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Episode 1

What cranial nerves pass through the superior orbital fissure?

Cranial nerves 3, 4, V1 (ophthalmic) branch of the trigeminal nerve, and 6 pass through the superior orbital fissure. Superior orbital fissure syndrome is dysfunction of these nerves that occurs due to something like trauma, tumor, or inflammation interrupting the function of these cranial nerves.

What cranial nerves pass through the inferior orbital fissure?

This is somewhat of a trick question as there is only one: the V2 (maxillary) branch of the trigeminal nerve. To be more specific, the maxillary branch of the trigeminal nerve gives off the zygomatic nerve, infra-orbital nerve, and orbital branches of the pterygopalatine ganglion that pass through the inferior orbital fissure.

What types of Le Fort fractures involve the orbit?

Le Fort Types 2 and 3 involve the orbit.

Le Fort Type 1: Palate dislocation with transverse fracture involving the lower nasal septum, pterygoid plates and maxillary sinuses.

Le Fort Type 2: Maxillofacial dislocation with oblique fracture involving the inferior orbital rim/orbital floor, nasal bridge and zygomaticomaxillary suture.

Le Fort Type 3: Craniofacial dislocation with fracture through lateral and medial orbital walls, nasofrontal suture, and zygomatic arch.

Note that all Le Fort fractures classically involve the pterygoid plate so if there is no pterygoid plate fracture, there is no Le Fort fracture.

What is a coloboma?

A coloboma is focal discontinuity of the orbital globe (usually posterior aspect of the globe) resulting from failure of the choroid fissure to close. If bilateral colobomas are present need to think of CHARGE syndrome where the C in CHARGE is for Coloboma, H=heart defects, A=atresia choanal, R=retardation developmental, G=genital hypoplasia, E=ear abnormalities. "Starts with eyes and ends with ears".

What entity presents with painful inflammation of the extraocular muscles, involving the myotendinous insertions, most commonly of the lateral rectus muscle?

Orbital pseudotumor which results from idiopathic inflammation of the extraocular muscles. Unlike Grave's the myotendinous insertions are involved. Steroid treatment leads to improvement.

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If findings of orbital pseudotumor are present and also involve the cavernous sinus, what is this disease entity called?

Tolosa Hunt syndrome. This is a painful ophthalmoplegia that involves the orbital apex and adjacent cavernous sinus. Steroid treatment leads to improvement.

What is the most common benign congenital mass of the orbit?

A dermoid of the orbit is the most common benign congenital orbital mass. These typically involve the superolateral aspect of the orbit and contain internal fat density.

What is the most common malignant orbital mass in a child?

Metastatic retinoblastoma to the orbit. Remember with retinoblastoma that there is a problem with the chromosome 13 retinoblastoma suppressor gene that is also associated with osteosarcoma. This means there is an association with facial osteosarcoma following radiation therapy for orbital retinoblastoma. About 1/3 will have bilateral retinoblastomas of the orbit. If you see calcification in the globe of a child this is classic for orbital retinoblastoma.

What is so called trilateral retinoblastoma?

Retinoblastoma involving both orbits as well as the pineal gland.

What is so called quadrilateral retinoblastoma?

Retinoblastoma involving both orbits as well as the pineal gland and suprasellar region.

What is the most common intra-ocular metastatic lesion in an adult?

Metastatic melanoma which often appears as an enhancing soft tissue mass in the posterior globe.

What are some key differences between an orbital lymphangioma and an orbital venous varix?

Orbital lymphangioma: Contains malformed veins and lymphatics together. Does not increase in size with Valsalva. Can show fluid-fluid levels, multiloculated cystic components, and trans-spatial involvement throughout orbit.

Orbital venous varix: Contains veins with weakened walls and lack of functional valves, therefore distends (sometimes massively) with Valsalva. Look for provided history of proptosis or diplopia upon straining or positional changes. May look normal on imaging without Valsalva and very abnormal upon Valsalva so provocative maneuvers can be key for imaging diagnosis with ultrasound, CT or MRI. A top cause of spontaneous orbital hemorrhage. Can also have painful thrombosis.

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What orbital mass is associated with chlamydia psittaci (bird fever)?

Orbital lymphoma which classically is associated with the lacrimal gland and conjunctiva (aka ocular adnexa). Look for a homogeneously enhancing mass with restricted diffusion on MRI.

What is the most common vascular orbital lesion in adults?

Orbital cavernous venous malformation aka cavernous hemangioma. These have weak arterial supply and therefore often show slow initial and delayed, incomplete enhancement. On imaging look for a well-circumscribed lesion that has slow and incomplete internal enhancement with delayed washout, most commonly located in the lateral intraconal compartment, sparing the orbital apex.

