Diagnosis and Management of Choroidal Folds

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Abstract

Choroidal folds are a rare clinical sign found in the retina that can present unilaterally or bilaterally. Its fundus appearance consists of alternating light and dark bands found in the macula that are usually oriented horizontally but can also take on a circumferential or radial pattern.[1] Even though choroidal folds often present idiopathically, they can also be an indication of a serious medical condition. Orbital tumors, hypotony, and idiopathic intracranial hypertension are some of the many possible etiologies.[2] As a result, when choroidal folds are first detected, it is important to complete a proper work up to rule out a potential vision or life threatening condition. This case report will review important steps a clinician should take before reaching a diagnosis of idiopathic choroidal folds as well as review ancillary testing needed to properly diagnose and manage this condition.

Case Report

A 69 year old male presented with blur in both eyes. The blur was constant at distance and near but he reported relief of symptoms with his current glasses. His last comprehensive eye exam was two years ago. His ocular history consisted of refractive amblyopia in the left eye, and mild cataracts in both eyes that were not visually significant. He had no history of ocular surgeries. His medical history consisted of hypertension, coronary arteriosclerosis, hyperlipidemia and hereditary thrombophilia. Systemic medications included apixaban, atorvastatin, hydrochlorothiazide and sildenafil with a previous allergic reaction to rivaroxaban.

Best-corrected visual acuity was 20/20 in the right eye and 20/40 in the left eye which was stable to the visual acuity measured three years ago. In addition, no changes were found from his habitual prescription of +3.25 -1.00 x 106 in the right eye and +7.50 -1.50 x 039 in the left eye on manifest refraction. Pupils were equal, round, and reactive to light with no afferent pupillary defect noted. Confrontation visual fields were full to finger counting in all quadrants and extraocular muscles were full in all fields of gaze with no diplopia or pain in the right and left eye. Cover test at distance and near revealed no tropias. Slit lamp evaluation revealed trace capped glands and pingueculae in both eyes, otherwise anterior segment was unremarkable. Intraocular pressure was found to be 17mmHg OD and 18mmHg OS at 9:01am with Goldmann applanation tonometry (GAT).

A dilated fundus exam was completed with 1 drop of 1% tropicamide and 1 drop of 2.5% phenylephrine in each eye. Posterior segment evaluation revealed a 1+ nuclear sclerotic cataract and trace anterior cortical spoking in both eyes. Optic nerve heads were both pink and healthy with distinct margins and a cup-to-disc ratio of 0.30 round. On the fundus exam, right eye findings presented with faint alternating light and dark bands that extended from the macula to the optic nerve head. These findings were not noted in previous dilated exams. The left macula was flat and avascular. The peripheral retina was intact with no holes, tears, or retinal detachments bilaterally. Fundus photos were completed at this visit allowing for better visualization of the alternating colored bands (see figures 1A-1C). Fundus autofluorescence (FAF) imaging of both eyes was also completed showing a diffuse, mild hyperfluorescent signal with no focal areas of hypo or hyperfluorescence (see figures 2A-2B).

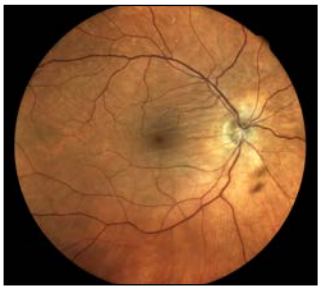

Figure 1A. Confocal Scanning Image of choroidal folds OD

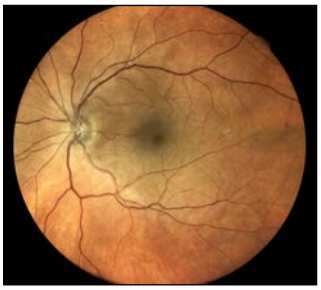

Figure 1B. Confocal Scanning Image fundus photo of a normal fundus OS

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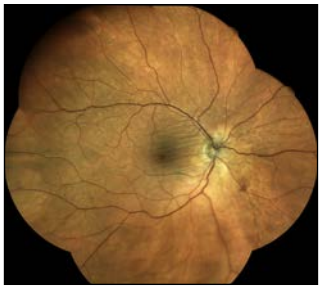

