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Case Report Open Access

Pattern Dystrophy Associated with Bilateral Submacular Fluid; a Multimodal Imaging Study

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Abstract

Purpose: To report multimodal imaging characteristics of a patient with pattern macular dystrophy with bilateral submacular fluid.

Methods: A 47 year-old man was referred with gradual decrease of visual acuity in both eyes. He had history of bilateral injection of intravitreal bevacizumab with the diagnosis of central serous chorioretinopathy without any clinical effect. Funduscopic and optical coherence tomography examinations revealed bilateral subretinal fluid in the macular area.

Results: Fundus autofluorescence, fluorescein angiography and indocyanin green angiography revealed characteristics of butterlfly-shaped macular dystrophy with no leakage in the macular area. Electro-oculography was abnormal.

Conclusion: Pattern macular dystrophy may be presented with bilateral subretinal fluid in macula.

Keywords: Pattern dystrophy; Central serous chorioretinopathy; Macular dystrophy

Introduction

Pattern dystrophies are a group of inherited macular disorders that include adult-onset foveomacular vitelliform dystrophy, butterfly-shaped macular dystrophy, reticular dystrophy of the retinal pigment epithelium (RPE), and fundus pulverulentus [1]. They are characterized by the deposition of yellow-orange-grayish materials above the RPE [1].

Pattern macular dystrophy is usually diagnosed in middle aged asymptomatic patients whose visual acuity remains normal or near normal for most of their lives [1,2]. The diagnosis is mainly clinical, based on the classic "pattern" appearance of the fundus. Multimodal imaging including specific autofluorescence and angiographic characteristics of the macula is used to confirm the diagnosis and evaluate for complications. Generally, dark adaptation, color vision, visual field, and electroretinogram (ERG) are normal, and the electro-oculogram (EOG) may be normal or subnormal [2]. Hereby, we report a patient with typical characteristics of pattern macular dystrophy presenting with bilateral subretinal fluid.

Case Report

A 47 year-old man referred to our retina clinic with the complaint of bilateral gradually decreased visual acuity since 2 years ago. He had a history of bilateral intravitreal injection of bevacizumab with the diagnosis of chronic central serous chorioretinopathy (CSC) six months before current presentation. Bilateral subretinal hypo-reflective spaces in the macula was evident on pre-bevacizumab injection optical coherence tomography (OCT) with no apparent change after injections. Past medical history and family history were unremarkable.

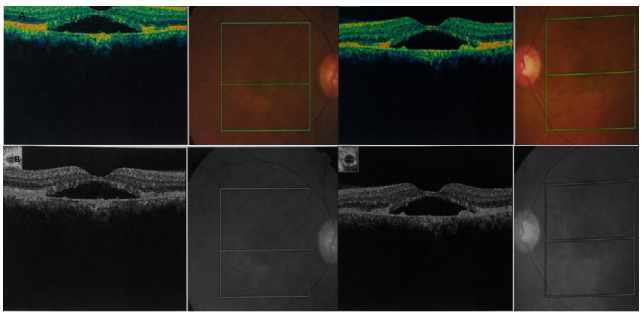

Figure 1: Optical coherence tomography shows bilateral submacular fluid.

Best-corrected visual acuity was 20/40 in right eye and 20/63 in left eye. Slit lamp examination was normal. Dilated fundus examination revealed subretinal fluid associated with RPE changes in the macula in both eyes.

Spectral domain OCT confirmed the presence of bilateral subretinal hypo-reflectivity in macula with mild RPE irregularities (Figure 1). Autofluorescence (AF, Figure 2), fluorescein angiography (FA) and indocyanin green angiography (ICG) images (Figure 3) displayed bilateral symmetric butterfly-shaped pattern of macular lesions. No obvious leakage was found in FA and ICG images.

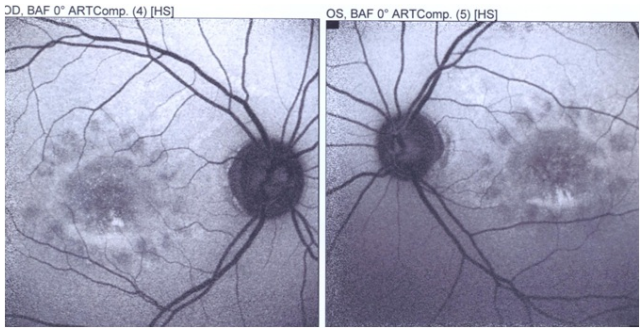

Figure 2: Fundus autofluorescence shows bilateral symmetrical hypo-autofluorescence in the macular area. Some hyper-autofluorescence is evident inferior to the fovea.

