy of the optic nerve

- In the orbit ON surrounded by orbital fat
- Optic canal lies within the lesser wing of the sphenoid
- The ON passes posteriorly and medially through the optic canal surrounded by its three sheaths.
- The dura fuses with the bony periosteum. The SAS is continuous with the intracranial space
- Accompanying the nerve through the canal are the ophthalmic artery and sympathetic nerves
- As the nerve passes backwards it migrates superiorly and medially to reach the optic chiasm.
- At the posterolateral aspects of the chiasm are the optic tracts



Clinical presentation

- 1) Intraocular gliomas
- May be found incidently
- Progressive loss of visual acuity
- 2) Intraorbital gliomas
- Decreased visual acuity
- Proptosis of the globe
- Loss of vision with extreme eye movements is a sign of ON compression
- Afferent pupillary defect

Clinical presentation

- 3) Intracranial gliomas
- Slowly progressive loss of vision
- Optic atrophy
- Chiasmal lesions produce bitemporal hemianopia
- Diminished visual acuity in a pt of NF 1 is a clinical hallmark of ONG.
- Loss of visual acuity in the first decade of life is due to ONG ,whereas slight loss of visual acuity in presence of an enlarged optic disc is more suggestive of a perioptic meningioma.
- Spasmus mutans is a form of nystagmus associated with ONG

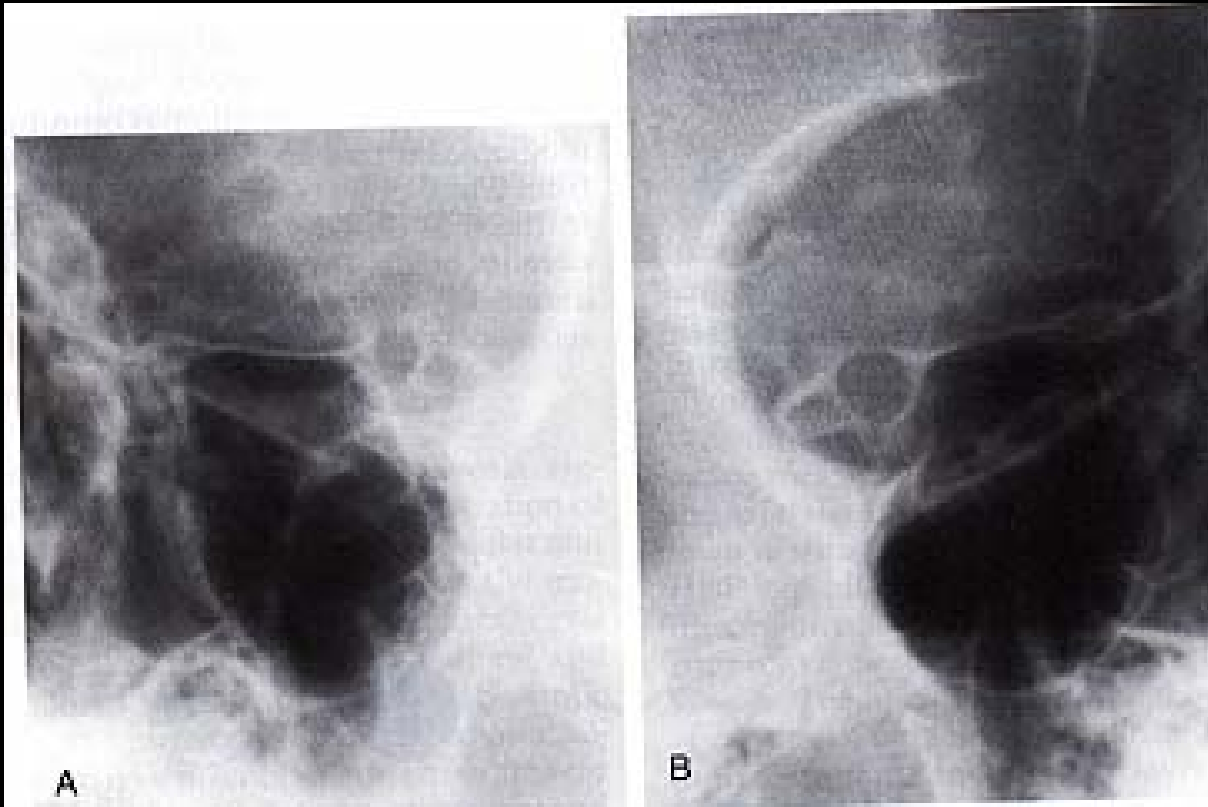
Clinical presentation

- OA and loss of vision occur secondary to destruction of the axons by astrocytes and by obliteration of the nutrient arteries by the tumour
- Visual field defect in intraorbital ONG is a central scotoma
- Lesions of the optic tracts give rise to incongruous homonymous hemianopia
- Lesions of the lateral geniculate body and optic radiations result in homonymous sector defects
- Lesions of the occipital lobes result in varied field defects, homonymous hemianopia homonymous quadrantanopia and splitting and sparing of fixation



Imaging

- Plain X rays
- Enlargement of the optic canal $> 5\text{mm}$
- Angiography
- Not used presently
- Angiographic sign of ONG –widening of the loop of the ophthalmic artery as it passes around the ON
- Meningiomas exhibit tumour vascularity which is not typically present in ONG.



Imaging

- CT scan
- 2-3mm axial and coronal sections with and without contrast
- For evaluation of the optic canal 1.5 mm axial and coronal images though the canal with bone filtering can be used.
- Contrast enhanced CT has limited application in evaluation of the ON.

