I. Case History

❖ Patient demographic
  ➢ 48 year old Hispanic female

❖ Chief complaint
  ➢ Dry eyes OU
  ➢ Headaches with increased frequency mostly in the frontal and temporal area.
  ➢ Blurry vision in the right eye more than the left, both with and without glasses.

❖ Ocular and Medical History
  ➢ Pertinent Ocular History:
    ▪ LEE: 9 months ago at an outside provider, cataract extraction OU recommended at that visit
  ➢ Medical History:
    ▪ (+)HTN, HLD, hypothyroidism
    ▪ (-)DM

II. Pertinent findings

❖ Clinical:
  ➢ Entering visual acuity with correction
    ▪ OD: 20/50+
    ▪ OS: 20/50+2
  ➢ Pupils: PERRL, (+)grade 1+ APD OD
  ➢ Extraocular motilities:
    ▪ OD: restriction in superior, supero-temporal and supero-nasal gazes; otherwise full
    ▪ OS: full without restrictions
  ➢ Confrontation Visual fields: Full to transilluminator OD+OS
  ➢ Refraction:
    ▪ OD: +1.25-1.00x090 ADD+2.25
      • DVA 20/30 PHNI
    ▪ OS:+1.25-3.00x090 ADD+2.25
      • DVA 20/30 PHNI
  ➢ Goldmann Applanation Tonometry:
    ▪ OD: 12 mmHg, OS: 12 mmHg

❖ Physical exam:
  ➢ Slit Lamp Exam
    ▪ Lids/Lashes: trace capped meibomian glands OU
    ▪ Conjunctiva: clear OU
    ▪ Cornea: clear OU
    ▪ Iris: flat and intact OU
    ▪ Anterior chamber: deep and quiet OU
    ▪ Angles 4x4 OU
  ➢ Dilated fundus exam (1 gtt 1% tropicamide and 2.5% phenylephrine OD/OS)
    ▪ Lens: trace nuclear sclerosis OU
    ▪ Vitreous: clear OU
C/D:
- OD: 0.35R pink and distinct, healthy, rim tissue elevated nasally, (-)edema
- OS: 0.30R, pink and distinct, healthy, rim tissue elevated superior-nasal, (-) edema
Macula:
- flat and clear, (-)FR OU
Vessels:
- Normal caliber OU
Posterior pole:
- Choroidal folds OD>>OS
Periphery:
- OD: ¼ DD chorioretinal scar in the inferior arcade, WsP temporal and nasal; otherwise flat and intact with no holes, tears or detachments.
- OS: WsP temporal, nasal and inferior-nasal; otherwise flat and intact with no holes, tears or detachments.

Macular OCT
- OD: sub-RPE disturbance with multiple folds, likely choroidal
- OS: trace ERM

ONH OCT:
- OD: ST and IT thinning
- OS: no thinning noted

B-scan
- Thickened choroid/sclera OD>OS, (-) detachments, (-) orbital mass

HVF 24-2
- OD: reliable, dense superior arcuate and inferior-temporal defect
- OS: reliable, superior defect

Fluorescein Angiography:
- Alternating hyperfluorescent and hypofluorescent bands in the choroid OD>OS

Exophthalmometry:
- Base 90, OD 15 mm, OS 14 mm

Axial Length:
- OD: 22.62 mm, OS: 22.68 mm

Lab work:
- TSH 3.342 (0.55-4.78) Negative

Radiology studies
- MRI:
  "There is a mass along the sphenoid wing which is extra-axial. The approximate size of the mass is 1.6x1.3x1.5 cm. This extra-axial mass shows homogenous enhancement and most likely differential is a meningioma. Other differentials could include metastatic lesions but these are felt to be less likely. The mass is compressing the apex of the right orbit and is compressing the right optic nerve. There is a suggestion of a possible invasion of the cavernous sinus regions also. There is a very mild mass effect on the medial temporal lobe on the right. There is no evidence of signal change within the temporal lobe. The right optic nerve is being
compressed by the mass. The optic chiasm appears to be intact without abnormality. The visualized optic tracts also show no focus abnormality.