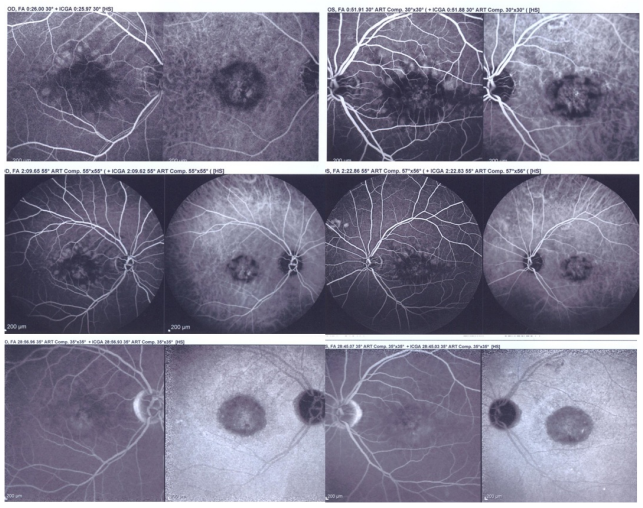

Figure 3: Combined fluorescein and indocyanin green angiography shows pattern-shaped areas of staining. No leakage is seen.

ERG was normal, however, Arden ratio was abnormal in both eyes (light peak/dark ratio was 1.24 in the right eye and 1.28 in the left eye).

Discussion

Patients with chronic CSC usually demonstrate pigmentary changes in the area of retinal detachment which is always associated with focal or diffuse leakage in FA and ICG [3]. In our patient, bilateral symmetric macular lesions with no apparent leakage in FA and ICG suggests an alternative diagnosis. Abnormal EOG was compatible with the diagnosis of pattern macular dystrophy.

Deutman et al, first described butterfly-shaped macular dystrophy; a peculiar bilateral butterfly-shaped pigmentation in the macular region of a white family, the pathology at the level of the RPE [2]. Different mutations of a photoreceptor disc membrane glycoprotein gene, encoded by retinal degeneration slow/peripherin gene located on human chromosome 6p2l, have been reported to be associated with this disease. Macular lesion appears as an area of depigmentation outlined a spoke-like pigment pattern which is similar to the shape of a butterfly. In autofluorescence imaging, butterfly-shaped macular dystrophy has a unique feature of hypo- fluorescence and hyper-fluorescence pattern [4]. FA shows early hyper-fluorescence outlined triradiate central, hypo-fluorescent, butterfly-shaped lesion, and helps to differentiate butterfly-shaped macular dystrophy from other pattern dystrophies [2]. In OCT examination, hyper- reflectivity between the RPE and Bruch's complex and disruption of the inner and outer segment junction of photoreceptors and external limiting membrane, and abnormal hyper-reflectivity originating from the RPE expansion towards the outer nuclear layer, have been reported [1]. Characteristic radiating pattern of the macular lesion in AF and FA is compatible with the diagnosis of the butterfly-shaped macular dystrophy. The origin of the subretinal fluid in our patient is unclear. Previous studies have shown that the type of the pattern dystrophy may vary in two eyes of a patient. Moreover, one type of pattern dystrophy may change to another type in the same eye [5]. Pinós et al., reported CSC occurrence in a young man with adult onset foveomacular vitelliform dystrophy [6]. Lee et al., reported the presence of subretinal fluid in 2 patients with adult vitelliform dystrophy misdiagnosed as chronic CSC. The subretinal fluid was proposed to be the result of the absorbed vitelliform lesion [7]. It is possible that our patient have had adult onset vitelliform dystrophy that later evolved to butterfly-shaped macular dystrophy. As there was no vascualr abnormality; and no active retinal or choroidal leakage evident in imaging, a response to intravitreal bevacizumab is not expected.

Butterlfly-shaped macular dystrophy rarely may be presented with bilateral subretinal fluid in macula which may resemble central serous chorioretinopathy. The latter has treatment while the first has not. Subretinal fluid in butterfly-shaped macular dystrophy in absence of CNV has not been reported, yet.

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