American Academy of Optometry: Case Report 5

Clinical Findings and Management of Idiopathic Central Serous Chorioretinopathy

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Abstract

Idiopathic central serous chorioretinopathy (ICSC), a disease affecting the central retina and choroid, presents a straightforward diagnostic profile based on patient demographics, symptoms, and clinical findings. Unfortunately, little is understood about the etiology of ICSC and treatment intervention can have devastating visual impact. The natural course of ICSC ends in favorable visual outcome most of the time. It is important for clinicians to appreciate the clinical findings, course of the disease, and weigh treatment options before pursuing intervention. This case report reviews the management of patients with ICSC, discussing clinical findings and treatment options as they relate to the case presented.

Key words*: Idiopathic central serous chorioretinopathy (ICSC), metamorphopsia, fluorescein angiography*

Introduction

Idiopathic central serous chorioretinopathy, also known as central serous retinopathy, central serous pigment epitheliopathy, and central serous retinitis, is a disease that appears to impair choroidal circulation leading to central retinal pigment epithelium (RPE) and serous detachment of the retina[1-4]. The disease typically presents in isolated unilateral episodes, but is considered a bilaterally disease of the choroid. There does not appear to be any clear predisposing factors that lead to the development of ICSC. It predominantly affects white males between the ages of twenty and fifty with a mean age of onset at thirty. No cases have been reported in individuals younger than twenty[1,5]. It affects males ten times more often then females and is associated with a type-A personality, patients with a competitive or aggressive disposition, or who may be under extreme physical or emotional stress. ICSC presentation has been associated with the use of vasoconstrictive agents such as epinephrine, endogenous hypercortisolism, and systemic corticosteroid use[1,6]. Several entities believed to elicit choroidal vascular dysfunction have been known to produce ocular findings that mirror ICSC. These include hypertension, pregnancy, dialysis, organ transplantation, and systemic lupus erythematosus[1]. An association with migraines has also been documented. The role of genetics is unclear. The vast majority of patients present without a family history of the disease; however, two families with an apparent inheritance pattern have been identified[7].

The sudden onset of unilateral metamorphopsia, blurred central vision, and less frequently abnormal color vision are classic symptoms of ICSC[1,3,5,8]. With extrafoveal presentations of ICSC discovered during routine examination, patients may be asymptomatic.

Case Report

Patient #5, an anxious 23-year-old white male, presented to our clinic on 25 January 1999 with complaints of reduced vision in his left eye at all distances for the last week. He reported awaking to what seemed like 'eye strain' on the first day of presentation and his vision had been steadily declining OS. At today's presentation he reported that it was as if he were "looking through a bubble" in the center of his vision. The patient's last eye examination was December 1997 and his medical record (including all prior eye examinations) has been lost. The patient's only past ocular history was occasional spectacle wear for distance vision. The patient's medical and family history was negative, and he denied taking any medications or having allergies to the same. The patient reports no history of migraines and his Blood Pressure measured 103/73. He was oriented to time, place and person.

His uncorrected visual acuity was 20/20 at distance and near OD, and 20/70 at distance, $20/30^{-2}$ at near OS. Best-corrected visual acuity was unchanged OD and OS, with a manifest refraction of +1.00 - 1.50 x 105 OD and +0.50 - 1.75 x 033 OS. Pinhole acuity at distance was 20/60 OS. Amsler grid testing was normal OD, however, a large area of central metamorphopsia was documented OS. Color vision testing with pseudoisochromatic plates was normal OU. Pupils were equally round and reactive to light, no afferent pupil defect was noted OU. Confrontation fields were full to finger count OU. Extraocular muscles were unrestricted in all gazes, and cover test demonstrated orthophoria at distance, and 2^{Δ} exophoria at near. Intraocular pressure was 6 mmHg OD and OS with non-contact tonometry. Anterior segment evaluation by slit lamp examination revealed a quiet bulbar and palpebral conjunctiva OU; an even tear film with tear