Figure 1C. Confocal Scanning Mosaic Image of choroidal folds OD

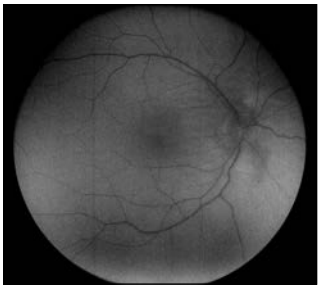

Figure 2A. Confocal Scanning Image with autofluorescence of choroidal folds OD

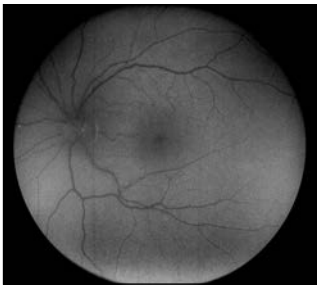

Figure 2B. Confocal Scanning Image with autofluorescence of a normal fundus OS

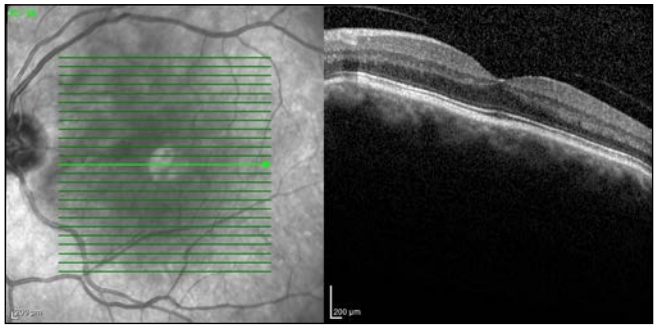

Figure 3A. Horizontal optical coherence tomography (OCT) macular scan of choroidal folds OD

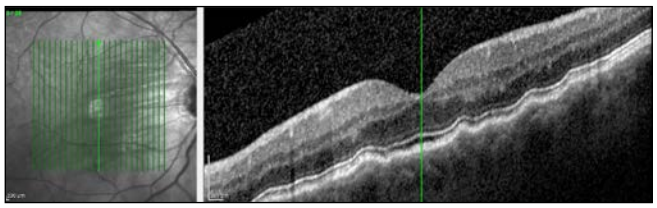

Figure 3B. Vertical optical coherence tomography (OCT) macular scan of choroidal folds OD

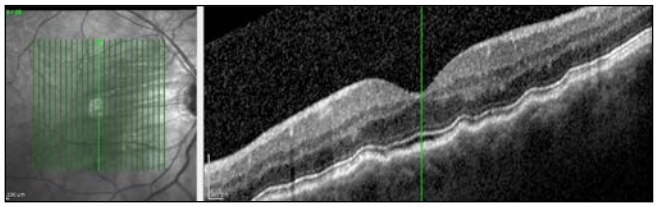

Figure 3C. Horizontal optical coherence tomography (OCT) macular scan of choroidal folds OS

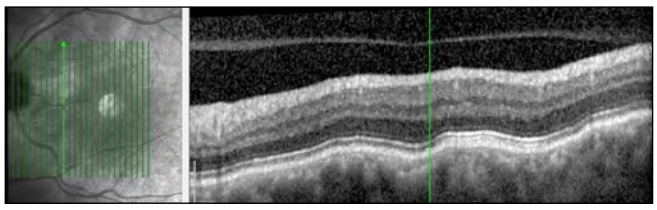

Figure 3D. Vertical optical coherence tomography (OCT) macular scan of choroidal folds OS

Due to the macular changes that were noted in the right eye, a macular OCT of both eyes was completed. The macular scan of the right eye horizontally appeared normal, with normal foveal contour and no obvious folds. When the macula was scanned vertically, normal foveal contour with chorio-retinal folds that correlated with the light and dark alternating bands seen on fundus photography were revealed. (see figures 3A-3B). Macular scan of the left eye also showed a normal foveal contour horizontally while the vertical scan showed large ripples in the choroid, consistent with mild choroidal folds. (see figures 3C-3D). As a result, despite no abnormal findings seen in the left eye on fundus photography, the macular scan revealed asymmetric but bilateral choroidal folds. There were no signs of a choroidal neovascular membrane (CNVM) or macular edema in either eye.

