FAVORITE CASES GRAND ROUNDS

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Pink sand beach in Bermuda
FINANCIAL DISCLOSURES

- None
CASE # 1

- Well I’ll be dog gone!
CASE # 1

- 41 year old white male
- In for yearly exam, “always had poor vision in left eye”
- Diagnosed with coloboma OS many years ago
- No medical history
- BVA 20/15 OD, hand motion @ five feet OS
CASE # 1

- 45 prism diopter constant LXT
- Other entrance testing and slit lamp findings unremarkable
- Fundus as shown OS, unremarkable OD
**Toxocara Canis (Toxocariasis)**

- Nematode carried by dogs. Usually southeastern US. Major problem worldwide.
- Ingested by humans as the result of eating tainted soil (dog feces) or occasionally undercooked meat.
- Eggs can remain viable in humans for years then activate into mobile larvae. Entire life cycle can be carried out in dogs, larvae only in humans.
- Larvae enter the eye via the blood stream, result in formation of a granuloma.
**Toxocara Canis**

- Two forms: VLM and OLM
- Do not typically co-exist together
- VLM occurs between ages one and four, OLM later in childhood into adolescence
- VLM symptoms of fever, weight loss, vomiting, etc. Vague nature of symptoms often prevents accurate diagnosis
**TOXOCARA CANIS**

- Typical ocular finding is a large granulomatous scar with fibrous bands radiating from the lesion and RPE hyperplasia.
- Associated vitritis and even iritis are possible.
- Treatment options include Albendazole 400mg BID in children (800 adults), Thiabendazole, cryotherapy, and photocoagulation.
TOXOCARA CANIS

- Steroids can be utilized to curb inflammation
- Success of treatment is very limited. Death of the organism leads to a greater inflammatory reaction so treatment is sometimes not indicated
Toxocariasis
TOXOCARIASIS
TRUE COLOBOMA
Toxoplasmosis
“Man.......I am just dying to give somebody Histo today.”
HISTOPLASMOSIS: FLORIDA STRAIN!
CASE # 2

- We’ve got it in the fold!
Case # 2

- 87 year old white female
- Chief complaint “blurry vision in right eye”
- History of cataract extraction and yag capsulotomy OS
- HTN
- Entrance testing non-contributory
CASE # 2

- BVA 20/70 OD, 20/20 OS
- NS cataract OD, centered IOL OS with open capsule
- Fundi as shown
CHOROIDAL FOLDS

- Shrinkage or compression of the scleral tissue leading to folding of the attached choroid/Bruch’s membrane/RPE complex
- Bilateral folds usually the result of age related scleral shrinkage in hyperopic females (as in this case). Benign
- Unilateral folds are more ominous and can be the result of orbital tumors, hypotony, CNVM’s, or posterior scleritis
CHOROIDAL FOLDS

- Alternating light and dark bands, show up clearly on IVFA as light and dark streaks
CHOROIDAL VS. RETINAL FOLDS

- **Choroidal...**
  - Usually roughly horizontal
  - Usually posterior pole
  - Light and dark streaks
  - Visible on IVFA

- **Retinal...**
  - Often stellate alignment
  - Can be anywhere
  - Similar but finer
  - Visible on IVFA only if vascular traction
CHOROIDAL FOLDS DUE TO HYPOTONY
WOUND LEAK (POST CATARACT SURGERY) WITH CHOROIDALS: SHALLOW / FLAT CHAMBER
WOUND LEAK POST REPAIR: CHAMBER BACK TO NORMAL DEPTH
Choroidal effusion (different pt.)
Choroidal effusion
Verhoeff’s streaks in choroidal effusion
HYPEROPIC CHOROIDAL FOLDS
PATTON’S FOLDS IN PAPILLEDEMA
CHOROIDAL FOLDS IN POSTERIOR SCLERITIS
CASE # 3

