Long-term Follow-up of Idiopathic Central Serous Chorioretinopathy by Fluorescein Angiography

RICHARD LEVINE, MD, ALEXANDER J. BRUCKER, MD, FANE ROBINSON, MD

Abstract: Idiopathic central serous chorioretinopathy (ICSC) is typically described as a self-limited, unilateral disease that affects healthy, young adult males. The authors studied 13 patients (14 eyes) who had documented spontaneous resolution of symptomatic macular detachments. These patients were evaluated in a longitudinal fashion to determine the fate of the retinal pigment epithelium (RPE) as viewed angiographically, both in the initially affected eyes and the fellow eyes. At the time of follow-up examination of the initially affected eyes, non leaking RPE defects had developed inside the areas of previous serous detachment in all cases. Nonleaking RPE defects had developed outside these areas in six (43%) eyes. Two (14%) eyes had new, asymptomatic macular detachments. Six (42%) fellow eyes had new RPE window defects; two (17%) of them also had active RPE dye leakage resulting in asymptomatic macular detachment at the final examination. Four (29%) originally involved eyes and one (8%) fellow eye lost more than two lines of Snellen visual acuity during the follow-up period. The authors' results suggest that ICSC may be a progressive bilateral disease that develops asymmetrically and causes diffuse RPE changes not localized to the area of serous detachment. Long-term follow-up of these patients may, therefore, be advisable. Ophthalmology 96:854-859, 1989

Idiopathic central serous chorioretinopathy (ICSC) is typically a sporadic, self-limited disease of young adult males. It is characterized by a unilateral serous detachment of the neurosensory retina, usually in the macular region. The focal accumulation of fluid in the subretinal space in these patients is probably related to a defect in the retinal pigment epithelium (RPE).

The purpose of this study is to evaluate RPE changes in patients studied longitudinally who had previously documented spontaneous resolution of a symptomatic macular detachment. The fellow eye was also evaluated to determine the presence or absence of any abnormalities.

PATIENTS AND METHODS

We reviewed the charts of 33 patients seen at the Scheie Eye Institute between 1977 and 1981 and diagnosed as having ICSC. Three patients were excluded from the study because of previous intraocular surgery, ocular trauma resulting in serous retinal detachment, and serous detachment of the macula secondary to posterior scleritis. Four additional patients who had received focal retinal laser photocoagulation therapy for treatment of ICSC...
were excluded. Seventeen of the remaining 26 patients met all of the following criteria and were eligible for inclusion into the study: (1) younger than 50 years of age at the time of initial diagnosis; (2) fluorescein angiographic evidence of dye leakage beneath the neurosensory retina typical of ICSC ("expanding pinpoint" or "smokestack"); (3) documentation of complete resorption of subretinal fluid during the follow-up period; (4) fluorescein angiographic photographs of the fellow eye. Of the 17 eligible patients, 13 patients (14 eyes) were available for follow-up examination and were entered into the study.

All follow-up examinations included best-corrected Snellen visual acuity, slit-lamp examination, direct and indirect ophthalmoscopy, stereoscopic color fundus photography, and intravenous fluorescein angiography of both eyes. The follow-up interval ranged from 7 to 12 years (mean, 8.2 years).

Fluorescein angiography was performed using 3 ml of 25% sodium fluorescein dye injected in the antecubital or adjacent vein over approximately 5 seconds. All pictures were taken with a Zeiss FF3-model Stereoscopic Fundus Camera using Kodak Tri-X film. The fluorescein angiograms were printed on Kodak Fine-Grain Positive film.

INITIAL CLINICAL CHARACTERISTICS

All patients were men. They ranged in age from 20 to 49 years (mean age, 40 years) when first diagnosed as having ICSC. There were nine whites, three blacks, and one Asian. Initial visual acuity in the affected eyes ranged from 6/6 to 6/24 (mean, 6/9).

