A. Imaging of the orbit: the globe and conal lesions

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Learning objectives

CT and MRI play crucial roles in the evaluation of orbital pathology. Familiarity with the radiologic appearance of common orbital lesions is important as many of these lesions will not be seen on physical examination. In this poster, we discuss some of the common non-traumatic orbital and ocular masses.
Background:

Familiarity with orbital and ocular anatomy is crucial to the understanding of disease processes of the orbit. Lesions of the orbit may be divided into those which are intraocular and those which are extraocular. The globe contains three layers:

-Sclera: The outer layer is composed of tough collagen-elastic tissue. It is continuous anteriorly with the cornea.

-Uvea: The middle layer is composed of the choroid, the ciliary body, and the iris.

-Retina: The inner layer is continuous with the optic nerve.

The extraocular compartment may be subdivided into three spaces:

-Pre-septal space: The space anterior to the orbital septum is the pre-septal space. The septum is composed of a fibrous sheet that is continuous with the periosteum of the orbit. It provides a strong barrier to the spread of infection.

-Intraconal space: The superior, inferior, lateral, and medial rectus muscles are connected by a fibrous membrane which provides the demarcation between the intraconal space and the extraconal space. These muscles originate from the annulus of Zinn near the orbital apex.

--Extraconal space: Defined as the space outside of the rectus muscles and their connecting sheath.

Nerves of the orbit:

-Optic nerve (CN 2)

-Oculomotor nerve (CN 3)

-Trochlear nerve (CN 4)

-Abducens nerve (CN 6)
Vasculature of the orbit:

-Ophthalmic artery: provides the main blood supply to the orbit. It branches from the internal carotid artery just distal to the cavernous sinus.

-Superior and inferior ophthalmic veins: valveless veins which either unite prior to or drain directly into the cavernous sinus.

The bony structure of the orbit is comprised of seven bones: the frontal bone, the sphenoid, the zygoma, the maxilla, the lacrimal, the palatine, and the ethmoid. The orbital floor is composed primarily of the maxilla. The lateral wall is composed of primarily the zygoma and the sphenoid. The medial wall is composed of the lacrimal bone, the sphenoid and the ethmoid (lamina papyracea). The orbital roof is composed primarily of the frontal bone.

Orbital foramen:

-Orbital canal: optic nerve, sympathetic fibers, ophthalmic artery

-Superior orbital fissure: occulomotor nerve, trochlear nerve, the ophthalmic branch of the trigeminal nerve, the abducens nerve, the superior ophthalmic vein, sympathetic fibers, the orbital branch of the middle meningeal artery

-Inferior orbital fissure: the maxillary division of the trigeminal nerve, the infraorbital artery and vein, the inferior ophthalmic vein

**Retinoblastoma:**

-The most common intraocular malignancy in the pediatric population.

-Leukocoria is the most common presenting sign of the disease. Generally, leukocoria is a nonspecific finding. It occurs when light is not reflected properly off of the retina. Any process or mass restricting the ability of light to travel through the vitreous and reflect normally off of the retina may result in leukocoria. Retinoblastoma is the most common cause of leukocoria.

-Other signs/symptoms include reduced vision, eye pain, and strabismus.

*Imaging findings:*
-CT is often the primary modality to assess patients with suspected retinoblastoma as it is exquisitely sensitive to intraocular calcifications.

-CT findings typically consist of a hyperattenuating mass in the posterior globe with extension into the vitreus or subretinal space.

-Contrast enhancement may or may not be present.

-Calcifications are present in 95% of cases of retinoblastoma and are highly suggestive of the diagnosis. They are helpful when trying to discriminate retinoblastoma from other pediatric ocular lesions, most commonly Coats disease. Coats disease is a vascular anomaly of the retina in which telangiectasia produces a lipoproteinaceous exudate in the subretinal space. This creates an appearance that is frequently difficult to distinguish from retinoblastoma. Calcification is rare in this disorder.