- Vasculitis? I don’t think so.........
Case # 3

- 23 year old AA female presenting for low vision consultation
- BCVA of 20 / 40 -2 OD and 20/800 OS
- Large, constant LXT, significant VF constriction OS > OD
- Other entrance tests and slit lamp findings unremarkable. Fundus appearance......
CASE # 3
Leber’s Hereditary Optic Neuropathy

- Hereditary mitochondrial disease process affecting the retinal ganglion cells
- Maternal inheritance pattern (mitochondrial DNA in embryo comes only from the egg)
- Genetic point mutations that have been fully identified / mapped
- Because of inheritance pattern, males can not pass to offspring
LHON

- Males more commonly afflicted but females can be
- Typically strikes in early adulthood, but can strike later
- Most commonly affects one eye followed by the fellow eye within several weeks
- Clinical trials ongoing with gene therapy; some encouraging early results
LHON

Reported associations

- Can get pseudopapilledema secondary to peripapillary NFL swelling
- Reports of vasculitis (older literature) and pseudovasculitis
- What are we looking at here? Pseudovasculitis from redundant ILM

Our patient......
LHON

- The ganglion cells die so quickly that the redundant internal limiting membrane of the retina collapses and folds on itself, especially around vessels.

- This appearance lasts several months then tends to go away.
CASE # 4

“Looks like drops of water on a windshield”
CASE # 4

- 60 YO Caucasian female
- In for exam to check on cataracts
- Arthritis, Asthma, OSA

- BCVA 20/25- OD, 20/20 OS
- IOP 13,12
- All preliminary testing normal
- 1+ NS, trace PSC OU
- ONH Drusen OU
- Multiple, small PED’s OU within the arcades
CASE # 4
MULTIPLE IDIOPATHIC PED SYNDROME

- Very rare, with few cases in the literature
- Multiple PED’s with little or no neurosensory retinal detachment
- Usually females, often related to pregnancy
- One theory is a variant of ICSC involving only the RPE
- May be related to sleep apnea, which has been linked to ICSC
CASE # 5

“We all have it doc....”
CASE # 5

- 58 year old Caucasian male
- Complaining of flashes OD for four days. Has floaters, but longstanding with no increase
- Told by a retinal specialist 17 years prior that he has a progressive retinal disease but would not go “completely blind”
- BCVA of 20 / 20 in each eye
- IOP 18 OD, 19 OS
- Entrance testing unremarkable
- Anterior segment unremarkable OU
- Posterior segment reveals significant drusenoid changes OU with pigment mottling OU
CASE # 5

- Posterior segment also reveals a fresh PVD OD with no holes, tears, or breaks
- Posterior segment appearance........
Case # 5
CASE # 5
CASE # 5

- So what have we here.......?
- Doyne’s Honeycomb Dystrophy!
- AKA: Mallattia-Leventinese Dystrophy
Case # 5

- Doyne’s Honeycomb Dystrophy
- Described by Doyne in England in 1898

- Malattia-Leventinese Dystrophy described by Alfred Vogt in Switzerland in 1925

- Now believed to be phenotypic variants of the same condition
- Both caused by genetic mutation in EFEMP1 gene
Case # 5

- The affected gene encodes a protein that is expressed in the retina and the RPE
- Leads to drusen formation early in life
- Located all throughout the posterior pole, including nasal to the disc

- The drusen coalesce over time leading to radial (MLD), honeycomb (DHD), or mosaic patterns (MLD and DHD)
- Symptoms are rare until early middle age
- Macula is affected with drusen, so can get vision loss and SRNVM
CASE # 5

- Because the condition is genetic, the role of nutritional supplements is unclear
- Manage with regular follow-up, amsler at home, possibly supplements
CASE # 5

- Differential diagnoses:
  - 1) AMD: affects macular area only. More likely to have RPE loss, more likely to have an SRNVM

- 2) Familial or basil laminar drusen: cluster in groups throughout the posterior pole, especially in arcades, but do not coalesce. No effect on vision.
CASE # 5