MR IMAGING

- Preferred imaging modality for ON
- Allows complete evaluation of the ON chiasm, tracts, lat geniculate bodies, occipital radiations and occipital cortex
- Subtle enhancement or enlargement of the ON can be easily identified with MRI.
- Subtle expansion or erosion of the optic canals more reliably seen with CT.
- Optic pathway evaluation-Helmholtz head coil
- Intraocular lesions –surface coils

MR IMAGING

- Complete MR evaluation of the optic pathway—HR axial images from mid maxilla to the mid portion of the frontal sinuses, coronal images from post globe thru cavernous sinus to the post aspect of the brainstem. T1 and T2 weighted images
- Slice thickness 3-5mm
- Interslice gap 1-2mm
- FOV 14-16 cm
- Resolution of 256 x 192 or 256 x 256
- Oblique sagittal images provides view of the entire nerve
- Gadolinium should be used in all ON studies.
- Fat suppression studies should be done post contrast

MR IMAGING

- Fat sat T2 weighted images are also useful.
- Complete evaluation of the optic pathway should include an evaluation of the brain to study the optic radiations and the occipital cortex.
- On both T1 and T2 Weighted images the ON is isointense to WM
- T2WI allows delineation of the ON from the the dural sheath with the hyperintense CSF space in between.
- On T1 weighted images the intracanalicular portion of the ON is outlined medially by the hypointense ethmoid sinus and laterally by the fatty marrow in the ant clinoid process



OPTIC NERVE GLIOMA

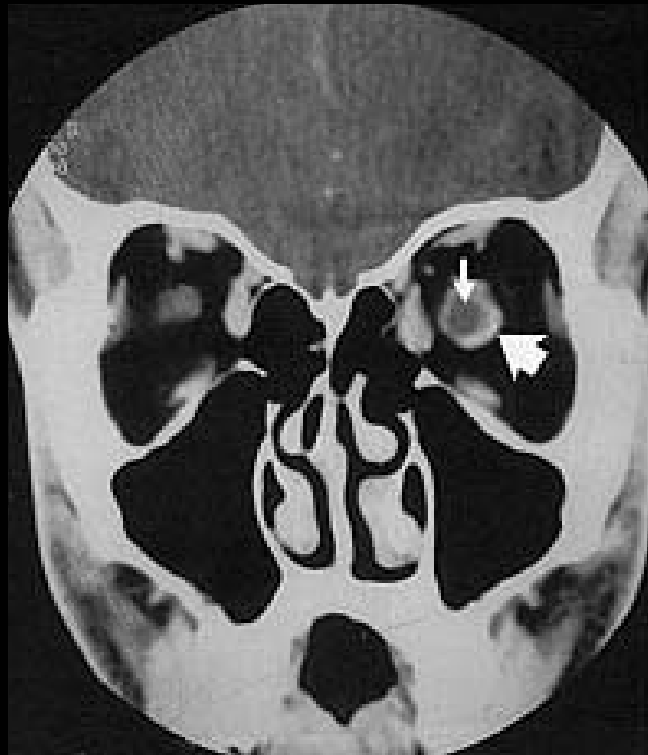
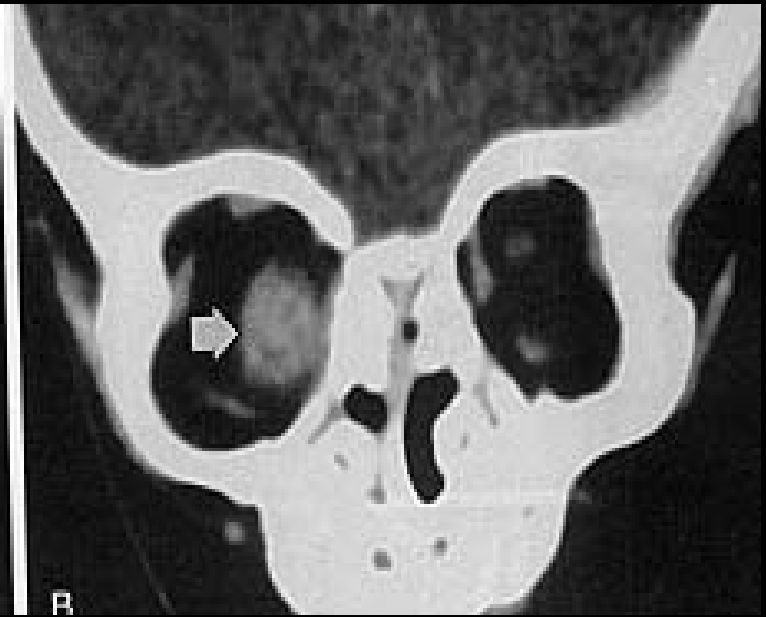
- Clinically and biologically two subsets
- Childhood form
- Adult form
- 1) Childhood form
- Found in young children
- Follows an indolent course
- Spontaneous regression has been reported
- Histologically juvenile pilocytic astrocytoma

OPTIC NERVE GLIOMA

- 2)Adult subgroup
- Middle aged pts
- Exhibit malignant behaviour with hypothalamic and temporal lobe infiltration
- Pathologically anaplastic astrocytoma or glioblastoma
- 50% involve the orbital portion of the nerve with remainder affecting the chiasm and orbit.
- Survival from time of presentation is usually less than 1 yr.

