Albrecht v. Graefes Arch. klin. exp. Ophthal.
206, 169–178 (1978)

Graefes Archiv
für klinische und experimentelle
Ophthalmologie

Central Serous Choroidopathy with Bullous Retinal Detachment

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Summary. We report here a new type of secondary retinal detachment that has never been clearly defined. The characteristic features of the disease are: (1) prevalence in middle-aged males, (2) bilateral involvement, (3) frequent existence of prodromal lesions that over long periods resemble central serous retinopathy, (4) in the evolution stage, appearance of multiple yellowish white exudative flecks of one-half to one disc in diameter at or near the posterior pole of the fundus, (5) fluorescein studies revealing pronounced leakage of dye from the choroid into the subretinal space at the sites of exudates, (6) retinal detachment of various degrees with shifting subretinal fluid and without tears, (7) no evidence of intraocular inflammation, (8) no filling abnormalities seen in the choroidal fluorescence, (9) no response to medical therapy, including steroids and antibiotics, (10) photocoagulation to leakage sites leading to rapid resolution of retinal detachment; otherwise, spontaneous healing of detachment occurring within about 7–9 months, leaving fibroblastic macular scars and marked visual loss, and (11) no evidence of systemic findings that may be of etiologic significance. From this characteristic clinical picture, the idea of a new clinical entity must be considered. Our findings in 35 eyes from 18 Japanese patients are discussed.

Introduction

A peculiar type of secondary retinal detachment has been reported by several authors from various independent institutions (Gass, 1973; Tsukahara and Morii, 1973; Tsukahara, 1974; Mimura et al., 1973a and b; Shimizu et al., 1974; Uyama and Tsukahara, 1977). This condition has apparently not been clearly defined in the past although similar cases were reported by Urayama et al. (1971). Characteristic features of this retinal detachment are: (1) almost exclusively, middle-aged males are affected, (2) bilateral involvement, (3) no intraocular inflammation, (4) multiple small exudations of one-half to one disc in diameter, gray or yellowish white in color and round or doughnut-

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0065-6100/78/0206/0169/$ 02.00

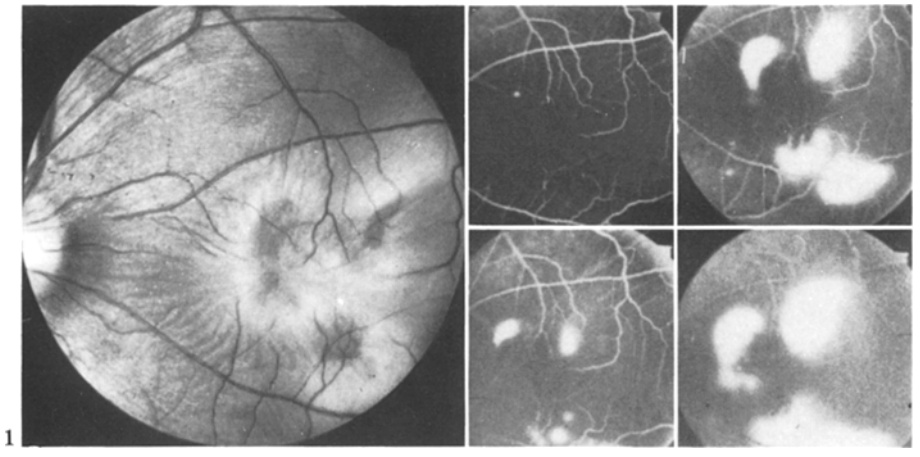


Fig. 1. Case 1, left eye. Primary lesions surrounded by grayish ring-shaped rim in the macula. Radial wrinkle of the retina around the lesions can be seen
Fig. 2. Case 1, left eye. Multiple leakage of the dye noted in the macular area

like in shape at or near the posterior pole of the fundus, (5) pronounced leakage of injected dye from the choroid into the subretinal space at the site of exudations, (6) retinal detachment of various degrees with shifting subretinal fluid and without retinal break, (7) no response to corticosteroids and antibiotics, (8) photocoagulation to the leakage sites leading to rapid resolution of detachment, (9) spontaneous healing of detachment occurring 7–9 months after the onset of the disease, leaving frequent macular scars and marked visual loss, and (10) extensive general examination failing to reveal any common finding that could be of etiologic significance. Our data on eyes from 18 Japanese patients are discussed.