True or False: Post-septal orbital infections usually originate from the face.

False. Post-septal orbital infections usually originate from the paranasal sinuses. Pre-septal orbital infections usually originate from the facial tissues.

Note that an orbital subperiosteal abscess often results from ethmoid sinusitis.

What is dacrocystitis?

Dacrocystitis is inflammation of the lacrimal sac. On imaging, look for a rim-enhancing lesion in the lacrimal fossa. Dacrocystitis-induced dilation of the lacrimal sac can cause obstruction and predisposes to staph and strep infections.

Episode 2

What are common causes of "raccoon eyes" on physical exam in a child?

Two leading differential considerations are metastatic neuroblastoma to the orbit as well as basilar skull fracture. The clinical history should obviously guide you towards one entity versus the other. Neuroblastoma is the top cause of orbital metastatic disease in children. These metastases often involve the bony orbital wall, frequently involving the lateral orbital wall and/or sphenoid bone with periostitis described as "hair on end". Look as well for additional sites of metastatic disease, including extradural metastases. Remember that on nuclear medicine, both MIBG and MDP (bone scan) can show sites of metastatic disease through different mechanisms—MIBG as a norepinephrine analogue and MDP through bone remodeling/calcium metabolism.

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Which subtype of breast cancer is more likely to metastasize to the orbit: invasive ductal carcinoma or invasive lobular carcinoma?

Invasive lobular carcinoma is more likely to metastasize to the orbit compared to invasive ductal carcinoma. Orbital metastases may occur many years after initial invasive lobular carcinoma diagnosis and may manifest clinically with visual acuity changes and/or double vision and enophthalmos which is posterior displacement of the globe in relation to the bony structures of the orbit. Note that enophthalmos should make you consider metastatic invasive lobular carcinoma whereas other orbital tumors more commonly cause proptosis.

What is the classic imaging appearance of optic neuritis?

Optic nerve enhancement and increased T2 signal. Optic neuritis is commonly unilateral. This may present with ocular pain.

What is neuromyelitis optica?

Neuromyelitis optica involves bilateral optic neuritis plus myelitis. Clinical symptoms include ocular pain/vision loss and potential paralysis. Is often a relapsing-remitting disease, similar to the clinical course of multiple sclerosis.

What is the classic triad of neuromyelitis optica?

Neuromyelitis has a classic (though not exclusive) triad of optic neuritis, myelitis, and anti-AQP4 (aquaporin 4) antibody.

What is the classic imaging appearance of thyroid orbitopathy?

Enlargement of the extraocular muscles, mostly involving the muscular bellies, with sparing of the tendons. The classic order of prevalence of extraocular enlargement is remembered with the mnemonic IMSLO, as follows: Inferior rectus>Medial rectus>Superior rectus>Lateral rectus>Oblique muscles. Increased volume of intraorbital fat and exophthalmos may also be present.

With thyroid orbitopathy, something like 10% will be euthyroid so TSH testing may not necessarily identify the etiology of orbital findings. More common in females, and more severe presentation in males. In the acute setting, you are more likely to see venous congestion with orbital fat stranding. In the chronic phase you are more likely to have diplopia and restrictive myopathy.

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What are orbital findings associated with neurofibromatosis type 1?

Neurofibromatosis type 1 can cause sphenoid dysplasia and buphthalmos due to a smaller volume orbit. Optic gliomas and Lisch nodules of the iris may also be present. On a frontal radiograph of the skull, the sphenoid dysplasia can produce the "empty orbit" sign wherein the affected orbit looks smaller and insufficient to accommodate an eye, therefore "empty" on radiography.

What are orbital findings associated with a cavernous carotid fistula?

A cavernous carotid fistula creates back pressure to the orbits which can cause pulsatile exophthalmos, proptosis, subconjunctival hemorrhage, and loss of visual acuity with cranial nerve 3, 4, V3, and 6 palsies. On imaging look for findings of orbital congestion that includes retroorbital fat stranding, extraocular muscle enlargement, enlargement of the superior ophthalmic vein, an enlarged cavernous sinus, and possible findings of intracranial hemorrhage.

A classic clinical history for a cavernous carotid fistula is trauma and weeks later development of an erythematous, painful, teary eye. With entities like Ehlers Danlos syndrome this can occur spontaneously. Confirmation is often with angiography via internal carotid contrast injection that shows the cavernous sinus and superior ophthalmic vein. Interventional radiology can also potentially treat this entity with coiling and often the patient will experience rapid or even immediate pain relief. If treatment is delayed, even beyond something like 24-36 hours, optic nerve infarction and blindness can result.

