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Case Reports

Concurrence of retinitis pigmentosa and central serous retinopathy

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Summary

We describe a patient with concurrent retinitis pigmentosa and central serous chorioretinopathy. Both conditions involve dysfunction of the retinal pigment epithelium and evince distinct clinical, angiographic, and electrophysiologic features. Potential pathophysiologic connections are explored.

Introduction

Central serous chorioretinopathy (CSR) is an idiopathic chorioretinal condition causing loss of central vision due to the accumulation of fluid in the subretinal pigment epithelium (RPE) space and subsequently in the subretinal space, creating a well-circumscribed serous retinal elevation.[1] The incidence of CSR has been estimated to be 6 per 100,000. It is not typically associated with other ocular diseases. The term *retinitis pigmentosa* (RP) describes a group of hereditary retinal dystrophies with characteristic retinal and electroretinogram (ERG) findings. Its prevalence is approximately 1 in 4000, affecting over a million people worldwide. It is characterized by degeneration of photoreceptor cells, predominantly rods, with secondary degeneration of the inner nuclear layer and retinal pigment epithelium. Fundus examination characteristically reveals bone spicule pigmentary changes in the midperiphery, attenuation of retinal vessels, waxy pallor of the optic disc, and a variable amount of pigmented cells in the vitreous.[2,3] These two entities have rarely been reported to co-occur. We report a patient with previously undiagnosed RP presenting with acute CSR.

Case Report

A 49-year-old African Canadian man presented with a one-week history of blurred vision in the left eye. He initially denied having any other ocular complaints, but on further questioning he admitted to experiencing poor night vision for approximately a year. The patient was taking glyburide and metformin for diabetes mellitus of 8 years' duration. He was also on amlodipine and candesartan for systemic hypertension. He had no previous personal or family history of eye disease. He was under considerable emotional stress, having recently lost his job.

On examination, visual acuity was 20/20 in the right and 20/200 in the left eye. The pupils were reactive, and no afferent pupillary defect was noted. Visual fields by confrontation were bilaterally constricted. Slit-lamp examination revealed moderate nuclear sclerosis bilaterally. No vitritis was noted. Intraocular pressure was normal in both eyes. Dilated fundus examination revealed bone spicule pigmentation in the midperiphery of both eyes (Figure 1). The left eye had a large accumulation of subretinal serous fluid in the macular region (Figure 1A-B). Fluorescein angiography demonstrated a "smokestack" pattern of hyperfluorescence (Figure 1C-D). There was no sign of diabetic retinopathy in either eye.

Goldman perimetry demonstrated bilateral ring scotomas. Full-field ERG performed after complete resolution of the CSR event was nonrecordable above noise in both

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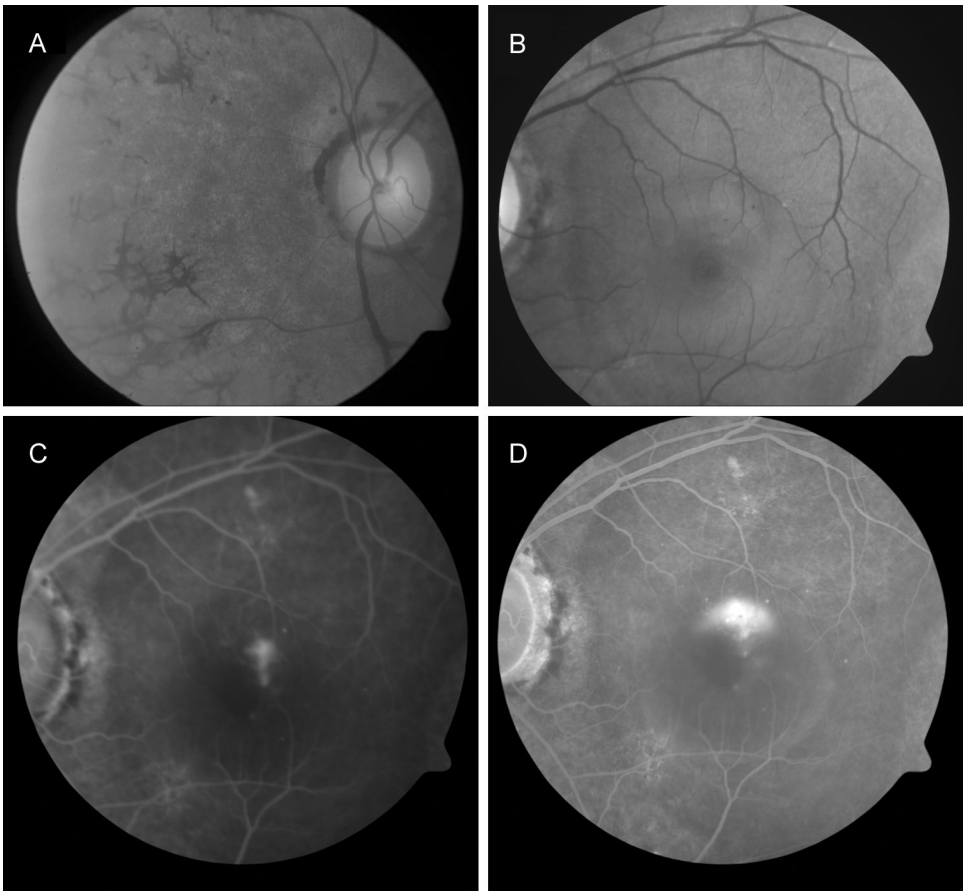