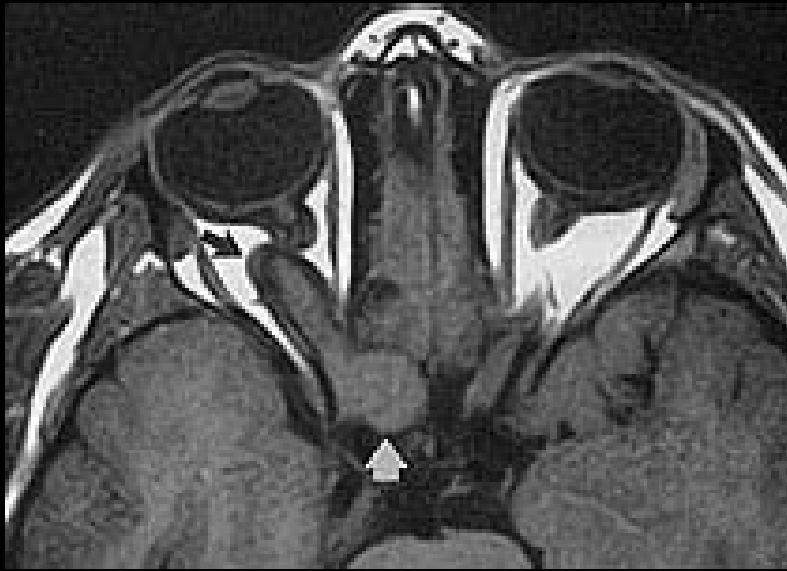
CT APPEARANCE OF ONG

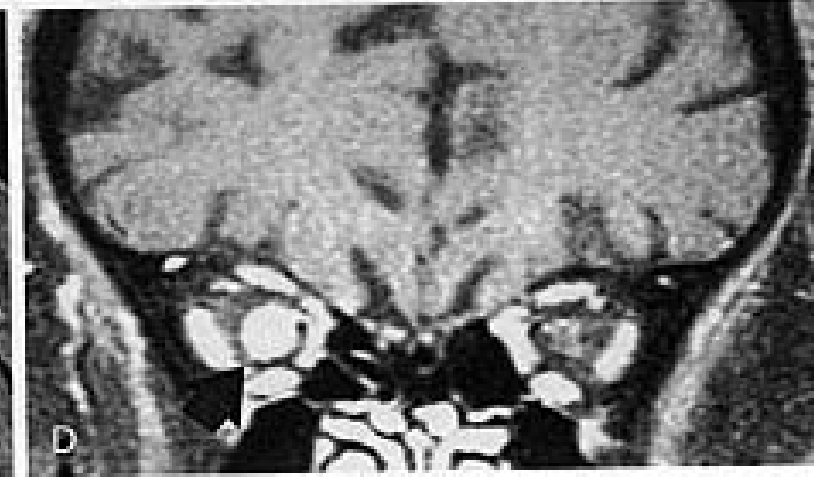
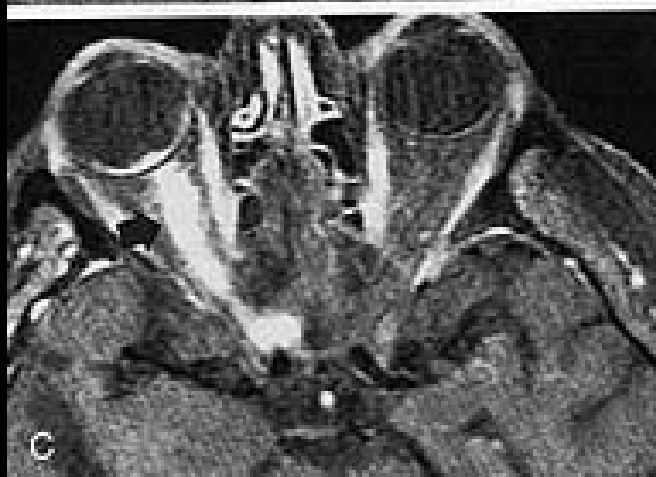
- Non contrast CT
- Enlargement, kinking and buckling of the ON
- Nerve becomes fusiform in shape
- Calcification not seen in previously untreated gliomas as compared to Meningiomas.
- Cystic changes are common
- Contrast enhancement is variable –moderate to intense-less than meningioma
- Optic nerve sheath can appear thickened due to arachnoidal hyperplasia associated with NF1.