With a diagnosis of bilateral asymmetric choroidal folds, further investigation was completed to help determine etiology. The patient was asked about headaches, nausea, and transient visual obscurations, which were all, denied. TSH, T3, and T4 levels were all tested and found within normal ranges. An MRI of the brain and orbit was completed and showed mild microvascular ischemic changes but no tumor was detected. As a result, with no symptoms reported by the patient, stable vision, no optic nerve edema, and a bilateral presentation of choroidal folds, the patient was educated on the idiopathic finding and asked to return in six months to monitor the condition by repeating dilation, fundus photos, and macular OCT. The patient was also informed on the importance of follow up and asked to return to clinic sooner if any changes in vision or new headaches occurred.

At the six month follow up the patient reported no changes in vision and no new symptoms. Best-corrected visual acuity was stable at 20/20 in the right eye and 20/40 in the left eye. Dilated fundus examination, fundus photos, and macular OCT were all stable at the follow up visit. Due to his stable findings and absence of symptoms, the patient was scheduled to return to clinic in one year for his annual comprehensive eye exam and to continue monitoring.

Discussion

Choroidal folds are a rare clinical sign that can be found in the posterior retina. Although rare, it is important to identify them due to their possible association with vision threatening, or even life threatening, pathological conditions.[2] Choroidal folds were first described clinically by Edward Nettleship in 1884 in a patient who had papilledema secondary to a space occupying lesion.[3] Since this observation, choroidal folds have been identified in a wide variety of conditions. Some of the possible causes include hypotony, posterior scleritis, idiopathic intracranial hypertension, choroidal neovascularization, thyroid eye disease, trauma, ocular surgery, and acquired hyperopia (see table I).[2,4-6] Choroidal folds can present unilaterally or bilaterally showing no significant preference to a specific race or age group.[7]

Etiologies of Choroidal Folds[2,10,19]

- Idiopathic
- High hyperopia
- Idiopathic intracranial hypertension
- Thyroid eye disease
- Posterior scleritis
- Orbital tumors
- Choroidal tumors
- Uveal effusion syndrome
- Uveitis
- Scleral buckling surgery
- Hypotony
- Age related macular degeneration
- Orbital myositis
- Orbital cellulitis

Although a large study has yet to be completed, several smaller studies have found that patients presenting with unilateral choroidal folds have a higher frequency of being linked with significant orbital or ocular disease compared to a bilateral presentation.[7] One study found that in a sample size of 54 patients, the top three etiologies of bilateral presenting folds were idiopathic, hyperopia, and macular degeneration. On the other hand, the top three etiologies for unilateral presenting folds were ocular tumors, posterior scleritis, and hypotony.[7] Although these studies were completed before the use of OCTs, they provide a good example of how differentials can change depending on the presentation of the choroidal folds.

Clinical Appearance and Pathophysiology

On funduscopy, choroidal folds present as alternating light and dark bands found in the posterior pole. The darker colored striae correspond to troughs of the folds where the RPE has been compressed. The lighter colored striae correspond to elevated ridges where the RPE has been stretched thin.[8] Choroidal folds are usually oriented horizontally but can also take on a circumferential or radial pattern.[2] Although they can vary in length they rarely extend beyond the equator and over time they tend to become wider and smoother in appearance.[2] Anatomically, choroidal folds are undulations of the inner choroid, Bruch's membrane, and the retinal pigmented epithelium. The undulations can also be seen in the neurosensory retina but this is not pathognomonic.[9] This lack of involvement from the neurosensory retina is a possible explanation why some patients with choroidal folds are asymptomatic and still maintain good vision. When the neurosensory retina is involved, the condition is referred to as chorioretinal folds.[10]

There are many theories that attempt to explain how choroidal folds arise. One theory believes that folds arise when there is a change in the anatomic relationship between the sclera and the choroid.[9] Some examples of these changes are an intraocular or extraocular force pushing on the sclera, thickening or shrinkage of the sclera, or thickening of the choroid. These changes can occur as a result of a pathological condition, ocular surgical procedure, or idiopathically.[2] Although pathophysiology can vary depending on etiology, it is well accepted that these changes, along with the strong adherence between Bruch's membrane and the underlying choriocapillaris, are what result in choroidal folds.[6]