Figure 1. Fundus photographs and fluorescein angiography of the left eye. A, Fundus photograph of left eye demonstrating peripapillary bone spicule pigmentation. B, Red-free photograph of left eye showing serous elevation of the neurosensory retina centered in the macular region. C-D, Fluorescein angiography at 1 minute and 6 minutes showing a "smokestack" pattern of hyperfluorescence within the area of the serous elevation.

eyes, as typical of RP. There was no significant difference in the ERG recordings between the eye with CSR and the contralateral eye.

No treatment was recommended. Two months later, the visual acuity had improved spontaneously to 20/40 in the affected left eye, with complete disappearance of the subretinal fluid. No recurrence has developed since the first attack of CSR two years ago.

Discussion

Our patient demonstrated features typical of both CSR and RP. His epidemiologic profile, symptoms, and fundus examination were typical of both conditions. Fluorescein angiography confirmed the diagnosis of acute CSR; the patient's elicited history of nyctalopia and bone-spicule pigmentation in the mid-periphery on fundus examination support the diagnosis of RP. These findings, in association with the extinguished ERG were highly suggestive of RP. Other diagnoses, such as vitamin A deficiency, metallosis, and retinal arterial occlusion were not compatible with the clinical history or fundus appearance. Although macular pathology can be found in up to 60% of RP patients,[3] most commonly it is atrophy of the neurosensory retina and/or the RPE. Other less common macular changes include macular cysts, holes, and/or cystoid macular edema.[2,3]

To our knowledge, CSR in association with RP has been reported only four times previously.[4–7] Meunier et al[6] presented images of a 43-year-old man with autosomal dominant, ERG-confirmed RP who presented with CSR; details of the case are not provided. Yamaguchi et al[7] reported a patient with pigmentary retinopathy who presented with CSR. The authors felt that the two entities were independent. Lewis described a case of a 30-year-old white woman who was found to have fundus, visual

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field, and ERG features consistent with RP.[5] At the time of original presentation, she had evidence of multiple serous RPE detachments. Seven years following the initial evaluation she developed a persistent serous detachment in the right eye that required laser therapy. Lewis speculated that RP may have contributed to the severity of the CSR; however, she concluded that the concurrence may have simply been a rare coincidence. Dorenboim et al described a case in which funduscopic examination, Humphrey visual field testing, fluorescein angiography, and ERG were all consistent with a dual diagnosis of CSR and RP.[4] Their patient required laser photocoagulation on two occasions. They speculated that the CSR in RP could be a result of atrophic changes in the RPE as a part of the RP disease process and further suggested that since postresolution CSR fundal changes can mimic RP changes in appearance, the incidence of CSR in RP may be underestimated.

The association in our case between RP and a first episode of CSR may be coincidental; however, elements of choroidal pathophysiology in both CSR and RP may explain the concurrence. In general, current theories tend to consider three aspects of RPE physiology in the development of CSR: (1) the balance of hydrostatic forces, (2) the continuity of the RPE, and (3) the pumping function of the RPE. To the first, both Piccolino and Spaide et al. suggest that increased choriocapillary vascular pressure, whether due to circulating catecholemines or hypoperfusion downstream, lead to increased hydrostatic pressure below the RPE, thus favoring extraversion of fluid into the sub-RPE and, potentially, subretinal spaces.[8,9] Others continue to explain that in order for fluid to accumulate there must also be either a break in the continuity of the RPE,[1,8–10] diffuse RPE pumping dysfunction,[11] or both.[8,10]

Histopathologic changes seen in RP may contribute to these processes and in turn lead to the development of CSR. Migration of RPE cells from Bruch's membrane to the inner retinal layers and perivascular areas in RP may lead to weakening of the retinal pigment epithelium and the formation of breaks in the RPE.[12] This may cause leakage into the sub-retinal space or interfere with the ability of the RPE to maintain negative pressure in this potential space. Additionally, reduced choriocapillary permeability found in RP could lead to increased upstream choriocapillary arterial pressure that may be a critical process in the development of CSR.[9,10]

The nature of any physiologic relationship between CSR and RP is speculative at this point and would require greater understanding of pathologic mechanisms that has thus far eluded researchers. Nevertheless, plausible pathophysiologic relationships between these disorders exist. It would be prudent to consider CSR as a possible etiology in a patient with RP and acute visual loss, and vice versa. An examination of the retinal periphery in patients with CSR may reveal early changes of RP, a finding that may have treatment implications.

Literature Search

We searched MEDLINE (January 1948 to September 2012) combining the following terms: *retinitis pimentosa* AND *central serous chorioretinopathy* OR *central serous retinopathy* OR *choroid diseases*. Survey of the results revealed three related references. Citation lists from each of these were then cross-referenced for appropriate reports. Finally, the ISI Web of Science database system was used to identify all articles citing the reports of interest; these results were surveyed for any applicable additions.

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