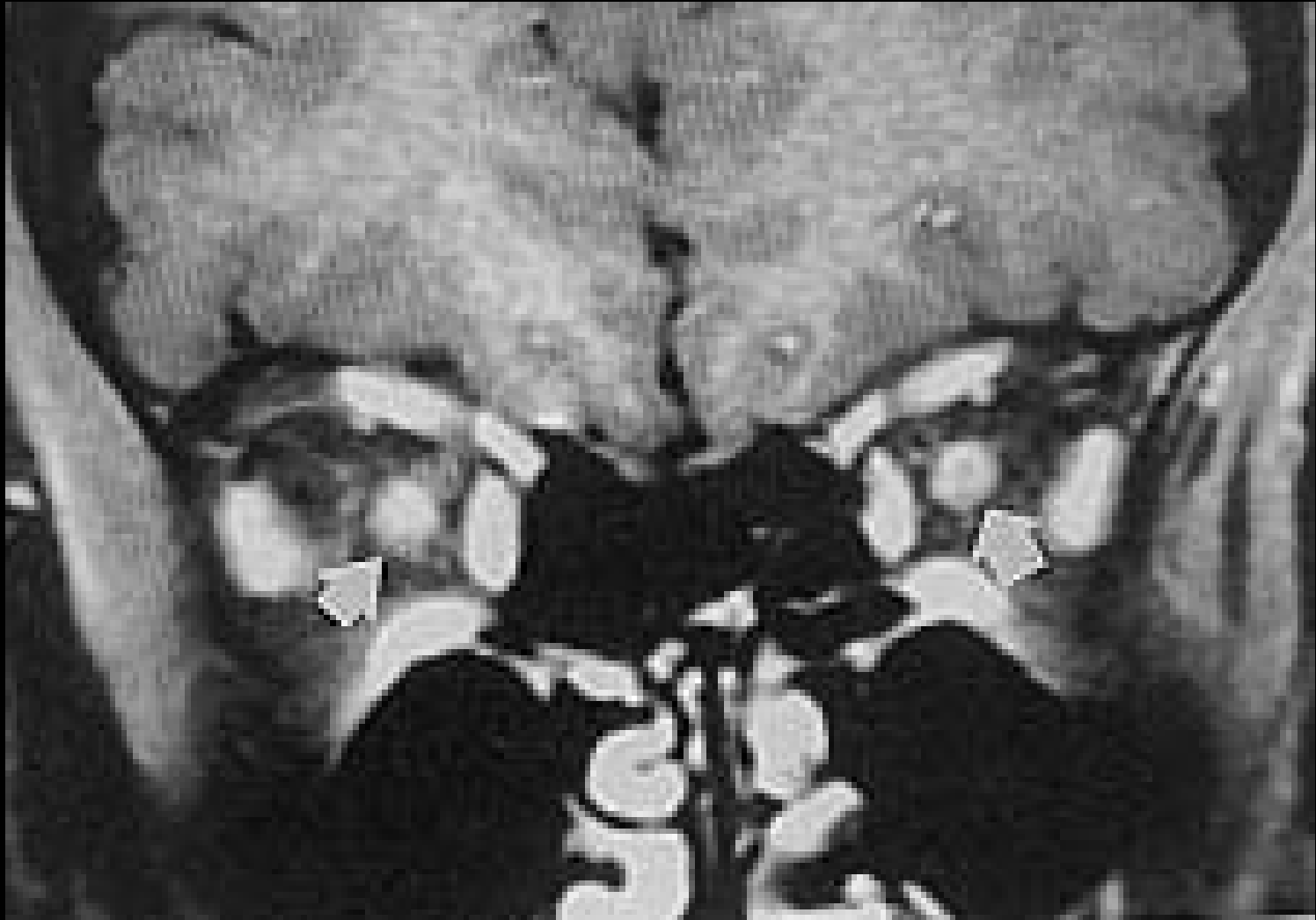


MR IMAGING OF ONG

- Precontrast T1 weighted images---nerve is isointense to cortex and appears enlarged , buckled and kinked .
- T2 weighted images—hyperintense to cerebral cortex, may appear heterogenous secondary to cystic areas
- Gliomas enhance variably and complete lack of enhancement can also occur
- Adult type ONG early imaging findings are non specific







TREATMENT

- Childhood ONGs grow slowly the most common therapeutic option is not to treat and re evaluate at frequent intervals
- radiation and chemotherapy are used to slow the tumour progression during growth spurts
- surgical debulking and enucleation are last resort treatment options.

TREATMENT

- Adult form ONGs grow rapidly ,invade the brain and are uniformly fatal.
- No therapy is effective in prolonging the survival.
- Differential diagnosis
- perioptic sheath meningioma, infectious or granulomatous neuritis, orbital pseudotumour lymphoma etc

MISCELLANEOUS OPTIC NERVE TUMOURS

- Malignant teratoid medulloepithelioma
- Ganglioglioma- optic nerve shows diffuse enlargement unlike that observed in ONG or meningioma.
- Haemangioblastoma
- Metastatic disease- ON involvement usually represents extension from retinochoroidal, intracranial or orbital metastatic localisation
- Lymphoproliferative diseases
- Choristoma



ORBITAL PSEUDOTUMOUR

- Non granulomatous inflammatory process in the orbit or the eye with no known local or systemic causes.
- Diagnosis by exclusion based on history clinical course , response to steroid treatment and biopsy in a limited number of patients.
- Usually occurs in adults but may also affect children.
- Paediatric orbital pseudotumour represents 6-16% of pseudotumours



CLINICAL PRESENTATION

- The disease is characterized according to the orbital structure predominantly involved.
- Myositis- one or more extraocular muscles
- dacryoadenitis-lacrimal gland
- periscleritis including Tenons space
- trochleitis
- perineuritis-outer dural sheath of the ON
- The disease may may be localized to these orbital structures but frequently there is associated fatty infiltration
- The inflammatory process may be localised simulating a tumour or may be diffuse within the fatty tissue

CLINICAL PRESENTATION

- Symptoms are a reflection of the degree inflammatory response and the location of the inflammatory tissue.
- Acute form---abrupt onset of pain , lid swelling and redness associated with diplopia and decreased vision.
- There may be ptosis ,proptosis and decreased orbital resilience with ballotment and pain on globe motion
- on slit lamp examination there is conjunctival chemosis and injection

CLINICAL PRESENTATION

- Chronic sclerosing form
- evolves over weeks to months
- signs of fixation of the globe and mass effect are prominent
- slowly progressive visual loss, diplopia and proptosis.
- Lacrimal gland: acute dacryoadenitis-pain in the superotemporal region. Lacrimal gland is enlarged and tender on palpation
- in the chronic forms -painless lacrimal fossa mass
-tumour cannot be excluded



CLINICAL PRESENTATION

- Extraocular muscles
- diplopia and pain exacerbated by eye movements
- localised conjunctival congestion and chemosis at the tendinous insertion of the involved muscles.
- If pseudotumour is located at the orbital apex-optic neuropathy and ophthalmoplegia

CLINICAL PRESENTATION

- Optic nerve dysfunction results from inflammation of the perineural tissue or compression of the ON from mass effect
- acute form of pseudotumour -responsive to high doses of systemically administered prednisolone
- variant of pseudotumour-Tolosa Hunt syndrome-characterised by inflammatory infiltration of the orbital apex including SOF and cavernous sinus.

TOLOSA HUNT SYNDROME

- Presents as painful ophthalmoplegia, minimal proptosis ,rare visual loss and external signs of orbital inflammation.
- Ophthalmoplegia-3,4 and 6 nerve palsies along with hypoesthesia of the periorbital skin due to involvement of V1.
- Generally unilateral but bilateral cases may occur.