Differential Diagnosis

It is important to differentiate choroidal folds from retinal folds. Although they can have a similar fundus appearance, retinal folds vary greatly from choroidal folds in their pathophysiology. Retinal folds are congenital or secondary folds that only involve the neurosensory retina. They are often associated with an abnormal vitreous or vitreal-retinal interface and are more likely to affect visual acuity or cause distortion in vision when located over the macula.[2,10] Although retinal folds can present with a horizontal striated appearance similar to some choroidal folds, the striations associated with retinal folds tend to have a more narrow and translucent appearance.[2] Some common etiologies of retinal folds are epiretinal membranes, optic disc swelling (known as Paton's lines), rhegmatogenous or tractional retinal detachments, retinopathy of prematurity, and toxocariasis.[11]

On fluorescein angiography, the presence of hyper and hypofluorescence of the choroid during the early stages of the test confirms a diagnosis of choroidal folds. Retinal folds do not appear on fluorescein angiography due to the lack of involvement from the retinal pigment layer.[3] When completing a macular scan with an OCT, one could easily differentiate between each diagnosis by assessing which retinal layers are undulated (see figures 4A-4B).

Imaging

In the past fluorescein angiography (FA) was often needed to confirm a diagnosis of choroidal folds.[3] Fortunately, with the increasing accessibility to SD-OCTs, FAs are now rarely needed unless there is a suspicion for a choroidal neovascular

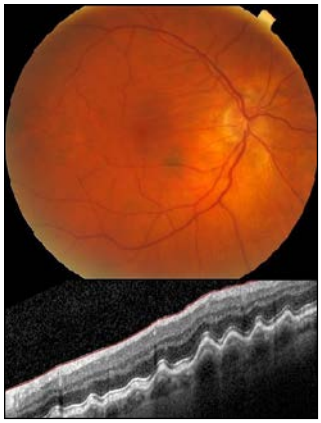

Figure 4A. Fundus photo of choroidal folds with its corresponding macular OCT image

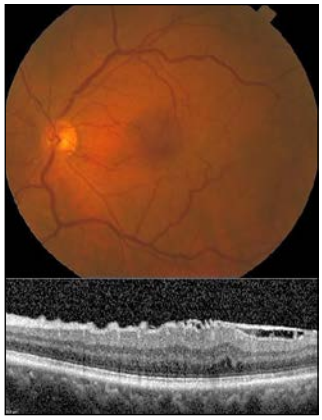

Figure 4B. Fundus photo of retinal folds with its corresponding macular OCT image

membrane.[4] The SD-OCT has also improved the detection of folds that would have otherwise been missed. In addition, due to its less invasive nature, the SD-OCT has facilitated the capability for a clinician to document and repeat imaging at follow up visits. As a result, the SD-OCT has greatly improved the ability for clinicians to detect and manage a patient with choroidal folds.[12]

When completing a SD-OCT scan to confirm the involvement of choroidal folds, it is important to place the scanning lines perpendicular to the orientation of the suspected folds. By doing this the undulations in the suspected area will become more prominent, making it easier to confirm the diagnosis.[13] Figures 5A and 5B show a macular scan of a patient with choroidal folds superior to the optic nerve head. On image 5A the OCT scan was completed with the default horizontal scanning orientation. On image 5B, the scan was completed with the scanning line placed perpendicular to the striations of the choroidal folds. As seen below, the undulations appear more prominent on image 5B than image 5A making it easier to confirm a diagnosis of choroidal folds. Also, it is important to use this as an example that choroidal folds can occur not just in the macula, but other areas as well, such as around the optic nerve or in the arcades.