Report of Representative Cases

Case 1. A healthy 40-year-old man noticed disturbances in his left visual field 2 months prior to visiting us on February 20, 1970. In June 1965 he developed central scotoma in the left eye and was treated for 6 months for central serous retinopathy. The condition relapsed in December 1968 and his eye was treated. This time, prior to referral to us, oral betamethasone was administered from January 20, 1970, to February 20, 1970, by another ophthalmologist, but there was no improvement. When he next visited our clinic, visual acuity in his right eye was 0.1 (20/200) and corrected to 1.0 (20/20) with −2.0D sph., and in his left eye 0.1 (20/200) (n.c.). Ocular tension in his right eye was normal and was unmeasurably soft in the left eye. The anterior segment of both eyes was normal. Fundus examination of the left eye in sitting position revealed a bullous detachment of the retina in the inferior half of the fundus. Subretinal fluid shifted rapidly with changes in positioning of the patient. Macular focal areas were surrounded by a grayish ring (Fig. 1), and fluorescein angiography revealed pronounced leakage of dye at the focal lesion (Fig. 2). In the macula, there was a flat retinal detachment and

radial wrinkles of the retina were present around the grayish lesions (Fig. 1). His right eye was normal until March 6, 1970, when he complained of blurred vision in it. Ophthalmoscopy of the right eye revealed a disc-shaped, flat elevation of the macula that resembled central serous retinopathy. Fluorescein angiography confirmed multiple leakage of dye into the macula. Small grayish spots and small retinal pigment epithelium detachments were evident at the leakage sites.

Retinal detachment failed to respond to chemotherapy, including corticosteroids and antibiotics. Detachment in the macula of the right eye extended from the macula to the equator inferiorly, but healed spontaneously about 18 weeks after its onset, leaving a mottling of the macula. Visual acuity in the right eye on October 7, 1971, was 0.5 (20/40) and was corrected to 1.0 (20/20) with −1.25D sph. Bullous detachment in the left eye started to subside after 4 months and disappeared 7 months after its onset. Macular scars with fibrous folds remained and visual acuity was 0.1 (20/200) (n.c). However, neovascularization developed on the disc of the left eye and vitreous hemorrhage occurred 4 times from June 23, 1970, to January 26, 1971. Xenon-arc photocoagulation to eradicate neovascularization was performed and recurrent vitreous hemorrhage was prevented. Visual acuity in the left eye on October 7, 1971, was 0.1 (20/200) and was corrected to 0.2 (20/100) with −2.OD sph. During convalescence from February 23 to July, 1970, extensive laboratory examinations were done, but there were no findings that might be of etiologic significance. Elevation of the cerebrospinal protein was not confirmed.

Case 2. A healthy 47-year-old man developed metamorphopsia of the left eye during a stay in Hong Kong 3 months before visiting us. He was treated for retinal hemorrhage for 2 months with no improvement. This time he was told he had a detached retina in the left eye and was referred to us on February 16, 1971. When first seen by us, visual acuity in his right eye was 0.6 (nearly 20/30) and correctable to 1.0 (20/20) with a +1.5D sph. Visual acuity in his left eye was 0.1 (20/200) and 0.2 (20/100) with a + 1.5D sph. Ocular tension in the right eye was 12 mm Hg and in the left eye was unmeasurably soft. The anterior segments of both eyes were normal. The aqueous was clear, and although a few floaters were present in the vitreous humor, there were no signs of inflammation. Ophthalmoscopy of the left eye done with the patient in a supine position revealed a total ballooning detachment of the retina, without breaks. Subretinal fluid moved quickly following change of head position and a highly ballooning inferior retinal detachment with a flat detachment in and around the macula in sitting position was present. The extreme periphery was examined carefully but an exudative focus could not be confirmed. On February 23, 1971, the patient complained of metamorphopsia of the right eye. Two bleb serous detachments of the retinal pigment epithelium nearly one disc in diameter were present in the macula, but there was no retinal detachment (Fig. 3). These detachments of the retinal pigment epithelium remained unchanged for the 5-year follow-up period.