RADIOLOGIC FINDINGS

- Lacrimal gland involvement
 - single
 - combined with other orbital tissue
- muscle involvement
 - single
 - multiple, often associated with orbital fat infiltration
- orbital fat involvement
 - diffuse ill defined infiltrations with or without involvement of the ON, lacrimal gland and muscles.
- Infiltration or mass at orbital apex
- tolosa hunt subtype
- globe involvement
 - tenons space-tenons fasciitis
- scleritis



RADIOLOGIC FINDINGS

- CT scans
- no specific density values
- variable enhancement after contrast
- MRI
- low signal on T1 and T2 weighted images
- depends on the degree of fibrosis with the sclerosing variety apt to reveal lesser degree of signal intensity on T2 weighted images.
- Marked enhancement on post gado scans

RADIOLOGIC FINDINGS

- Lacrimal gland
- most frequently involved orbital structure
- diffuse oblong enlargement of the lacrimal gland with preservation of the shape of the gland
- marked expansion along the AP diameter along the lateral orbital wall and lateral rectus muscle
- inflammatory reaction in the periglandular tissue imparts a poor definition to the margins of the gland



RADIOLOGIC FINDINGS

- No density or intensity characters on CT or MRI to differentiate enlargement from other causes
- prompt response to steroids with radiologic findings supports diagnosis of pseudotumour.

RADIOLOGIC FINDINGS

- Extraocular muscles
- single or combination of muscles
- tendons enlarge along with the muscle bundles and lead to a tubular configuration
- contrast to thyroid ophthalmopathy-spindle shaped configuration with normal muscle tendons
- enlarged inflammed muscles reveal intense enhancement which also occurs in normal muscles and therefore does not have any diagnostic implications



RADIOLOGIC FINDINGS

- The enlarged muscles may regress after steroids.
- There may also be ill defined infiltrates through out the orbital fat with enlargement of the lacrimal gland.
- May manifest as diffuse infiltrations of the orbital fat enveloping the globe and surrounding the ON sheath complex similar to lymphoproliferative diseases.



RADIOLOGIC FINDINGS

- At the orbital apex
- it may assume appearance of a mass or as
- Infiltration of the orbital apex
- May compress ,obliterate or displace the ON
- Subcategory of pseudotumour-sclerosing pseudotumour
- Presents as diffuse increase in density of the orbital fat with obliteration of the ON, muscles and circumferential involvement of the globe.
- Complete fixation of the intraorbital structures with no motion of the globe



RADIOLOGIC FINDINGS

- If the inflammation extends from the orbital apex to the cavernous sinus –Tolosa Hunt syndrome
- Enlargement of the cavernous sinus on the involved side
- Diffuse enhancement on post contrast scans.
- Involvement of the globe is not uncommon in pseudotumour

RADIOLOGIC FINDINGS

- Diffuse enlargement of the sclero-uveal coat
- Inflammatory process is located in the Tenons space a potential space between the Tenons capsule and the sclera
- There is usually enhancement of the sclera following contrast.

DIFFERENTIAL DIAGNOSIS

- Bacterial infection-orbital cellulitis
- Commonly caused by sinusitis
- Lid swelling which may be limited to the preseptal or may extend into the post septal space
- Fungal infection
- Sarcoidosis
- Sjogrens syndrome
- Wegeners granulomatosis

LYMPHOPROLIFERATIVE DISEASES OF THE ORBIT

- LYMPHOMAS
- NHL is the common in the orbit
- Extranodal involvement in NHL IS common but incidence of orbital involvement is quite low.
- 70-75% of orbital lymphoma pts have systemic disease
- Controversy whether malignant lymphomas originate in the orbit and metastatize to other sites or they are part of systemic process from the beginning.

LYMPHOMAS

- In the orbit incidence of systemic lymphoma is high with involvement of the eyelids and less common occurrence in conjunctiva, orbit and lacrimal gland.
- Pts with bilateral orbital or lacrimal gland lymphoma do not have an increased incidence of systemic lymphoma compared to those with unilateral disease.
- 85% of NHLs are B cell
- 15% of NHLs are T cell

CLINICAL PRESENTATION

- 50-70 yrs
- Females > males
- Painless proptosis
- Ptosis ,diplopia motility disorders and impairment of vision.
- Lymphoid lesions of conjunctiva manifest as salmon coloured elevations
- Orbital lesions frequently involve the superior and anterior orbits causing proptosis and downward displacement of the globe.

CLINICAL PRESENTATION

- Extraocular muscles are rarely involved but are often obliterated from adjacent lymphoid tumours especially levator -superior rectus complex.
- A biopsy is indicated on suspicion
- Total excision is contraindicated as many of these lesions are diffuse and infiltrating.
- Conjunctival epibulbar and lacrimal gland lesions are easily accessible
- Orbital lesions located anteriorly and superiorly can be approached thru a subperiosteal brow incision
- For more posterior lesions a lateral orbitotomy with bone removal may be necessary(Kronlein procedure)

CLINICAL PRESENTATION

- Fine needle biopsy with specific surface marker studies may avoid incisional surgery.
- In lacrimal gland lesions epithelial tumour needs to be differentiated from lymphoma
- In such cases a FNAC or incisional biopsy is contraindicated because of possibility of spillage of tumour cells into surrounding tissue and a complete gland removal is indicated.
- In bilateral cases only one side needs to be biopsied.

PATHOLOGY

- Benign reactive hyperplasia is characterised by benign appearing lymphoid follicles with reactive germinal centres surrounded by lymphocytes histiocytes and plasma cells
- There is a preponderance of T cell lymphocytes and polyclonal B cells.

