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Case Report

Posterior Polar Hemispheric Choroidal Dystrophy

Introduction

The term "posterior polar hemispheric choroidal dystrophy" was first used by Yannuzzi to describe the annular, hemispheric loss of retinal pigment epithelium and choriocapillaris [1]. Since then, there have been no further reports on this rare condition.

Case Report

A 71-year-old Japanese woman presented with gradual deterioration of the visual fields in both eyes for the past five years. She did not have a history of previous ocular or systemic illnesses. There was no family history of a similar disease. Best corrected visual acuity was 0.8 and 0.9 in her right and left eyes, respectively. The fundi showed bilateral symmetrical annular bands of depigmentation beginning at the lower part of the optic disc that passed along the course of the lower temporal vessels leading to increased visibility of the choroidal blood vessels (Figure 1). Bone spicule pigmentation of the retina was absent, and there was no evidence of narrowing of the retinal blood vessel. Fundus autofluorescence imaging (Heidelberg Retina Angiograph 2; Heidelberg Engineering, Heidelberg, Germany) showed clearly defined hypofluorescent areas that corresponded to the aforementioned lesions (Figure 2). Automated perimetry (Humphrey Field Analyzer model 750; Humphrey Instruments, Inc, Dublin, California) revealed an incomplete ring scotoma that corresponded to the above lesions (Figure 3).

Discussion

Posterior polar hemispheric choroidal dystrophy is characterized by annular, hemispheric loss of retinal pigment epithelium and choriocapillaris. There is visual field loss that corresponds to the choroidal atrophy. This rare condition was most readily detected with the fundus autofluorescence images.

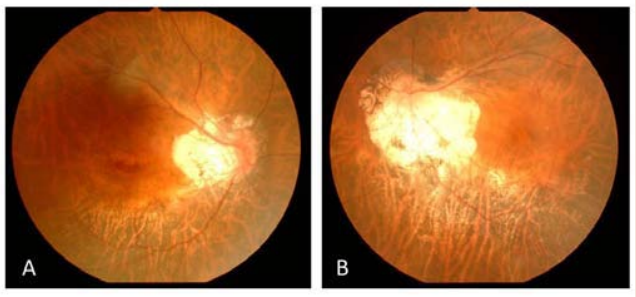

Figure 1: Right (A) and left (B) fundus photopraphs showing bilateral symmetrical annular bands of depigmentation beginning at the lower part of the optic disc that pass along the course of the lower temporal vessels.

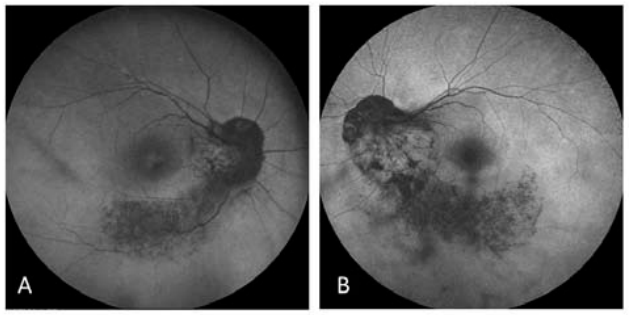

Figure 2: Right (A) and left (B) fundus autofluorescence images showing bilateral symmetrical hypofluorescent areas.

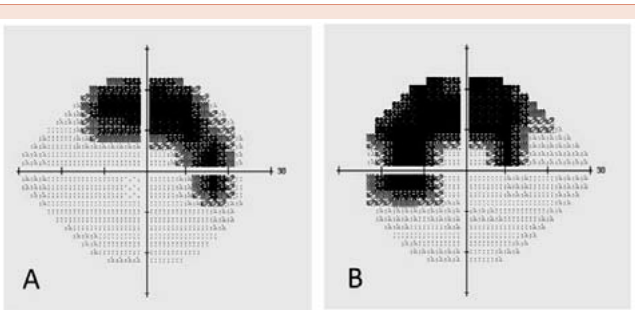

Figure 3: Right (A) and left (B) visual fields showing anincomplete ring scotoma as indicated by the black shaded regions.

The case presented herein has many findings that are consistent with those described by Yannuzzi as posterior polar hemispheric choroidal dystrophy [1].

The differential diagnosis of this condition included pericentral retinitis pigmentosa[2], pericentralpigmentary retinopathy [3], pericentral retinal dystrophy [4], circinatechoroidal sclerosis [5], annular choroidal sclerosis [6], choroidal sclerosis [7], and central areolar choroidal dystrophy [8].

Patients with pericentral retinitis pigmentosa may present with slight central or night vision difficulty and occasional awareness of peripheral vision loss[2]. Examination of the fundus typically reveals midperipheral retinal pigment epithelial attenuation or bone spicule

pigmentation, a normal appearing disc and macula, minimal retinal arteriolar narrowing, and a normal appearance to the far periphery. An annular scotoma, corresponding to the region of retinal pigment epithelium attenuation or bone spicule pigmentation, is a common finding. However, in this present case, she did not complaine night blindness. In addition, there was no evidenceof bone spicule pigmentation and retinal arteriolar narrowing.

Choroidal sclerosis is a term used to describe a group of genetically heterogeneous retinal dystrophies that share a common funduscopic phenotype of scalloped, well-circumscribed, and confluent chorioretinal atrophic lesions that extend beyond the macula [7]. The term choroidal sclerosis was initially coined because of the sclerotic appearance of the choroidal blood vessels, which frequently appear white or light yellow. However, histologic studies have shown the term to be a misnomer because the principal abnormality is the loss of choriocapillaris, retinal pigment epithelium, and the outer retina in the absence of abnormal sclerotic changes in the choroidal vasculature. Circinatechoroidal sclerosis and annular choroidal sclerosis are rare forms of choroidal sclerosis that are limited to a circular or annular area around the macula. Interestingly, posterior polar hemispheric discoloration showed in this present case.

Central areolar choroidal dystrophy is a hereditary retinal disorder that affects the macula, resulting in progressive and usually profound visual loss [8]. The hallmark feature of the disorder is a well-defined atrophy of the retinal pigment epithelium and the choriocapillaris involved the fovea. In contrast, foveal sparing showed in this present case.

My findings were based on a single case of posterior polar hemispheric choroidal dystrophy, and further studies with additional cases are necessary.

Conflict of Interest

The author has no financial or proprietary interests related to this paper.

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