![Fig. 1: Pre contrast soft tissue and bone windows and coronal post contrast CT images of a pediatric patient with retinoblastoma. Note the high attenuation mass in the posterior vitreous of the right eye with prominent calcifications (arrows). Mild enhancement is seen on the post contrast image.](image)

References: Radiology, Lund University - Lund/SE

-MRI is a complimentary study to CT in the evaluation of retinoblastoma as it is more sensitive in assessing the extent of the disease.

-Close attention should be paid to both globes as retinoblastoma may be bilateral greater than 25% of the time.

-On MRI retinoblastoma will typically be slightly hyperintense to the vitreous on T1-weighted imaging and markedly hypointense to the vitreous on T2-weighted imaging. Post gadolinium enhancement will typically be seen.

-Patterns of spread: Retinoblastoma may spread by direct invasion into the vitreous, the orbit, or the optic nerve. Closely scrutinize the pineal and suprasellar areas for trilateral and quadrilateral retinoblastomas. Once involving the optic nerve, tumor cells may spread...
via the subarachnoid space to involve the intracranial compartment and even the spinal canal. Hematogenous metastases commonly involve bone, brain, and lungs.

- Coronal images are helpful in detecting enlargement/enhancement of the optic nerve suggestive of disease involvement.

**Fig. 2:** Axial T1 pre, T1 FS post, and T2 weighted images as well as coronal T1 FS post and T2 weighted images of a patient with bilateral retinoblastomas demonstrate a large enhancing T1 hyper, T2 hypointense right ocular mass with direct extension into the vitreous (thin arrows). Also note the small left retinoblastoma (thick arrows).

**References:** Images courtesy of Dr. Andrea Rossi, M.D., Dept. of Neuroradiology, G. Gaslini Children's Hospital, Italy.

**Rhabdomyosarcoma:**

- The most common malignant extraocular orbital tumor in the pediatric population. Mean age of presentation is 7 years.

- Thought to develop from pleuripotent mesenchymal cells.

- Prevalence is roughly 1/10 that of retinoblastoma.

- Head and neck rhabdomyosarcomas constitute roughly 1/3 of all pediatric rhabdomyosarcomas and approximately 25-35% of those occurring in the head and neck are orbital primaries.
Tumors are always unilateral.

Patients typically present with rapidly progressive unilateral proptosis, eyelid/conjunctival edema, and ophthalmoplegia.

-3 histologic variants: Embryonal, alveolar, and pleomorphic types. Embryonal is the most common.

**Imaging Findings:**

- The tumor most commonly arises in the extraconal space.

- The common CT appearance of the lesion is an irregular ovoid, well circumscribed extraconal mass.

- Marked enhancement is typical.

- Larger tumors tend to be less well defined, may have necrosis and hemorrhage, and are more heterogeneous in appearance.

- Extraocular muscle encasement is more typical than muscle invasion/enlargement.

- The globe is frequently displaced/distorted but is rarely invaded.

- On MRI, tumors are typically isointense to muscle and brain on T1 weighted images. On T2 weighted images, tumors are typically hyperintense to muscle or brain. Restricted diffusion may also be seen.

- CT is preferred for evaluating bone involvement which is present in roughly 50% of patients at initial diagnosis.

- MRI is preferred for evaluating intracranial extension.

- Hematogenous metastases most commonly involve the lung or bone.

- Local lymph node metastases are rare.

- Important differential diagnostic considerations include orbital cellulitis with abscess, subperiosteal hemorrhage from trauma, and leukemia/lymphoma. Thus, correlation of imaging findings with complete blood count and history is important. Vascular malformations, Langerhans cell histiocytosis, and neuroblastoma metastases should also be considered.
Fig. 3: Pre and post coronal T1 weighted, coronal T2, axial T1 post, and diffusion weighted images of a patient with primary orbital rhabdomyosarcoma. Notice the extensive enhancement (thin arrows) and restricted diffusion (thick arrow).