III. Differential Diagnosis:

- Differential Diagnosis for this case:
  - Primary and leading diagnosis: Compressive lesion
  - Posterior scleritis (ruled out by symptoms and exam findings)
  - Thyroid Eye Disease (ruled out by bloodwork)

- Differential diagnosis for choroidal folds (Ling Yung, et al, 2005):
  - Papilledema
  - Posterior scleritis
  - Orbital/ocular mass
  - Choroidal neovascular membrane associated with age related macular degeneration
  - Scleral buckle
  - Hyperopia
  - Hypotony

IV. Diagnosis and Discussion:

- Choroidal folds develop from compressive stress on the choroid, Bruch’s membrane and the retina. This causes symptoms such as metamorphopsia as well as reduced vision if there is a hyperopic shift or if the choroidal folds are found within the macula. (Jacobsen et al, 2015)
  - Choroidal folds appear narrow initially, broadening and whitening in appearance with time. (Jaworski et al, 1999)

- It is likely that the above patient had idiopathic choroidal folds in each eye, with exacerbated folds in the right eye due to the meningioma compressing on the right optic nerve.
  - In most cases patient’s with a meningioma will present with headaches, seizures, hemiparesis, personality change, sphincter problems and ocular motor dysfunction. Rarely is the initial diagnosis made on finding choroidal folds alone. (Ling Yeung et al, 2005)

- Raised intracranial pressure can be an independent cause of chorioretinal folds. Chorioretinal folds in the above patient may have been a sign of localized elevated intracranial pressure within the subarachnoid space surrounding the optic nerve. The tumor may have caused fluid to be trapped in the subarachnoid space of the optic nerve elevating the pressure within this specific area. If this were the case, the pressure in the brain would not be elevated. (Taban et al, 2006)
  - It is rare but possible for chorioretinal folds to precede signs of optic nerve edema. (Taban et al., 2006)

V. Treatment and Management

- Treatment
  - A referral was made to neuro-ophthalmology and neurology departments to further assess and evaluate the mass found on MRI and any ocular involvement.
It is likely that the patient will either undergo radiation therapy or surgery to remove the mass/prevent further growth.
- Results pending

The patient will return to eye clinic in 3 months to assess changes in vision and visual fields. It is important to closely monitor the patient through the coming years because patients with chronic chorioretinal folds can develop choroidal neovascularization. (Olsen et al, 2014)
- Patients with chronic chorioretinal folds, more often older patients, have been found to develop chorioretinal fold maculopathy, exhibiting a yellow luteal macular appearance or RPE atrophy. Fluorescein angiography testing reveals both staining and mild leakage in patients found to have this maculopathy. The visual acuity of patients with chorioretinal folds maculopathy did not improve with treatment anti-VEGF agents. (Olsen et al, 2014)

Long term follow up is recommended to monitor the reoccurrence of a mass.
- In some cases, despite treatment and removal of the mass, choroidal folds and acquired hyperopia remain present. This may be due to scleral remodeling after long-standing compression on the sclera and choroid. (Jacobsen, et al 2015)

Management
- Standard evaluation in patients with choroidal folds includes a complete dilated exam, intraocular pressure reading, fluorescein angiography, OCT and B-scan.
  - B scan may show flattening of the posterior pole, thickening of the choroid and/or sclera and enlarged optic nerve subarachnoid space.
  - Fluorescein angiography will differentiate retinal folds from choroidal or chorioretinal folds. Folds involving only the retina will not be seen on FA.
    - Folds within the pigment epithelium cause some sections to have increased density of pigment epithelium. This obscures the light and causes some areas to seem darker than others on FA. (Jaworski et al, 1999)
  - OCT can provide information as to whether the sensory retina is involved in the folds seen on funduscopic examination. (Gasperini et al 2006)
    - It is important to assess whether the inner retina is flat on OCT imaging to determine whether the folds are choroidal, chorioretinal or retinal. (Giuffre et al, 2007)
  - MRI should be performed only in cases where no other etiology is found to explain the presence of choroidal or chorioretinal folds.
    - Patients with newly acquired hyperopia with choroidal folds should undergo an MRI of the brain. (Jaworski et al, 1999)
  - A lumbar puncture is only necessary if imaging is inconclusive. (Taban et al, 2006)
    - Rarely, a patient with increased intracranial pressure can present with choroidal folds and no optic nerve head swelling. Thus, lumbar puncture must be considered in patients who have any symptoms of increased intracranial pressure (Ling Yeung et al, 2005).
    - Only after all testing is found to be unremarkable, can one diagnose choroidal folds as idiopathic
• Idiopathic folds are usually symmetrically distributed in the posterior pole of each eye. (Jaworski et al, 1999)

VI. Conclusion

- Choroidal folds are associated with many different ocular and systemic conditions. Although choroidal folds are a rare finding, thorough testing is needed in order to properly assess their cause.
- Patients presenting with choroidal folds will frequently need to be co-managed with ophthalmologists, neurologists and/or neuro-ophthalmologists.

References