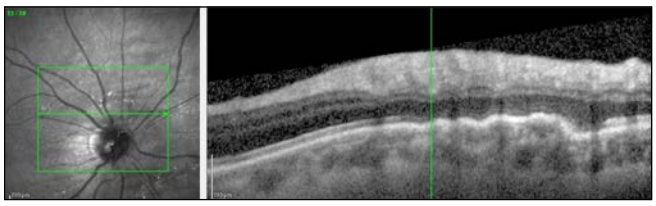

Figure 5A. OCT image of choroidal folds with line scan oriented parallel to horizontal choroidal folds located superior to the optic nerve head OD

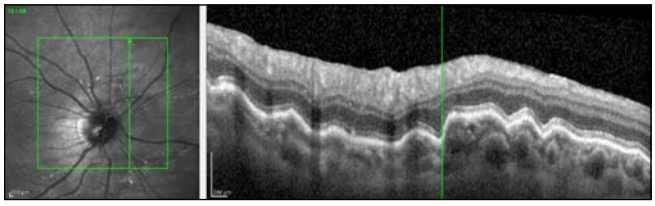

Figure 5B. OCT image of choroidal folds with line scan oriented perpendicular to horizontal choroidal folds located superior to the optic nerve head OD

Etiologies

Once the presence of choroidal folds has been confirmed, there are important questions and tests one should complete in order to help determine a possible cause. With an extensive list of potential etiologies, these questions and tests should be guided by the patient's chief complaint, case history, and abnormal findings throughout the course of the exam (see table II).[2,4] The proper work up for a patient with choroidal folds will be different every time depending on the suspected etiologies and in the likely case that no pathological condition is found, choroidal folds can also be idiopathic in nature.[2,4] Still, even though choroidal folds are commonly found to be idiopathic, it is only once an extensive investigation has been completed that a clinician can consider this a possibility.[2]

Work Up for Choroidal Folds[2,4]
1-Visual acuity
2-Pupils
3-Extraocular muscle movements
4-Visual field
5-Intraocular pressure
6-Dilated fundus examination
7-Colored Fundus photos
8-Optical coherence tomography of macula
9-A-scan/B-Scan ultrasound
10-Fluorescein angiography
11-Magnetic resonance imaging (MRI)

A patient that presents with a new onset of choroidal folds should cause a clinician to put intraorbital conditions high on their list of differentials such as orbital tumors, Grave's disease, and orbital myositis.[2] Important signs that can help gauge the level of concern for an intraorbital pathological involvement are the presence of an afferent pupillary defect, an extraocular muscle restriction, proptosis, and changes in visual acuity.[2,3] Choroidal folds secondary to an intraorbital condition are more commonly found in patients who also present with proptosis secondary to an anteriorly located orbital tumor.[14] As a result, even if all tests are unremarkable, it is important to complete an MRI on these patients, with greater urgency for patients who present with new onset of unilateral choroidal folds.[2]

Choroidal folds can also present secondary to ocular hypotony. Ocular hypotony can occur due to overfiltration, ocular surgery, or after ocular trauma where penetration of

the globe has occurred.[13] Overfiltration after trabeculectomy surgery is one of the most common causes of choroidal folds secondary to hypotony. This is especially true in patients where antimetabolite adjuncts were used during the surgical procedure.[13] "Hypotony maculopathy" is a term that was first coined by Gass in 1977 and it is used to describe when choroidal folds secondary to hypotony extend into the macula causing a reduction in vision.[13] Fortunately, if normalization of the intraocular pressure is regained in a timely manner, most of the changes in vision can be expected to resolve. However, in situations where normalization of the intraocular pressure is delayed for over 4 months, permanent macular chorioretinal changes, as well as changes in vision, can occur.[13] As a result, management of this condition should be directed towards promptly finding a resolution to the cause of hypotony.

Choroidal folds in an asymptomatic patient who has completed imaging and has ruled out pathological conditions can be classified as idiopathic. However, before reaching this diagnosis, it is important to take the time and consider the possibility of idiopathic intracranial hypertension (IIH).[8,15,16] In most cases, patients with IIH will present with symptoms of headaches, nausea, and bilateral disc edema, but it may prove difficult to detect when only mild symptoms are present. Although rare, it is possible for IIH to present with choroidal folds in the absence of papilledema.[8] It has also been hypothesized that the presence of choroidal folds with a lack of papilledema or any other pathological conditions could alternatively be associated with a past episode of IIH that has since resolved.[17] This is why it is important to confirm the absence of IIH symptoms when evaluating choroidal folds. In addition, searching for an enlarged optic nerve subarachnoid space or flattened posterior ocular wall when completing a B scan or MRI can also be helpful.[16,18] Some studies suggest that a lumbar puncture should always be considered before diagnosing a patient with idiopathic choroidal folds.[1] Whether or not a lumbar puncture is completed, all patients classified with idiopathic choroidal folds should be regularly monitored for any changes.[8]