During convalescence from February 20 to April 17, 1971, extensive laboratory tests, including a general physical examination, hemogram, blood chemistry, and chest x-ray, were carried out, but results were negative. The efficacy of any form of medical treatment, including corticosteroids and antibiotics, was not established.

Case 3. A healthy 36-year-old male complained of blurred vision in both eyes and was referred to us on November 2, 1972. His right eye had been treated for central serous

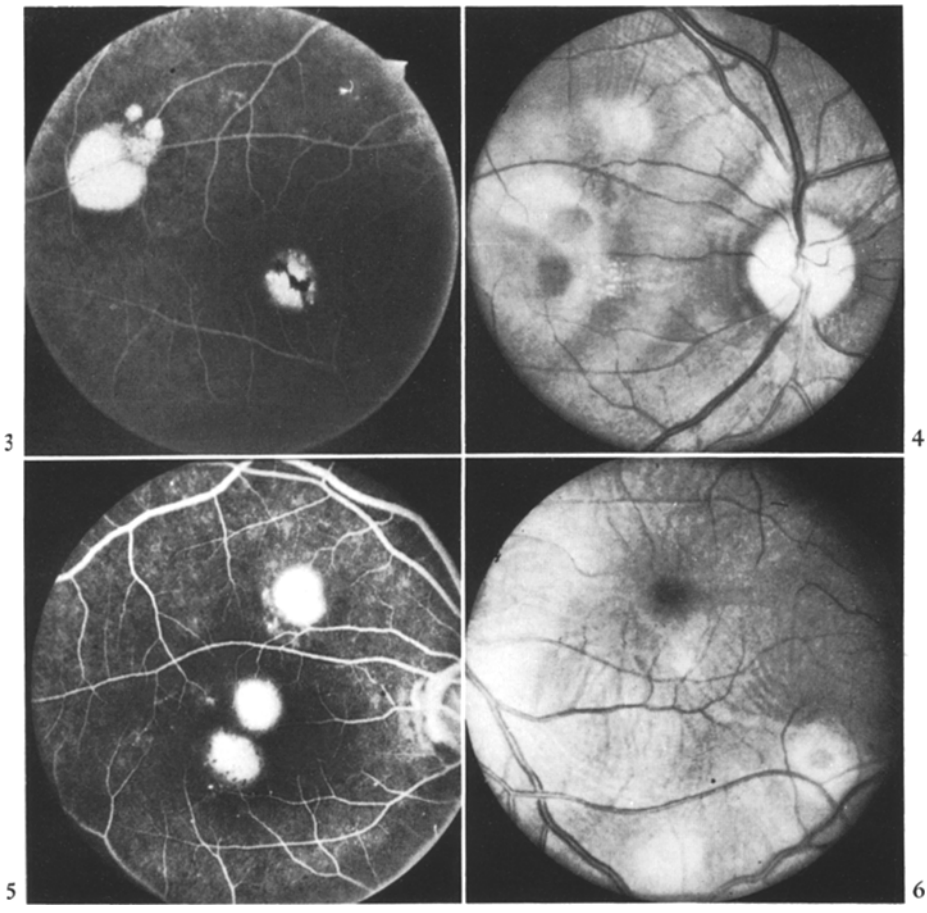


Fig. 3. Case 2, right eye. Fluorescein angiogram. Two serous retinal pigment epithelium detachments without subretinal leakage are noted in the macular area
Fig. 4. Case 3, right eye. Exudative focal areas in the macula show a doughnutlike appearance. Radial retinal wrinkles and a small amount of hard exudates are visible
Fig. 5. Case 3, right eye. Fluorescein angiogram demonstrates three retinal pigment epithelium detachments with subretinal leakage
Fig. 6. Case 3, left eye. Two exudative focal areas near the macula