- This patient monitored over time with regular follow-up and amsler
- Remains symptom free with good vision
Another Doyne’s Example
Case # 5
Another Doyne’s example
CASE # 5
ANOTHER DOYNE’S EXAMPLE
CASE # 5: ANOTHER DOYNE’S EXAMPLE
Case # 5
Familial / Basil Laminar Drusen Example
CASE # 5
AMD EXAMPLE
CASE # 6

“Are you sure this picture is not upside down?”
CASE # 6

- 43 year old AA male complaining of poor vision, pain, itching and watering OD
- History of RD OD due to trauma and subsequent repair 10 years prior
- Taking Naphcon-A and Hydrocodone
- BCVA of LP OD, 20 / 30 + OS
- IOP 50 OD, 15 OS
- EOM’s normal
- VF normal OS
- Pupil unreactive OD, but “reverse APD” OS
- White appearance to upper half of iris with the naked eye
CASE # 6

- Anterior segment evaluation
  - Eyelid edema OD
  - Aphakic OD
  - Solid, clear bubble in central AC OD with top half of chamber filled with a white substance
  - 2+ Conj. injection OU

- “reverse pseudohypopyon”
  - Anterior segment OS unremarkable
  - Posterior segment OS unremarkable. No view of fundus OD

- Anterior segment appearance
CASE # 6
Case # 6
CASE # 6: LOOKS LIKE STYROFOAM!
CASE # 6

- So what have we here.........?
- Silicone oil emulsification!
CASE # 6

- Silicone oil tamponade is routinely used in complicated retinal detachment repair
- It must be removed later to avoid potential complications
- One complication is emulsification
CASE # 6

- Emulsification basically means turning to soap
- Silicone oil turns into smaller droplets of soap-like material when surface tension decreases significantly
- This occurs when the oil contacts various biological products
CASE # 6

- Proteins, lipids, and phospholipids
- Particularly HDL
- Occurs up to 50% of the time if oil is not removed
- Happens in the vitreal cavity, then droplets travel to the AC, especially in an aphakic patient
CASE # 6

- “inverse pseudohypopyon”
- Leads to corneal endothelial toxicity and edema, band keratopathy, and increased IOP / glaucoma
- In a seeing eye, prompt removal is indicated
CASE # 6

- This patient was given cycloplega for ciliary spasm
- Also offered pressure lowering agents for comfort, but deferred
- Sent for consideration of surgical removal of emulsified oil
- Lost to follow up
ANOTHER EXAMPLE OF EMULSIFICATION
MORE AC oil with AC IOL (say that 10 times quickly!)
CASE # 7

- Maybe it IS a tumor!
CASE # 7

- 39 year old white male
- Chief complaint: spot on right eye
- Reports that spot has been present since high school but has just recently gotten much larger
- BCVA: 20/20 OD & OS
- Entrance testing unremarkable
CASE # 7

- Slit lamp exam revealed findings seen as well as an inferior cortical cataract OD, OS unremarkable
- DFE unremarkable OU
- B-scan ultrasound of posterior segment OD unremarkable
AMELANOTIC IRIS MELANOMA

- Pathology revealed spindle cell morphology (relatively non-aggressive)
- Systemic work-up revealed no sign of metastases
- Iris melanomas account for only a small portion of uveal tumors (< 10%)
- Seen most frequently in blue irides
IRIS MELANOMA

- Inferiorly located due to sun exposure, may distort pupil
- Rarely metastasize (3-5%) because visibility typically leads to early detection
- Satellite lesions can be seen, increased IOP common with seeding of tumor cells into the TM
- Differential diagnoses include iris nevi, Lisch nodules, Koeppe and Busacca nodules, etc.
- Sector iridectomy performed for complete resection (see post-op picture next)
- Pt. did very well with an opaque contact lens to give him a new pupil
ANOTHER IRIS MELANOMA
THE END!

- Any questions or comments........

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