break up time of 13 seconds/OU; clear lashes OU; intact and clear corneas OU; irises were brown OU; anterior chamber appeared clear without cells or flare and the estimate of the anterior chamber angles was 4/4 by VonHerrick. The patient was dilated using one drop Tetracaine, one drop 1% Mydriacyl and one drop 2.5% Phenylephrine OU. Once the patient was fully dilated, an evaluation of the posterior segment by slit lamp with 78D lens and by Binocular Indirect Opthalmoloscope revealed a few focal mid-peripheral cortical opacities of the lens consistent with congenital cataracts. The central media of the lenses in both eyes were optically clear. Fundus assessment revealed normal optic nerves with a cup-to-disc ratio of .4/.4 OU. The neuroretinal rims were healthy and intact. Retinal vessels appeared normal with an arterial-venous ratio of 2/3 noted OU. Both eyes presented with abnormal macular findings. The macula of the right eye had a focal area approximately 1-disc diameter (DD) temporal to the fovea that appeared as a region of relative retinal pigment epithelial hypopigmentation with questionable elevation. The macula of the left eye had a clearly defined central serous retinal detachment approximately 1DD in size centered over the fovea. No retinal pigment epithelial detachment was noted. The dome of the detachment was clear and the serous fluid within the detachment appeared optically clear. The peripheral retina was flat and intact with no pathology noted OU.

The differential diagnoses considered in this case include:

Age-related macular degeneration (ARMD)
Optic pit
Macular detachment due to a rhegmatogenous retinal detachment
Choroidal tumor
Pigment epithelial detachment
Idiopathic central serous chorioretinopathy (ICSC)

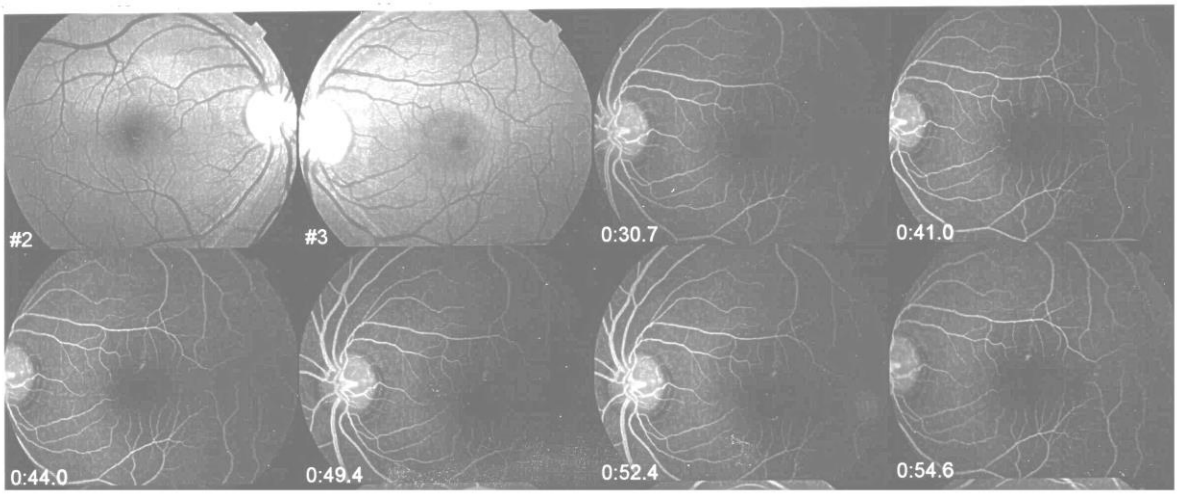
- ARMD generally appears in patients over 50 years of age. It is often bilateral and accompanied by drusen, pigment epithelial alternations and, in some instances, a chorodial neovascular membrane.
- An optic pit causes a small defect in the nerve tissue and a serous retinal detachment that may be contiguous with the optic disc.
- A macular detachment due to rhegmatogenous retinal detachment is accompanied by a hole or tear in the retina.
- A choroidal tumor or mass is generally evident during a binocular indirect ophthalmoscopy examination.
- The margins of a pigmented epithelial detachment (PED) would be distinct, however, a PED may accompany ARMD or ICSC.
- ICSC generally appears as a localized detachment of the sensory retina from the underlying pigment epithelium by a clear serous fluid in the macular area. The margins are sloping and merge gradually into the attached retina.

There was no evidence of drusen or defects in the nerve tissue of the optic disc. There were no holes or tears in the retina, no tumors in the choroids, and no distinct margins to the defects. Therefore, the patient was diagnosed with idiopathic central serious chorioretinopathy (ICSC) OS and tentatively diagnosed with resolved ICSC with secondary RPE changes OD. He was also diagnosed with congenital cataracts OU. The patient was counseled on the nature of ICSC and instructed to return for a fluorescein angiography to be conducted by a clinic technician later that afternoon to confirm the diagnosis. The fluorescein angiography would also reveal a