IMAGING

- Common feature of lymphoid tumours is their tendency to mould or plaster themselves along the globe where such contact exists.
- Fairly well defined round to oval in shape elongated along the extraconal space
- In some cases the margins are ill defined secondary to infiltrations
- In extraconal lesions the medial and lateral rectii are often obliterated and this is due to intimate association of the tumour with the muscle margin though in some instances there is lymphoid infiltration into the muscle



IMAGING

- Intraconal tumour follows contour of the posterior aspect of the globe with no indentation of the globe.
- Infiltrations of the Tenons space causes diffuse thickening of the sclero uveal coat
- There may be small polypoid projections from Tenons space into adjacent orbital fat.
- Perioptic lymphoma causes diffuse enlargement of the optic nerve sheath complex.
- Tumours arising from the conjunctiva seen as a homogenous mass conforming to the anterior globe margins
- Involvement of the eyelids can manifest as anterior masses adjacent to the globe



IMAGING

- Less frequently lymphoma can present as multiple ill defined infiltrations or as diffuse homogenous mass causing complete obliteration of the intraorbital spaces.
- Lacrimal gland involvement characterised by diffuse enlargement of the gland which is elongated in shape on axial views and conforms to the contour of the adjacent globe.
- Coronal CT shows superoinferior elongation of the lacrimal gland moulding itself along the lateral orbital wall and adjacent globe
- Orbital lymphomas can extend thru IOF into the pterygopalatine fossa or thru the SOF into the cavernous sinus area
- In most cases there is no associated bony destruction

IMAGING

- CT MRI USG are studies of choice
- CT study-3-5 mm axial and coronal sections with soft tissue and bone windows
- MRI T1 and T2 weighted images 3-5 mm sections in axial plane and T1 axial and coronal images before and after contrast preferably fat suppressed.

IMAGING

- Anterior preseptal -post septal process, usually involving the superior aspect of the orbit
- infiltration extends to eyelids and underneath the conjunctiva with formation of a mass. Additional extension of infiltration to soft tissues overlying the frontal and maxillary bone may be observed

IMAGING

- Lacrimal gland disease that may be limited to the orbital part of the gland or extend to the palpebral portion.
- The lacrimal gland becomes enlarged with smooth erosion of the adjacent lateral orbital wall
- main differential is pleomorphic adenoma

IMAGING

- Retrobulbar involvement
- tumour infiltrates and replaces the orbital fat
- involvement may be diffuse and poorly defined or as well circumscribed masses
- tendency to mould around existing orbital structures thus no indentation of the globe.
- Infiltration of the perioptic space produces diffuse enlargement of the ON sheath complex.

IMAGING

- Extension of a lymphomatous lesion
- from the sinonasal cavities
- there are no CT or MRI features that allow differentiation of pseudolymphoma from malignant lymphoma
- preferred method of evaluation of lymphoproliferative disease is a CT scan
- T1 weighted images-low signal
- T2 signal varies according to the cellular composition of the lymphoma
- moderate to marked enhancement on the post contrast scans



LACRIMAL GLAND TUMOURS

- Lacrimal gland is about the size & shape of an almond.
- Two lobes-Palpebral and orbital separated by levator aponeurosis
- lacrimal gland tumours classified as epithelial or non epithelial
- epithelial tumours arise from the lacrimal gland acini
- non epithelial lesions consist of congenital , inflammatory and neoplastic conditions



LACRIMAL GLAND TUMOURS

- Pts with lacrimal fossa mass divided in to two grps
- pts with suspected pleomorphic adenoma
- pts with other other lacrimal gland carcinomas, lymphomas, pseudotumours and inflammatory lesions.

EPITHELIAL TUMOURS

- Classification of lacrimal tumours similar other major salivary glands
- epithelial tumours comprise 40-50% of all lacrimal masses.
- Half of these tumours are benign mixed tumors and half are malignant.

EPITHELIAL TUMOURS

- Benign mixed tumour of pleomorphic adenoma of the lacrimal gland
- most common tumour of the lacrimal gland
- term mixed tumour relates to histologic findings of both epithelial and mesenchymal components of these tumours.
- 4th- 5th decade of life
- no sex or racial predilection
- presenting signs include slow growing mass in the lacrimal region
- masses that extend posteriorly may present with proptosis and limited extraocular motility



EPITHELIAL TUMOURS

- Pain is typically absent
- prs of pain, diplopia or rapid growth almost always indicates malignant transformation.
- Involves the orbital portion of the gland
- although benign these tumours have potential to undergo malignant transformation and if not excised en bloc have a high rate of local recurrence.
- Treatment is mainly surgical -complete excision of the tumour

EPITHELIAL TUMOURS

- Pathology
- small tumours-encapsulated with smooth margins
- larger tumours may show an irregular surface
- on microscopy tumour composed of epithelial and mesenchymal components
- epithelial cells are arranged in sheets or cords
- the mesenchymal component can be varied-myxoid, chondroid or mucinous. Bone formation related to bony metaplasia has also been described.



ADENOCYSTIC CARCINOMA

- Uncommon tumour
- second most commonly occurring epithelial tumour affecting the lacrimal gland.
- Most frequent malignant epithelial neoplasm of the gland.
- Occur in a slightly younger age group than mixed tumours with a peak around the fourth decade.
- Pts present with firm to hard mass in the lacrimal gland region
- tumour shows rapid infiltrative growth with predilection for perineural and vascular invasion

ADENOCYSTIC CARCINOMA

- The aggressive infiltrative nature of the disease may lead to limitation in extraocular motility and involvement of the surrounding bone
- complete surgical excision is often impossible.

PATHOLOGY

- Not encapsulated and frequently invade surrounding tissues and bone.
- Histology- benign appearing sheets of cells that surround spaces of varying shape and size--cribriform or Swiss cheese pattern of the tumour
- mesenchymal matrix seen in mixed tumours is absent.
- Classified as different histological types-- tubular cribriform sclerosing comedocarcinoma and basilooid



IMAGING

- Benign mixed tumours -round to oval rather than almond shaped
- well circumscribed masses
- nodularity or infiltrative nature should raise the question of carcinoma rather than pleomorphic adenoma.
- Calcification more common in carcinomas
- bone erosion strongly favours a malignancy
- pleomorphic adenomas are hypo on T1 & hyper on T2 weighted images. Moderate to marked contrast enhancement . Heterogenous appearance may be noted



IMAGING

- Malignant lacrimal gland tumours have an overall rounded configuration and frequently distort the globe and orbital contents
- irregular infiltrative edges and bony erosion may be noted.
- Calcification more commonly seen in malignant tumours
- both benign and malignant tumours can produce bone sclerosis.