retinopathy for 3 years. Three days prior to his visit to us he developed blurred vision in his left eye. He noticed no alopecia, polliosis, vitiligo, or dysacousis. When first seen by us, his vision was finger counted at 2 m in the right eye and 0.2 (20/100), correctable to 0.3 (20/70) with +0.5D sph. in the left eye. Ocular tension was normal. The anterior segment of both eyes was normal and the vitreous clear. Ophthalmoscopy of the right eye revealed two exudative flecks one-half disc in diameter in the macula. Those exudative focal areas were surrounded by a gray ring and had a doughnutlike appearance (Fig. 4). Superior to those foci was another grayish lesion of the same size. A left retinal detachment involved the macula and there was no bullous detachment.

Radial retinal wrinkles surrounded the exudative flecks and a small amount of yellowish, hard exudates near the nasal margin of the foci was noted (Fig. 4). Fluorescein angiography confirmed serous detachment of the pigment epithelium with subretinal leakage of the dye at those gray exudative areas (Fig. 5). Ophthalmoscopy of the left eye revealed a flat macular elevation that extended inferiorly. A gray exudative fleck one-half disc in diameter was observed at 3 disc-diameter and temporal-superior from the disc. Fluorescein angiography revealed a prominent leakage of dye. Xenon-arc photocoagulation was applied at the leakage site, detachment of the macula subsided rapidly, and visual acuity recovered, reaching 0.3 (20/70) to 0.7 (20/30). After 2 weeks, however, a flat, macular elevation reappeared and two newly developed gray exudative areas were confirmed. A few days later, a gray color in the center of those flecks faded and a doughnutlike appearance was seen (Fig. 6). Fluorescein angiography confirmed leakage of dye at these newly developed foci. Reviewing the fluorescein angiograms that were taken before recurrence of the macular detachment, it is interesting to note that newly developed leakage sites were observed as small, round fluorescent areas and were interpreted to be small detachments of the retinal pigment epithelium without diffusion of dye into the subretinal space. This time, spontaneous resolution of new detachment was soon quite evident and the macula became flat within 3 months, without photocoagulation. The visual acuity of the left eye improved to 1.0 (20/20). Retinal detachment of the right eye healed spontaneously 8 months after the first visit, but visual acuity was finger counted at 4 m because of the macular scar. Extensive clinical investigations revealed all values to be within normal limits. The efficacy of any form of medical treatment was not established.

Summary of Cases

In the past 7 years a new type of secondary retinal detachment, not clearly recognized in the past, has been encountered in our clinic in 35 eyes from 18 patients. Characteristic features are summarized as follows:

1. Age, Sex, and Bilaterality. Age distribution, sex, and bilaterality of the disease are summarized in Table 1. Almost exclusively, middle-aged males are affected. Of 18 patients, only one had unilateral involvement.

2. Ocular Findings. Representative case histories often reveal repeated recurrence of central serous retinopathylike lesion in the prodromal stage and the eyes are treated as cases of central serous retinopathy for many years. There are no signs of inflammation

Table 1. Distribution Factors

Age (years)	Cases	Sex
21–30	1	Male 15 # (83 %)
31–40	7	Female 3 (17 %)
41–50	9	
51–60	0	
61–70	1	
Average, 41 years		