The macula was detached at the time of initial examination of all 14 eyes, with foveal sparing in 2 eyes. The size of the serous retinal detachment (measured using its largest diameter) varied from 1.5 to 4.0 disc diameters (DD) (mean, 2.3 DD). This measurement was made in relation to the optic disc size of each patient. A clinically definable RPE detachment within the serous detachment was seen in two (14%) eyes. All affected eyes were comprehensively evaluated to determine if RPE changes were...
Fig 2. Case 4. A, fluorescein angiogram of superotemporal arcade 9 years later, at final examination. The area of RPE staining, seen in Figure 1D, has enlarged approximately fourfold since the initial examination. The adjacent area, previously a window defect, now shows fluorescein dye leakage associated with a sensory retinal detachment. B, fluorescein angiogram of same patient, at final examination. Extensive RPE window defects and pigment migration have occurred inside the area of previous sensory retinal detachment.

present in addition to the serous detachment. Pigmentary changes at the level of the RPE were present in eight (57%) eyes outside of the serous detachment. Subretinal precipitates were found below the detached neurosensory retina in three (21%) eyes. Drusen in the pattern of Doyne's honeycomb dystrophy were present in one (7%) eye, not related to the area of RPE leakage.

INITIAL ANGIOGRAPHIC CHARACTERISTICS

The initial fluorescein angiograms demonstrated active leakage of dye into the subretinal space in all cases. The leakage occurred as a gradually expanding dot of hyperfluorescence in ten (71%) eyes, and as a classic "smokestack" leak in four (29%) eyes (Fig 1). The leakage site varied in size from a pinpoint to approximately one-half DD. The leakage occurred within the foveal avascular zone in eight (57%) eyes; extrafoveally, but within the macula in six (43%) eyes of which two (14%) were within the papillomacular bundle.

CLINICAL FINDINGS DURING INTERIM FOLLOW-UP PERIOD

Eleven of the 13 patients denied symptomatic recurrences of ICSC during the follow-up period. Two patients (cases 6 and 12) had recurrent ipsilateral episodes of ICSC documented clinically and angiographically. These cases recurred 2 years (case 6) and 6 years (case 12) after the initial diagnosis. Both cases resolved fully during the interim follow-up period.

FOLLOW-UP CLINICAL AND ANGIOGRAPHIC CHARACTERISTICS

Serous detachments of the macula were present in four (31%) eyes at the most recent follow-up examination. These were all associated with active dye leakage angiographically. In one (case 12) of these four eyes, the leakage occurred inside the area of previous serous retinal detachment. The remaining three patients had leaks occurring outside the area of previous serous retinal detachment. One occurred ipsilaterally, above the superotem-
poral arcade (case 4, right eye; Fig 2A), and two developed in fellow eyes (case 4, left eye; case 5, left eye). The four patients with macular detachments at this final examination were asymptomatic.

The visual acuity in the initially affected eyes ranged from 6/6 to 6/24 at the final examination. Loss of more than two lines of visual acuity during the follow-up period was experienced by four patients. Only one of these patients (case 12) had a macular detachment (involving the fovea) at the final examination. None of the initially unaffected eyes lost more than two lines of visual acuity during the follow-up period.

All focal pigment epithelial leaks evolved into clinically observable RPE atrophic areas, which corresponded angiographically to RPE window defects. All patients developed RPE atrophy and varying degrees of associated RPE clumping in the areas of prior serous detachment of the neurosensory retina (Fig 2B). Although pigmentary changes at the level of the RPE were present in eight (57%) eyes outside of the serous detachment at the initial examination, new RPE changes were later found outside of the area of original serous detachment in six (43%) eyes at the final examination. These changes were exemplified in case 6 (Figs 3, 4). A curvilinear, vertically oriented RPE atrophic tract developed in one eye (case 10) originating in the infranasal macula that descended inferiorly to the equator (Figs 5, 6). This was not associated with active leakage nor with a dependent peripheral serous detachment when seen at the follow-up examination.

The fellow eyes of all patients in the study were examined both clinically and by fluorescein angiography. New RPE lesions appeared in six (50%) of the fellow eyes. Retinal pigment epithelial changes seen at the time of initial examination had enlarged in seven (57%) of the fellow eyes; in one of these eyes (case 4, left eye) an active juxtapapillary leak with a serous detachment sparing the...
fovea was seen (Fig 7). No RPE changes were present in this area of leakage on the original physical examination or fluorescein angiogram. Table 1 summarizes the clinical and angiographic findings of the long-term follow-up of these patients.