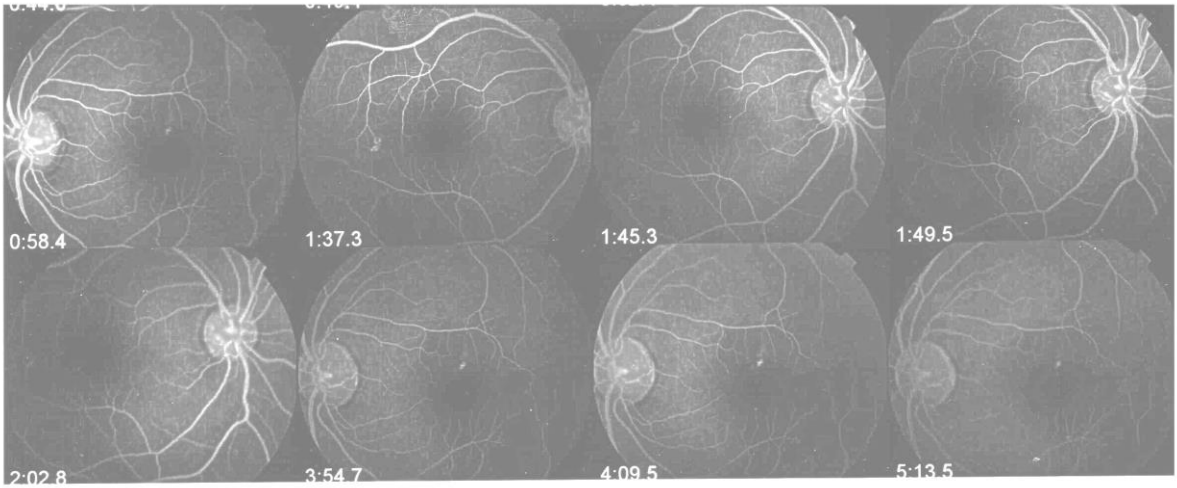
choroidal neovascular membrane and would also be needed if laser treatment were to be instituted. The patient was advised to follow up with me to review the results and discuss treatment the next day.

Follow up # 1

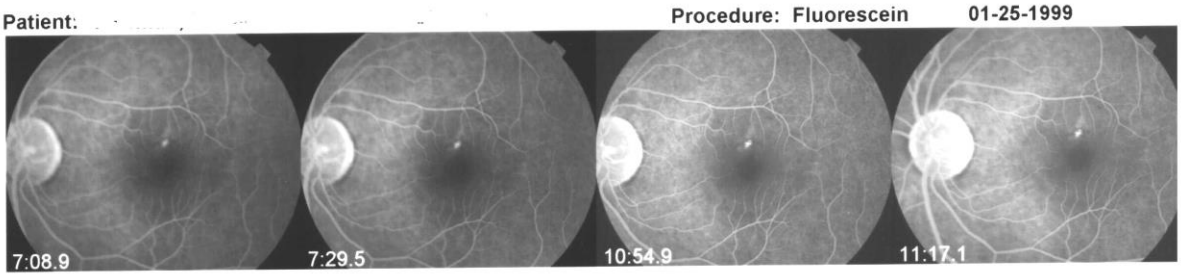
The patient returned on 26 January 1999 to review the fluorescein angiography OU. His visual acuity remained stable with uncorrected acuities of 20/20 OD and 20/70 OS. His results are presented in the next three series.



Fluorescein angiography of the right eye (continued in series below) revealed results consistent with retinal pigment epithelial atrophy and hypopigmentation. The lesion temporal to the fovea demonstrates early phase hyperfluorescence accompanying the choroidal flush and arterial phase of the angiography. As the fluorescein abates, with transition to the venous stage of the angiography, so do the corresponding levels of fluorescence from the lesion. By the end of the arterial phase of the angiography at 2 minutes the lesion is barely visible. The fluorescein angiography of the left eye is notable different. The dome of serous neurosensory retinal detachment is easily visualized by the reflections from the internal limiting lamina (above). A focal retinal pigment epithelial lesion is noted approximately ½ DD superior to the fovea and begins to hyperfluoresce with the onset of the choroidal and arterial phases of the angiography.



The lesion continues to hyperfluoresce through the venous phase and remains hyperfluorescent even at eleven minutes. Also noted is a subtle superior extension of hyperfluorescence just superior to the initially well-defined lesion through out the course of the imaging.



Both patterns confirm the initial diagnosis of focal RPE atrophy (likely secondary to an earlier bought of ICSC) OD, and active retinal pigment epithelial detachment with overlying serous sensory detachment consistent with ICSC OS.