NON EPITHELIAL TUMOURS

- Are inflammatory and lymphoproliferative in nature
- acute and chronic dacroadenitis, pseudotumour, sarcoidosis and the lymphoepithelial lesion that may occur either in Sjogrens syndrome or as a localised lacrimal gland lesion--Mikulicz syndrome.
- Lymphoid lesions both benign and malignant occur commonly in the lacrimal gland



CLINICAL PRESENTATION

- Slowly progressive, painless loss of vision and proptosis
- Age at presentation varies from 3-76 yrs
- Presence of optociliary venous shunt of the disc, accompanied by disc pallor or visual loss is highly suggestive of optic nerve sheath meningioma

RADIOLOGIC DIAGNOSIS

- Arise from meningothelial cells of the arachnoid situated along the ON sheath or from extension of an intracranial meningioma into the orbit.
- Tumours may also arise from the ectopic arachnoid cells in the orbit
- Plain Xrays- enlargement of the optic canal and hyperostotic changes of the optic canal.

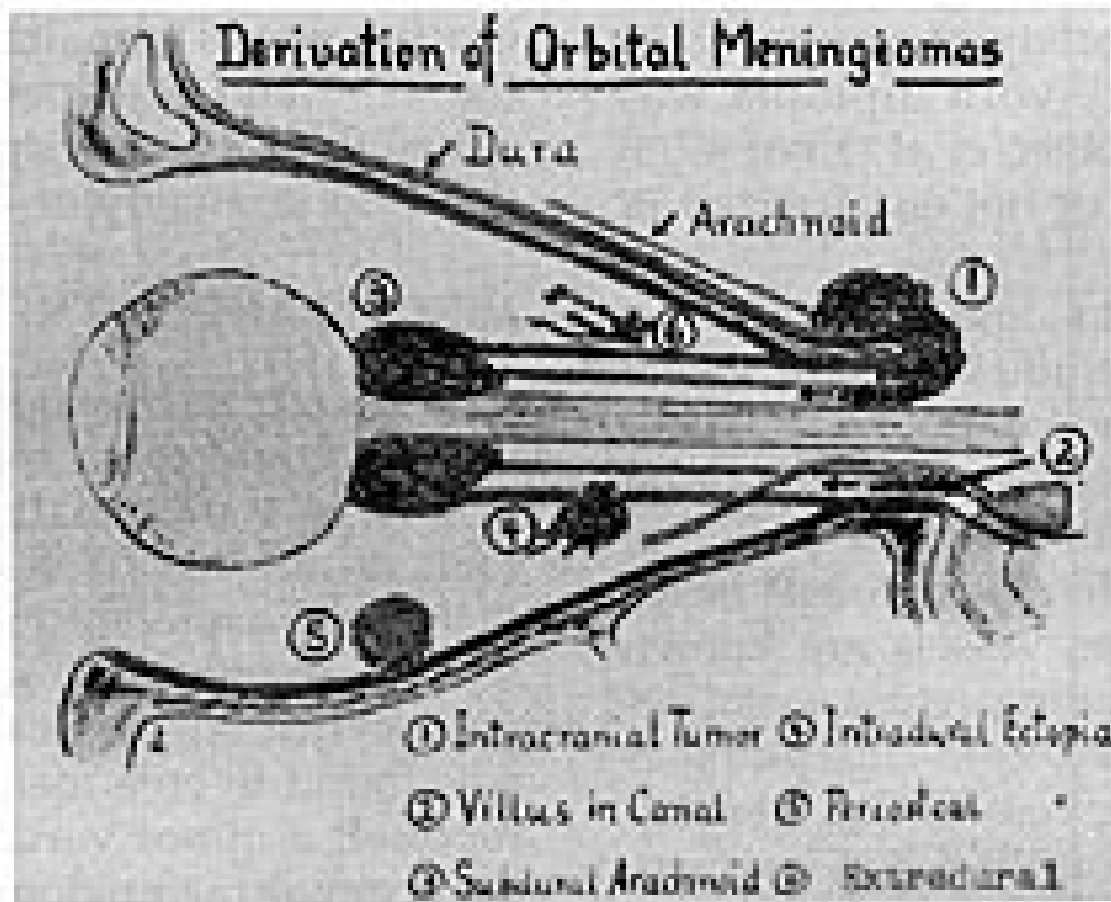


Figure 3. The various locations of meningiomas in the orbit. (From Walsh FB: Meningiomas primary within the orbit and optic canal. Neuro-ophthalmology: Symposium of the University of Miami and the Bascom Palmer Eye Institute. Saint Louis, C.V. Mosby, 1975, pp 166-190; with permission.)

RADIOLOGIC DIAGNOSIS

- Imaging diagnosis based on CT and MRI
- CT is an excellent imaging study for evaluating ONSM
- CT obtained before and after contrast enhancement. Thin sections 1.5-3mm required to visualize exact extent of the tumour.
- ONSM confined to the dura thus seen as well defined tubular thickening of the ON
- ONSM commonly seen as diffuse tubular enlargement or eccentric expansion of the of the ON.



RADIOLOGIC DIAGNOSIS

- ONSM show homogenous and well defined contrast enhancement
- can be infiltrative and show irregular and serrated appearance.
- Tram track sign originally described for ONSM -enhanced CT scans show lucency (ON) in the centre of an enlarged and enhancing ONSM.
- DDs-optic nerve lymphoma, optic neuritis and pseudotumours.



