Case of Idiopathic Chorioretinal Folds

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Abstract

Chorioretinal folds are well documented but uncommonly seen clinically. In the advent of new investigation methods, i.e. Optical Coherence Tomography and fundus angiography, improvements in the understanding of the pathogenesis of chorioretinal folds are seen. Prior to these, >85% of cases were diagnosed as Idiopathic. However, with current understanding, it is postulated only 15% are truly idiopathic and new diagnostic algorithms were proposed.

Case Presentation

The patient is a 76 year old diabetic female, who has been undergoing routine diabetic retinopathy screening for the past 5 years. Her blood sugar levels remained well controlled and there were no ocular abnormalities detected throughout this period. She also has hypertension and dyslipidemia. She did not have any visual complaints and denied having recent change in her spectacles prescription which would suggest a hyperopic shift. There was no history of ocular trauma. She did not have complaints of ocular pain or frequent headaches. She did undergo two uneventful phacoemulsifications and intraocular implantation for both eyes 4 years ago.

Upon examination, her unaided visual acuity was right eye (OD) 6/9 and left eye (OS) 6/18.75 9+0.501.0. The intraocular pressures were 14 mmHg bilaterally. The anterior segment was normal bilaterally with stable posterior chamber intraocular lenses. There was no proptosis or ptosis. The extraocular muscles movements were full in all directions of gaze. There were no eye signs to suggest thyroid eye disease. Fundus examination of both eyes (Figure 1a, 1b) revealed obliquely placed chorioretinal folds, primarily located at the supero-temporal and infero-temporal aspect of the arcades. The retina was normal with no diabetic retinopathy changes. The optic discs were pink with a cup disc ratio of 0.4 on the right, and 0.3 on the left. There was no disc swelling or hyperemia. The maculae were normal with no abnormalities seen. There were no drusen, retinal hemorrhages, choroidal effusions or signs to suggest uveitis.

Investigations

B-scan ultrasonography revealed a normal B-scan with complete posterior vitreous detachment bilaterally. There was no thickened posterior sclera, fluid beneath the sub tenon’s space or choroidal effusions detected. The retina was flat. No retro-bulbar mass was seen during this scan (Figure 2). The axial lengths were OD 22.37 mm and OS 22.39 mm. Optical Coherence Tomography (OCT) revealed no subretinal fluid or retinal pigment epithelial detachment to suggest age-related macular degeneration. However, full thickness choroidal folds were seen (Figure 3). There were no signs of choroidal neovascularization seen in the patient. Computed Tomography (CT) of the brain revealed no abnormalities.

Discussion

The choroid constitutes the vascular layer of the eye and supplies blood to the outer 1/3rd of the retina. As it is difficult to observe the choroid clinically, various imaging studies have been described for the diagnosis of choroidal folds like color fundus photography, fluorescein and Indocyanine Green (ICG) angiography, auto fluorescence, Optical Coherence Tomography (OCT) and ultrasonography [1]. The Pathogenesis of choroidal folds are postulated to be caused by conditions that thicken or shortens the sclera, choroidal thickening. This may be caused by external compression, conditions causing hypotony, or increase in intracranial pressure. The causes of choroidal folds can be broadly divided into ocular and systemic causes [2]. However, it remains a diagnosis of exclusion as many authors have concluded that idiopathic choroidal folds in hyperopic individuals only represent 15% to 17% of the cases. Iatrogenic caused hypotony have been reported to cause choroidal folds [3]. Patients have underwent bilateral cataract extraction;
Figure 1a (OD) and 1b (OS): Color fundus photo showing chorioretinal folds exhibited by alternating light and dark circumferential lines. White arrows indicating light lines whilst dark arrows indicating dark lines.

Figure 2: (OD): Optical Coherence Tomography (OCT) through the folds. Choroidal folds marked with stars.

Figure 3: (OD) Indocyanine green & fluorescein fundus angiography. Figure 3a reveals a normal fundus angiography at the posterior pole. No leakage or pooling seen.
however, they were not immediately seen post-operatively. Choroidal folds have been seen among choroidal neovascularisation, however, these folds present as radial choroidal folds [4]. Common systemic causes of choroidal fold causes compression of the globe posteriorly. Examples of systemic causes are orbital tumors, idiopathic intracranial hypertension, fronto-medial mucoceles, meningioma. Advance thyroid eye disease causing severe inflammation and proptosis have been reported to cause posterior compression of the globe [5]. Posterior scleritis secondary to inflammatory disease, i.e. Rheumatoid arthritis, Vogt-Koyanagi-Harada disease, systemic lupus erythematosus, have been reported to cause posterior pole mass and consequently causing choroidal folds [3]. These conditions were not associated with the patient. Choroidal folds have also been reported among astronauts after prolonged space flight. It is suggested that microgravity condition have caused cephalic fluids shift, akin to the pathogenesis of Idiopathic Intracranial Hypertension [6]. Hyperopic shift was also seen among these astronauts [6]. Investigations were carried out according to algorithm to exclude the causes mentioned above [3]. Patient did not show any signs and symptoms of inflammatory disease. B-scan ultrasonography was performed to rule out posterior scleritis and posterior pole mass. Fluorescein fundus angiography and indocyanine green angiography was performed to rule out choroidal neovascularisation. CT Brain was performed to exclude any insidious masses that have not presented clinically. After the exclusion of the major causes of the choroidal folds, patient is diagnosed with bilateral idiopathic choroidal folds. She is also under follow-up annually for her diabetic retinopathy screening.

References