The patient was counseled that little was known about the cause of ICSC and that it was often related to a type-A personality. The patient confirmed that this personality trait did in fact apply to him. The patient was counseled about the use of laser treatment in treating ICSC versus simply monitoring for resolution over time and that the odds were very good that his vision would return to normal without intervention. The patient was instructed on the use of the Amsler Grid and verbalized his understanding clearly and concisely. The patient was given an Amsler grid to monitor daily at home for changes in his vision and was instructed to return in six weeks for follow up, or sooner if any changes were noted.

Follow up # 2

The patient returned for follow up on 10 March 1999. He denied any changes in his medical history and denied taking any medications. He reported some improvement in his left eye, but his vision remained hazy OS. Entering acuities were 20/20 OD and $20/40^{-2}$ OS, no improvement with pinhole. Amsler grid testing continued to show a central area of metamorphopsia OS. Color vision was normal and pupils were equally round and reactive to light with no afferent

pupil defect noted OU. One drop of Fluress was instilled to measure the intraocular pressures by applanation tonometry. The intraocular pressures were 8 mmHg OD, and 10mmHg OS. Slit lamp findings remained unchanged OU. Dilated fundus assessment OS revealed a much smaller area of involved retina, with an appearance consistent with retinal edema and thickening as opposed to frank serous detachment.

The patient was counseled on the good recovery he was making and to remain patient as the retinal swelling continued to resolve. He was instructed to return again in six weeks for follow up and to continue to monitor vision at home with Amsler grid testing.

Follow up # 3

The patient returned on 16 April 1999 for follow up. Again, the patient denied any change in his medical history. He did report improvement in his vision OS but it 'still didn't seem right'. Visual acuities were 20/20 OD and 20/30 OS, no improvement with pinhole. Manifest refraction OS was +0.25 – 1.00 x 077 yielding a visual acuity of $20/25^{-2}$. Amsler grid testing demonstrated a focal area of central metamorphopsia OS. Pupils were equally round and reactive to light, no afferent pupil defect was noted OU. Intraocular pressures by non-contact tonometry were 7 mmHg OD, and 8 mmHg OS. Slit lamp findings remained unchanged OU. Dilated fundus assessment OS revealed a focal area of RPE mottling just superior to the fovea and an area of questionable retinal thickening less than ¼ DD in size centered over the fovea.

Again, the patient was counseled on the good progress in visual recovery noted OS. He was counseled to continue with home monitoring and return again in six weeks.

Follow up # 4

The patient returned on 2 June 1999, reporting good vision OU. He denied any change in medical history and reported that his vision now seemed 'normal' OU. Uncorrected visual acuities were 20/20 OD, $20/20^{-1}$ OS corrected to 20/20 with +0.25 – 1.00 x 081. Amsler grid testing was normal OS. Color vision remained normal OU. Pupils were equally round and reactive to light, no afferent pupil defect was noted OU. Intraocular pressures by non-contact tonometry were 6 mmHg OD, and 7 mmHg OS. Slit lamp findings were unchanged OU. Dilated fundus assessment OS revealed only a faint focal area of RPE mottling just superior to the fovea. No retinal thickening was apparent.

The patient was counseled on his good visual recovery. He was reminded about the possibility of recurrence and instructed to continue with home Amsler grid testing on a weekly basis OU. As long as he did not experience any changes in his vision, he was counseled to return in six months for a comprehensive annual examination.

Discussion

The pathogenesis of idiopathic central serous chorioretinopathy remains obscure; however recent studies are shedding light on its vascular origin. Indocyanine green angiography studies suggest that ICSC is a bilateral disease of the choroid that results in reduced perfusion of choroidal capillary lobules[2]. The reduced choroidal perfusion is believed to occur secondary to vasomotor

instability or sympathetic nervous system excitation[4, 5,13]. This lends credence to the psychological stereotype attributed to ICSC patients and demonstrated in LC. In regions of reduced choroidal perfusion, localized ischemia leads to serous fluid leakage from the choroid, creating a detachment and break in the retinal pigment epithelium with secondary serous fluid accumulation between the RPE and neurosensory retina. The propensity for ICSC to affect the central macular region over peripheral retina is both a function of the symptoms produced leading to detection, and the greater level of hemodynamic activity. The demographic profile of patients affected by ICSC has resulted in limited histological studies; however in those conducted, the choroid, RPE, and retinal vasculature appeared normal. The only changes observed were serous RPE and Bruch's membrane detachments as well as cystic degeneration in the outer retinal layers of the detached neural retina[1].