**DISCUSSION**

The natural history of untreated ICSC was described previously. That study prospectively evaluated the clinical course and visual acuity outcome of patients with nontreated ICSC. Retinal pigment epithelial defects were noted angiographically in the originally involved eye and the fellow eye; however, the authors never commented on the relationship of new RPE changes to previous serous detachments or changes in the RPE of the fellow eye. The most extensive retrospective long-term follow-up study to date has evaluated visual outcome, incidence recurrence, and pigment epithelial leakage characteristics of ICSC. That study, however, differs from ours because their patients were selected for evaluation when they had symptomatic disease.

We studied a group of patients with nontreated ICSC in whom resorption of subretinal fluid had been documented. We sought to determine what, if any, pigment epithelial abnormalities would later develop in these asymptomatic patients. All patients were called and asked to return for follow-up. We found clinically silent macular detachments were present in 31% of eyes at follow-up examination. New leaks occurred with approximately equal frequency both inside and outside areas of previous serous detachment.

In all of our cases, focal pigment epithelial leaks evolved into window defects angiographically, and diffuse RPE atrophic changes developed within previously detached areas. One patient manifested an extramacular RPE atrophic tract, a feature of long-standing subretinal fluid accumulation previously reported by Gass and Yannuzzi. The finding of new pigment epithelial changes outside areas of previously documented serous detachments has not been described previously. One explanation for this finding is that these areas may be the manifestation of a diffuse, progressive RPE disturbance that is accompanied, at some time in the course of the disease, by an episode of ICSC. Alternatively, these may represent sequelae of asymptomatic detachments in that location. Our finding of clinically silent macular detachments in four patients supports this possibility.

The fellow eye, evaluated as a secondary part of the study, was the site of progressive, clinically observable pigment epithelial abnormalities in the majority of cases. As previously illustrated, one patient had asymptomatic macular detachments with angiographic evidence of leakage in both eyes at the most recent follow-up examination.

The choice of nontreated cases of ICSC for prospective evaluation was made to remove a confounding variable from the evaluation of the natural history of the disease. Arguably, excluding treated patients may have introduced a selection bias in favor of more clinically benign and uncomplicated cases. On the other hand, the fact that fluorescein angiography was performed initially in these patients might indicate that they represented a high-risk group in that angiography is not routinely done on all patients at the first presentation with ICSC. Thus some patients with ICSC were not included in the study because fluorescein angiograms had not been done initially. It should also be noted that the average age of our patients was 40 years, somewhat older than the figure quoted in previous studies. Also, 23% of our patients were black, which is somewhat unusual in this disorder. Therefore, it may be that our patients include a demographic subset who are more likely to acquire a progressive RPE disturbance.

This is the first study to document angiographically, in a longitudinal fashion, progressive RPE abnormalities that develop after resolution of ICSC. We studied patients with resolved, angiographically documented ICSC and found new angiographic abnormalities including RPE window defects both inside and outside areas of previous neurosensory retinal detachment and new, active leakage sites.
Table 1. Summary of Clinical and Angiographic Findings of Long-term Follow-up of Idiopathic Central Serous Chorioretinopathy

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Race/ Sex</th>
<th>Age (yrs)</th>
<th>Affected Eye</th>
<th>Visual Acuity</th>
<th>SDR Size (mm)*</th>
<th>Fovea Involved</th>
<th>Leak Site</th>
<th>RPE Changes</th>
<th>Duration (yrs)</th>
<th>Follow-up Acuity</th>
<th>Fate</th>
<th>New ICSC</th>
<th>Comments</th>
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<td>Bilateral ICSC at presentation; new RPE changes within areas of SDR at final examination</td>
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SDR = serous detachment of retina; DO = disc diameter; RPE = retinal pigment epithelium; ICSC = idiopathic central serous choroidopathy; W = white; OD = right eye; OS = left eye; FAZ = foveal avascular zone; PMB = papillomacular bundle; EF = extrafoveal; R = resolved; B = black; A = Asian.

* Size of serous detachment of retina was measured in larger diameter.
† Fate of serous detachment: clinical and angiographic appearance of serous detachment at most recent (follow-up) examination.
‡ The affected eye.

Our results suggest that ICSC may less often be the benign and self-limited condition than it is thought to be and in certain patients may herald a bilateral, progressive RPE disturbance necessitating long-term follow-up of the patient.

REFERENCES

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