RADIOLOGIC DIAGNOSIS

- CT scans may show linear, plaque like or granular calcifications within or along the ON mass.
- ONSM surround the ON & thus the caliber of the ON is attenuated within the surrounding tumour.cf ONG where the nerve itself is expanded.
- Pneumosinus dilatans may be noted.



Optic Nerve Sheath Meningioma

RADIOLOGIC DIAGNOSIS

- MR scans less sensitive than CT for detection of calcification.
- Is the imaging modality of choice for ONSM.
- ONSM seen as localized or fusiform dilatation of the ON
- tumour retains isointense appearance to ON and brain tissue on most MR pulse sequences.
- Post contrast images reveal moderate to marked contrast enhancement
- enhanced fat suppressed are most valuable for definition of ONSM
- en plaque meningiomas can only be diagnosed on Fat sat post contrast images.



RADIOLOGIC DIAGNOSIS

- Childhood ONSM are often associated with NF2 and is more aggressive than the adult form.
- Another rare grp of intraorbital meningiomas consists of tumours arising from ectopic arachnoid cells in the orbital cavity.
- Frequently associated with localised expansion of the adjacent ethmoid air cells the so called blistering.



DIFFERENTIAL DIAGNOSIS

- Sarcoidosis
- optic neuritis (demyelinating type)
- optic nerve gliomas

PERIPHERAL NERVE TUMOURS OF THE ORBIT

- Orbit is a host to many peripheral nerves
- V1, 3RD 4TH and 6th CNs
- 4% of orbital tumours
- malignant peripheral nerve sheath tumours in the orbit are extremely rare.
- Sensory nerves give rise to PNT with greater frequency as compared to motor nerves.
- VA not affected unless tumour compresses the ON.



NEUROFIBROMA

- Plexiform
- diffuse
- localised or circumscribed
- post amputation neuromas

PLEXIFORM NEUROFIBROMAS

- Present in infancy and childhood
- most commonly involve the eyelids
- presenting sign is visible or palpable mass in the eyelid with subsequent ptosis.
- The tumour can extend to the forehead , temple and superior orbit leading to downward displacement of the globe.
- Plexiform neurofibroma of the lid is virtually diagnostic of Von Recklinghausens disease



NEUROFIBROMAS

- Diffuse neurofibromas have an appearance similar to plexiform neurofibromas.
- Localised neurofibromas present as slow growing tumours that exert mass effect with displacement of the globe in the direction opposite that of the tumour.
- Tumour more common in the superior quadrants.
- Commonly seen in the 3rd -5th decade.



PATHOLOGY

- Plexiform neurofibromas are unencapsulated with proliferating units surrounded by perineurium enclosing axons, Schwann cells and endoneurial fibroblasts.
- There is marked increase in vascularity which leads to profuse bleeding during surgery.
- Increased vascularity is reflected by marked contrast enhancement on CT and MR images.

PATHOLOGY

- Solitary neurofibromas often demonstrate a pseudocapsule, but a true perineurium is not seen.
- Composed of wavy bundles of peripheral nerve sheath cells with comma shaped nuclei and hyaluronic acid and collagen in the stroma.

SCHWANNOMAS

- Clinically schwannomas and neurofibromas are indistinguishable since both arise from sensory nerves and demonstrate slow progressive growth with ocular displacement.
- Schwannomas are encapsulated cf neurofibromas.
- Commonly have a oval or fusiform shape.
- Schwannomas and neurofibromas are differentiated histologically.

PATHOLOGY

- Encapsulated by the perineurium of the nerve of origin.
- Classic feature is the alternation within the lesion of solid cellular areas termed as Antoni A pattern and loose myxoid tissue with stellate or ovoid nuclei termed as Antoni B pattern. Nuclear pallasading is more common than in neurofibromas
- highly organised picket fence appearance of the nuclei referred to as Verocay bodies.



IMAGING

- PLAIN FILMS
- enlargement of the bony orbit
- CT demonstrates a well circumscribed homogenous tumour with density similar to brain.
- Heterogenous center may be seen in tumours that have undergone central necrosis or lesions with varying internal cellularity such as Schwannomas.
- Both tumours enhance with contrast.

IMAGING

- Plexiform neurofibromas and diffuse neurofibromas are well enhancing lesions with irregular borders and an infiltrative appearance.
- These lesions can extend to any orbital structure
- in case of plexiform neurofibromas the tumour involves the eyelid and extends to surrounding areas such as forehead and temporal fossa.
- When evaluating a pt of plexiform neurofibroma one should also look for other findings consistent with neurofibromatosis such as sphenoid wing dysplasia and orbital roof defects

IMAGING

- MR imaging
- solitary neurofibromas/schwannomas: well circumscribed oval or fusiform mass that is hypointense to orbital fat and isointense to extraocular muscles and brain on T1 weighted images.
- The tumours are hyperintense to orbital fat on T2 weighted images.
- The myxoid regions of the tumour with greater water content show greater signal intensity on T2 weighted images as compared to the more cellular components.

IMAGING

- Both tumours show heterogenous enhancement on the post contrast scans with the mucoid matrix and cystic components demonstrating greater enhancement.
- Orbital Plexiform neurofibromas and diffuse neurofibromas appear as ill defined masses with extension into the surrounding tissues .
- Heterogenously hypointense on T1 weighted images relative to orbital fat. It shows high signal intensity on T2 weighted images relative to fat .
- There is variable enhancement better seen with fat suppression techniques.



TREATMENT

- Both tumours are readily amenable to surgical excision.
- surgical excision is relatively blood less and most tumours can be removed intact.
- The primary reason to distinguish between neurofibroma and schwannoma is that neurofibromas are more likely to undergo malignant transformation.
- Treatment of Plexiform neurofibromas is less satisfying. These lesions are quite vascular and diffusely infiltrative .
- Complete removal without total exentration is unacheivable.





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