Unilateral, localized serous RPE and neurosensory retinal detachment at the macula produce characteristic clinical findings. Typically visual acuity is moderately reduced in the affected eye and may be correctable to 20/20 with a hyperopic shift in a patient's prior habitual spectacle correction. Depending on the size of the neurosensory retinal detachment, a mild afferent pupillary defect and relative scotoma may be present[9, 10]. Complaints of metamorphopsia and abnormal color vision are readily documented with Amsler grid and color testing[11]. Patients with ICSC may also demonstrate impaired contrast sensitivity, dark adaptation, delayed photostress recovery times, and abnormal multifocal electroretinograms[9,12,13].

The typical ophthalmoscopic appearance of an acute episode of ICSC is that of a transparent dome, or blister, with fluid accumulation between the neurosensory retina and RPE in the posterior pole[1,3,5,8]. The neurosensory detachment is best viewed at the slit lamp with an indirect fundus contact lens and off-axis light source observing separation of the beam and shadows created as the light source traverses the detachment and highlights elevation of the neurosensory retina and retinal vessels. The size and height of the dome will vary and binocular depth cues, such as parallax, color variation, and glistening reflections from the internal limiting membrane must be employed to appreciate the full limits of the neurosensory detachment. The fluid within the neurosensory detachment may be clear or turbid. A focal yellow-gray area surrounded by a faint gray halo, typically less than 1/3 DD, may be observed within the base of the dome and represents the underlying RPE detachment[1,5,8].

Rarely, ICSC will present as a bullous peripheral nonrhegmatogenous retinal detachment located inferiorly, thus confusing diagnosis. The bullous sensory detachment shows shifting subretinal fluid and yellowish-white subretinal exudates[1,8]. Fluorescein and indocyanine green angiography will illuminate the source of leakage and confirm the diagnosis in these cases.

The appearance of chronic or slowly resolving cases of ICSC varies[1,5,8,14]. The fluid within the dome may become more turbid, and focal protein precipitates may form on the posterior neurosensory retina and anterior RPE, appearing as small focal yellow deposits. As the fluid resolves, RPE mottling or clumping may develop as well as RPE hypopigmentation and cyst-like changes within the macula. These RPE changes suggest a chronic episode of ICSC and may serve as evidence of prior episodes. These signs of longevity suggest poorer visual prognosis. Unfortunately, 5% of ICSC patients will go on to manifest a progressive variant of the disease with persistent diffuse retinal pigment epithelium dysfunction and persistent or intermittent subretinal fluid[1]. The area of RPE hypopigmentation observed in the patient's right eye was

consistent with the appearance of a prior episode of ICSC. Given its location 1DD temporal to the fovea, the patient may have remained asymptomatic during that episode.

Angiographic imaging is important to exclude the presence of other sources of neurosensory retinal detachment, particularly choroidal neovascularization. Two angiographic techniques may be employed: fluorescein and indocyanine green angiography. In active cases of ICSC, both techniques will show a focal staining within the base of the neurosensory detachment that stains early and continues to enlarge throughout the series. This represents the RPE detachment. The stain may diffuse through the break in the RPE and begin to stain the neurosensory retinal detachment if the RPE remains compromised. Filling of the neurosensory detachment may take on the appearance of a smoke stack emanating from the RPE break, or that of gradually enlarging circle of hyperfluorescence. It is postulated that the convection currents of the serous exudate within the neurosensory detachment cause the smoke stack appearance. Both techniques will show persistent staining into the late stages. Indocyanine green angiography can help to clarify the diagnosis of ICSC when ruling out the presence of neovascularization if fluorescein angiography is inconclusive[1,15]. Indocyanine green will only show staining at the site of RPE and neurosensory detachment in the case of neovascularization. In cases of ICSC, indocyanine green will show bilateral hyperfluorescence in both affected and unaffected areas of the choroid with peak fluorescence at mid-phase[15]. When indocyanine green angiography is unavailable and initial fluorescein angiography is inconclusive for choroidal neovascularization, a second fluorescein angiography may be repeated several days later at which time ICSC will appear stable or remitting as opposed to the growth seen with neovascularization. The patient showed a classic RPE and 'smoke stack' neurosensory detachment with fluorescein angiography in the left eye that persisted well into the late phases of testing. Fluorescein angiography of the right eye was consistent with an RPE 'window' defect whose hyperfluorescence tracked the choroidal filling and voiding phases.