What are some differences between retinal detachment and choroidal detachment?

Retinal detachment often produces a v-shaped morphology in the orbit as the retina has anchor points anterior and posterior in the orbit and detaches between the anchor points. On the other hand, choroidal detachment has more of a u-shaped morphology, or only involves the lateral but not posterior aspects of the orbit as the choroid does not attach at the optic disc. Choroidal detachment diverges at the optic disc whereas retinal detachment converges at the optic disc.

What are top differential considerations for presence of leukocoria?

Leukocoria is an abnormal whitish reflection from the retina. The top cause is retinoblastoma, followed by persistent hyperplastic primary vitreous (due to congenital anomaly with lack of regression of fetal primary vitreous, look for microphthalmos, mis-shaped lens, possible retinal detachment, and so called hyaloid canal extending from abnormal lens posteriorly in the orbit), Coats disease (aka exudative retinitis wherein leaking of lipoproteins causes retinal detachment), and larval granulomatosis and other entities. If you see leukocoria in a child on a board exam think retinoblastoma first.

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What is the top site of metastatic disease from uveal melanoma?

Uveal melanomas have a propensity to metastasize to the liver which is the most common site of metastatic disease.

What is the mechanism whereby permanent diplopia can result from an orbital blowout fracture?

Herniation of the inferior rectus muscle through the orbital floor fracture can cause entrapment and restrictive myopathy, thereby leading to permanent diplopia if not surgically fixed following fracture. Therefore, look for and report presence of inferior rectus hernation in cases of orbital floor blowout fractures.

On a similar note, an orbital apex fracture is in close proximity to the optic nerve and if hemorrhage occurs this can compress the optic nerve and potentially cause permanent blindness.

What is phthisis bulbi?

Phthisis bulbi is a shrunken globe, often related to decreased vitreous or aqueous humor. Causes can be due to trauma causing loss of vitreous or aqueous humor or inflammation or other causes of loss of aqueous or vitreous humor production.

What are classic imaging findings for an optic nerve sheath meningioma?

Tram-track enhancement (most common—results from enhancing tumor around non-enhancing optic nerve) and possible calcifications with possible hyperostosis of the adjacent bone. Clinically may present with proptosis and loss of vision. If bilateral think neurofibromatosis type 2.

Remember that the optic nerve has a blood brain barrier and will only enhance is diseased in some way.

Remember: bilateral optic nerve gliomas->NF1

Bilateral optic nerve meningiomas->NF2

True or false: The majority of cases of orbital lymphoma will also have systemic lymphoma.

True. About 75% of orbital lymphoma patients will have systemic lymphoma.

Orbital lymphoma, like other orbital masses, can present with proptosis and diplopia.

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What are classic features of an orbital fibrous histiocytoma?

An orbital fibrous histiocytoma is a circumscribed enhancing orbital mesenchymal tumor, mostly benign, most common in middle aged adults. A classic history could be an individual with prior treated retinoblastoma, or other orbital tumor treated with radiation, presenting now with an orbital mass that is a post-radiation induced fibrous histiocytoma. If post-radiation history this is more likely to be malignant.

What entity should you first consider in a child or young adult who develops exophthalmos when they get a viral infection?

Orbital lymphangioma due to lymphoid tissue hypertrophy reacting to viral illness. Can also present with proptosis following only mild orbital trauma. Can hemorrhage and cause fluid-fluid levels on MRI. Differential diagnosis includes an orbital rhabdomyosarcoma.

True or false: orbital rhabdomyosarcomas are typically painless?

True. Orbital rhabdomyosarcomas are almost all found in children and teenagers and are more common in males. These present clinically with a rapidly enlarging painless mass with proptosis and visual changes. 5-year survival is over 90% with chemotherapy and radiation.

Bonus tips: Ophthalmic artery enters the orbit through the optic nerve canal with the optic nerve and the ophthalmic vein exits the orbit through the superior orbital fissure. The superior orbital fissure and optic nerve canal are separated by the clinoid process and the superior orbital fissure is more lateral than the optic nerve canal.

Remember the posterior chamber of the eye is the space around the lens that is actually anterior to the vitreous humor.

Retinopathy of prematurity and congenital toxoplasmosis can both result in a small calcified globe.

Bilateral lacrimal gland enlargement can be classically seen with lymphoma, sarcoidosis and Sjogren's.

If you see bilateral lacrimal gland uptake on a nuclear medicine scan and are asked to name the scan, think of a gallium scan first, also consider Tc-99m pertechnetate.

Orbital lymphomas classically show restricted diffusion whereas orbital pseudotumors do not.

Why is the orbit called the orbit if it doesn't orbit anything?

Nobody knows.