References: Radiology, Lund University - Lund/SE

Capillary Hemangioma:

-A benign neoplasm composed of endothelial and capillary proliferations.

-The most common vascular lesion of the orbit in the pediatric population.

-These tumors will typically become apparent by one month of age. Many are present at birth.

- Rapid growth ensues for the next 6-8 months followed by a plateau period.

-Most tumors spontaneously involute by adolescence.

-60% of these neoplasms occur in the head and neck. When they occur in the orbit, they are more often extraconal than intraconal.
-Most of these tumors may be observed, however, some become quite large and may cause complications such as amblyopia, visual axis occlusion, or stretching of the optic nerve.

-May be isolated or associated with PHACES syndrome.

**Imaging Findings:**

-Doppler ultrasound is often useful in initial evaluation of capillary hemangiomas. Will show increased arterial and venous flow velocity with low flow resistivity.

-Larger lesions require CT and/or MRI to evaluate the complete extent of the lesion.

-Lesions are typically not well circumscribed.

-CT will show a markedly enhancing mass that is higher in attenuation than muscle and brain due to increased blood content. Bone remodeling may be seen in large lesions. Bone destruction is virtually never seen.

-On MRI, lesions are mildly hyperintense to muscle on both T1 and T2 weighted images. The presence of multiple flow voids help to distinguish this tumor from other lesions.

-With valsalva, lesions may enlarge and worsen proptosis.

![Fig. 4: T1 pre, T1 post, and T2 weighted images of a patient with a large right capillary hemangioma involving the skin, subcutaneous tissues and extraconal space. The lesion diffusely enhances. Note the flow voids seen on the T2 weighted image (arrow).](image)

**References:** Images courtesy of Dr. Kasim Abul-Kasim, M.D., Ph.D., Lund University, SUS, Malmoe, Sweden.
- Rhabdomyosarcoma is an important differential diagnosis. Diffusion weighted imaging is helpful in equivocal cases as rhabdomyosarcomas will restrict diffusion and capillary hemangiomas will not.

- Other differential diagnostic considerations include other vascular malformations, infantile fibromatosis, and infantile fibrosarcoma.

**Choroidal Melanoma:**

- The most common intraocular malignancy in adults.
- Occurs in roughly 5-7 out of every 1,000,000 individuals.

**Imaging findings:**

- Uveal tract melanomas on CT are typically hyperattenuating and have marked enhancement making them relatively difficult to differentiate from choroidal hemangiomas on CT.

![Fig. 5: This diffusely enhancing right ocular mass was pathologically proven to be a uveal tract melanoma.](image)

**References:** Radiology, Lund University - Lund/SE

- Most choroidal melanomas have a characteristic MRI appearance due to their high melanin content and are hyperintense on T1 weighted imaging and hypointense on T2 weighted imaging.
- Amelanotic melanomas do occur in some instances. These will have imaging characteristics similar to other tumors.

- Retinal detachment, associated with uveal tract melanoma, confers a worse prognosis.

**Fig. 6:** Pre contrast T1 fat sat image of the liver of a patient with metastatic choroidal melanoma. Note the innumerable T1 hyperintense hepatic metastases (arrows).

**References:** Image courtesy of Dr. Peter Liu, M.D., University of Michigan Department of Radiology, Ann Arbor, MI, USA.

**Optic Pathway Glioma:**

- The most common intraconal tumor in the pediatric population.

- Constitutes roughly 3% of all orbital tumors.

- Mean age at diagnosis is 5 years old with most tumors becoming apparent by age 20.

- The majority of tumors are WHO grade I juvenile pilocytic astrocytomas. These tumors tend to be slow growing and indolent.
- Tumors in adults rarely occur. If they do occur, they are usually highly malignant.

- The tumor arises from the optic nerve itself, thus, decreased visual acuity is a common presenting symptom.

- Tumors may occur anywhere along the optic tract (orbital, intracranial pre-chiasmal, chiasmal, intracranial post chiasmal).