Conclusion

Choroidal folds are a clinical sign that requires a thorough evaluation to rule out possible underlying causes.[9,15] Orbital tumors, hypotony, idiopathic intracranial hypertension, posterior scleritis, thyroid eye disease, trauma, or ocular surgery are important etiologies a clinician should consider. In a new presentation of unilateral choroidal folds it is especially important to rule out the involvement of an orbital tumor by completing an MRI of the brain and orbit.[2] Once a clear MRI has been confirmed, a lumbar puncture should also be considered if there is suspicion for the involvement of idiopathic intracranial hypertension.[1,17] The use of OCT has also changed the way choroidal folds are detected and are an important tool to differentiate from retinal folds. With a careful case history and proper testing or imaging, clinicians should confidently be able to manage choroidal folds after determining the cause.

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POST-COURSE TEST

Diagnosis & Management of Choroidal Folds

1 Which of the following statements regarding choroidal folds is false?
- ☐ Choroidal folds are a rare clinical sign that presents either unilaterally or bilaterally.
- ☐ Choroidal folds are characterized by alternating light and dark bands found in the macula.
- ☐ Choroidal folds are always a result of a serious underlying medical condition.
- ☐ Choroidal folds require a complete and proper work up to determine their etiology.

2 Which condition is not considered to be one of the top 3 etiologies of choroidal folds presenting bilaterally?
- ☐ diopathic
- ☐ hyperopia
- ☐ glaucoma
- ☐ macular degeneration

3 Which condition is not considered to be one of the top 3 etiologies of choroidal folds presenting unilaterally?
- ☐ Ocular Tumors
- ☐ Posterior scleritis
- ☐ Hypotony
- ☐ Uveitis

4 Which of the following statements regarding choroidal folds is true?

- ☐ Choroidal folds are only oriented horizontally,
- ☐ Choroidal folds do not vary in length and usually extend well beyond the beyond the equator.
- ☐ The dark-colored striae of choroidal folds correspond to ridges and light-colored striae correspond to troughs.
- ☐ Choroidal folds are undulations of the: inner choroid, Bruch's membrane, and the RPE

5 Which statement about retinal folds is incorrect?

- ☐ Retinal folds are either congenital or secondary folds
- ☐ Retinal folds are often associated with an abnormal vitreous or vitreal-retinal interface.
- ☐ Retinal folds tend to be indistinguishable from choroidal folds.
- ☐ Retinal folds do not appear on fluorescein angiography.

6 Which of the following is not required inorder to determine the etiology of a choroidal fold?

- ☐ A description of the patient's chief complaint.
- ☐ A detailed description of the physical appearance and orientation of the folds.
- ☐ A detailed patient work up and case history.
- ☐ A review of the abnormal scan and imaging results

7 Which statement regarding where choroidal folds occur is incorrect?

- ☐ Choroidal folds can occur in the macula
- ☐ Choroidal folds can occur around the optic nerve
- ☐ Choroidal folds can occur in the arcades
- ☐ Choroidal folds can occur in multiple sites across the retina at the same time

8 When patients present with a new onset of choroidal folds which condition should not be considered as being a diagnostic differential?

- ☐ Orbital tumours
- ☐ Elevated Intraocular Pressure
- ☐ Grave's disease
- ☐ Orbital myositis

9 With the onset of new choroidal folds which sign is not a considered to be an important concern for an intraorbital pathological involvement?

- ☐ Presence of an afferent pupillary defect
- ☐ An extraocular muscle restriction
- ☐ An elevation in intraocular pressure
- ☐ Changes to visual acuity

10 Which condition will not result in a choroidal fold secondary to ocular hypotony?

- ☐ Narrow angle glaucoma
- ☐ Overfiltration after trabeculectomy
- ☐ Ocular surgery
- ☐ Ocular trauma including penetration of the globe.

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