Optical coherence tomography is another tool that can be used to track the course and resolution of ICSC. This imaging technique can identify the presence of subretinal fluid with high resolution, and the degree of subretinal fluid found correlates well with ophthalmoscopic findings and visual function[16].

Differential diagnosis of ICSC includes any entity that can produce a RPE and or neurosensory nonrhegmatogenous retinal detachment. This includes diseases of the choroid, RPE, and retina. Most of these can be differentiated by other ophthalmoscopic findings; however, it is essential to rule out the presence of choroidal neovascular membranes. These membranes share many of the same characteristics of ICSC and can be seen at any age. Both young adults with presumed ocular histoplasmosis and older patients with age related maculopathy may develop choroidal neovacular membranes. For this reason, fluorescein angiography should be considered in all cases of ICSC. Other causes must be considered in the differential diagnosis of ICSC. For example: optic disk pits, optic disc coloboma, optic neuritis, papilledema, macular holes, choroidal tumors, uveitis, vitreous traction, peripheral rhegmatogenous retinal breaks, and systemic hypertension can all produce neurosensory retinal detachments[17].

The initial management of ICSC centers on the single decision of intervention versus monitoring. Either choice will end in the same visual outcome. The majority of acute episodes of ICSC left untreated will resolve within six to twelve months of initial onset, and vision will return to a

normal functional level in most patients[8]. Up to 60% of patients will be completely asymptomatic upon resolution, achieving 20/20 visual acuity[5]. The only form of intervention that has demonstrated some improvement in patients with ICSC is direct laser photocoagulation at the site of RPE leakage[18,19]. No pharmacologic agents have yet been demonstrated to be useful in treating ICSC. It is important to realize that laser photocoagulation has no impact on final visual outcome, it simply speeds the rate of resolution of the serous detachment. Visual function of patients treated with laser photocoagulation will improve four times faster than those simply monitored, but the final visual outcome for treated versus non-treated patients is the same. Laser photocoagulation does not change the recurrence rate for ICSC, nor does it impact the likelihood of developing the more severe progressive form of the disease. The use of laser photocoagulation does not come without risk. It has the potential to induce the formation of choroidal neovascularization and, if directed too close to the foveal avascular zone, may permanently scar central vision. For this reason, laser photocoagulation should only be considered after four to six months of monitoring in the majority of patients. The timetable for photocoagulation may be pushed forward in patients who develop choroidal neovascularization or who have permanent changes from ICSC in the fellow eye or a history of multiple recurrences. Laser photocoagulation may be considered for those who require improved vision for work, but only after thorough counseling of the risks involved.

Fortunately in the management of this patient, laser photocoagulation was not indicated. This patient, as with most patients, demonstrated frustration at the lack of intervention early in the course of management. Patience exercised in delaying treatment was rewarded with good visual outcome and the patient was spared from the potential adverse side effects of photocoagulation. The prognosis for the patient remains guarded. The findings of his right eye support a prior history of acute ICSC and underscore the bilateral nature of this disease. Recurrence is a significant issue with ICSC and patients must be warned that there is 30-40% likelihood that they will experience another episode of acute ICSC in the same or fellow eye in the future[1,5]. The prognosis for most patients with ICSC is very good. As noted earlier, the vast majority of patients will return to normal, functional vision after a few months of resolution. However, despite the relative absence of symptoms following resolution of the RPE and neurosensory detachments, most patients will demonstrate some residual metamorphopsia, altered color vision, or reduced brightness perception with testing [1,5,8]. The minority of patients that go on to develop the chronic progressive form of the disease face the greatest threat to vision. These patients develop a generalized loss of RPE function, and may demonstrate RPE atrophy, drusen, persistent choroidal exudation, or choroidal neovascularization. This chronic form can result in significant visual damage to both eyes leaving the patient functionally disabled.

Conclusion

This case demonstrates the role of patient history, clinical observation, and angiographic studies in the diagnosis of idiopathic central serous chorioretinopathy. In most cases the diagnosis can confidently be made without angiographic testing; however, the information derived from angiography is critical to detect the extent of the retinal abnormalities and to exclude the presence of other sources of serous RPE and neurosensory retinal detachment. It is important for clinicians to appreciate their role in educating patients about the natural course of this bilateral disease and to resist the temptation to intervene with photocoagulation before it is truly indicated. Thus avoiding potential complications. Although the prognosis for patients with

ICSC is generally favorable, patients should continue to self-monitor their vision even after resolution, as recurrence and choroidal neovascular membrane development is always a possibility. Furthermore, patients with resolved ICSC should be followed annually to monitor for any occult retinal changes.

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