- Greater than 1/3 of patients with optic gliomas have neurofibromatosis type I. In these cases, the tumors are almost always bilateral.

- Optic pathway glioma is the most common intracranial malignancy in patients with neurofibromatosis type I.

**Imaging Findings:**

- MRI is best for assessing tumor extent.

- On MRI, lesions tend to be iso- to slightly hypointense to muscle on T1WI, and hyperintense to muscle on T2WI.

- Enhancement patterns tend to be homogeneous and may be moderate to marked in intensity.

- In NF1 patients, optic gliomas tend to cause smooth thickening/enlargement of the optic pathway without distorting its native configuration of the nerve. Non syndromic optic gliomas tend to cause fusiform/eccentric enlargement of the optic nerve and cystic components are much more common.
Fig. 7: Axial T1 post, coronal T2 and T1 post, and axial T1 post weighted and T2 Flair images show fusiform enlargement of the left optic nerve (thin arrows). Not the diffuse enhancement. This optic pathway glioma involves the intracranial compartment extending just posteriorly to the optic canal (thick arrow).

References: Radiology, Lund University - Lund/SE

- In patients with bilateral optic nerve gliomas, it is important to assess for other stigmata of neurofibromatosis including sphenoid wing dysplasia, bupthalmos, cutaneous neurofibromas, and neurofibromatosis spots.

- Important differential diagnoses to consider include nerve sheath meningiomas (which typically occur in a different age group), sarcoidosis, and optic neuritis.
Fig. 8: Axial and coronal T1 weighted images of a second patient with an optic pathway glioma arising within the right optic nerve (arrows).

References: Radiology, Lund University - Lund/SE

Optic Nerve Sheath Meningioma:

- While middle aged patients are most commonly affected, pediatric patients with neurofibromatosis type 2 may develop the tumor.

- Patients commonly experience slowly progressive visual loss. This is in contrast to patients with optic pathway gliomas in whom vision usually remains stable or spontaneously improves.

Imaging findings:

- Tumors tend to be slightly hypointense to muscle on T1 weighted imaging and slightly hyperintense to muscle on T2 weighted imaging.

- Often, the optic nerve is seen as a distinct entity that is separate from the mass. This is useful in distinguishing an optic nerve sheath meningioma from an optic pathway glioma in which the nerve will not clearly be seen.
Fig. 9: Axial T1 pre and T1 post and coronal post contrast images with fat saturation in a patient with an optic nerve meningioma. Note how the optic nerve can be readily distinguished from the enhancing mass on the coronal images (arrow).

References: Radiology, Lund University - Lund/SE

 Orbital Lymphoma:

- May occur as a primary lesion, but is more commonly associated with systemic disease, most commonly mucosa-associated lymphoid tissue lymphoma.

- Occurs mostly in older patients.

- Often presents with slowly progressive periorbital swelling and proptosis.

 Imaging Findings:

- May be unilateral or bilateral, extraconal or intraconal.

- Lesions are infiltrative and ill defined.

- Obscuration/destruction of normal orbital anatomy occurs with advanced cases, however, lesions typically conform to the shape of the orbit. Bone destruction is rare.

- Lesions may be radiologically indistinguishable from orbital pseudotumor.

- Lesions tend to be isointense to muscle on T1 weighted imaging and slightly hyperintense to muscle on T2 weighted imaging.
Fig. 10: Axial T1 pre, T1 post, and T2 fat sat images of a 54 year old female with bilateral orbital lymphoma. Note the diffusely infiltrative nature of the lesion which obscures normal orbital anatomy yet conforms to the normal osseous structure of the orbit. There is mild post contrast enhancement (arrow).

References: Radiology, Lund University - Lund/SE

Conclusion:

Non traumatic orbital and ocular masses are uncommon. However, familiarity with some of the more common lesions and their imaging appearances is important as many lesions lead to substantial morbidity and mortality.
References