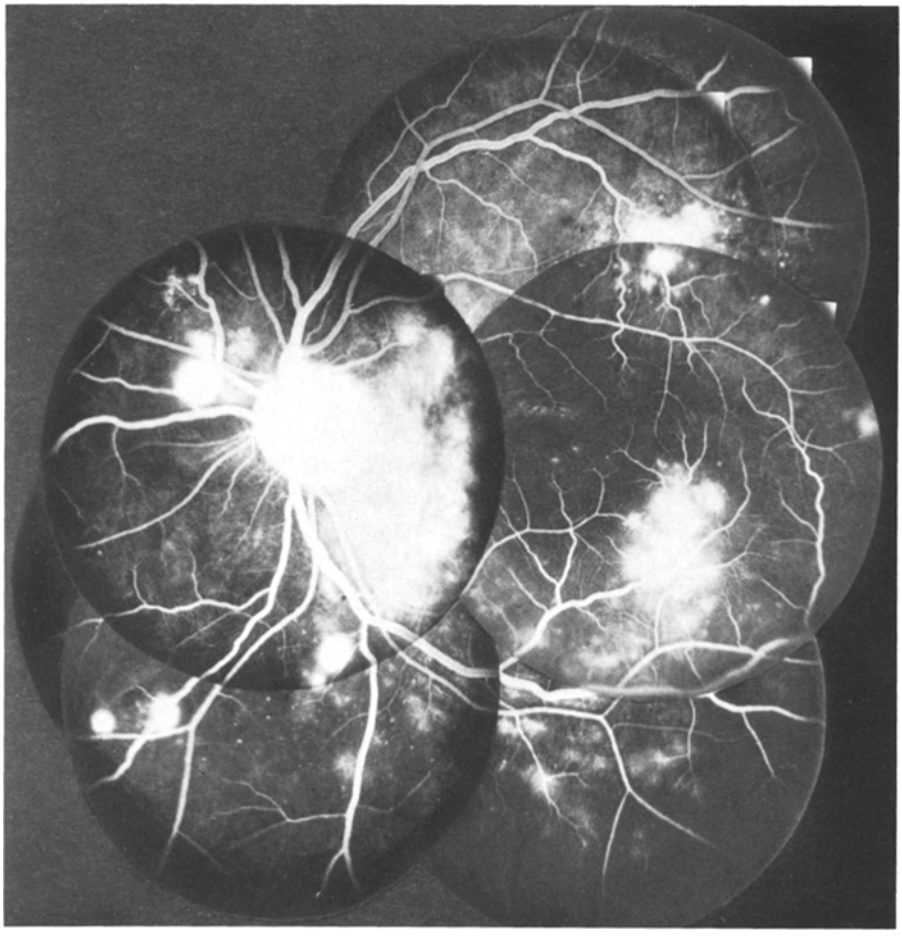
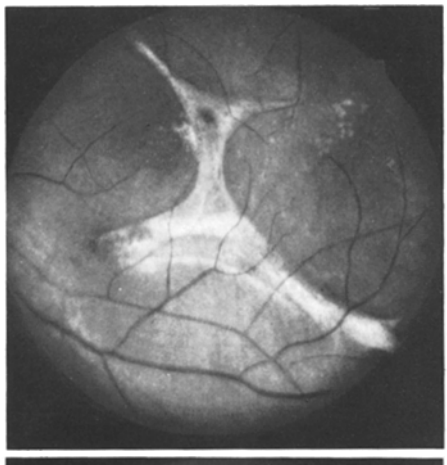


Fig. 7. Fluorescein angiogram. Left eye of a 44-year-old man. Multiple leakages of fluorescein were present in and around the macula

in the anterior and posterior portions of the eye. A characteristic picture is noted in the fundus. In the evolutional stage, a bullous retinal detachment without breaks is observed. Subretinal fluid is readily movable and detachment rapidly changes following change in head position. In cases where fundus examination is done with the patient in the supine position, total ballooning retinal detachment is revealed, and a highly ballooning inferior detachment with a flat detachment in and around the macula is confirmed in the sitting position (Fig. 9). Even careful examination of the extreme periphery of the fundus produces no evidence of inflammatory exudative foci. Multiple primary lesions (Figs. 1, 2, 4, and 7) are present at or near the posterior pole of the fundus and are commonly identified as grayish or yellowish gray focal areas of one-half or one disc in diameter or as discrete areas surrounded by a grayish rim with a doughnutlike appearance (Figs. 1 and 4). In the initial stage, the focal area is diffusely gray in color, but this gray



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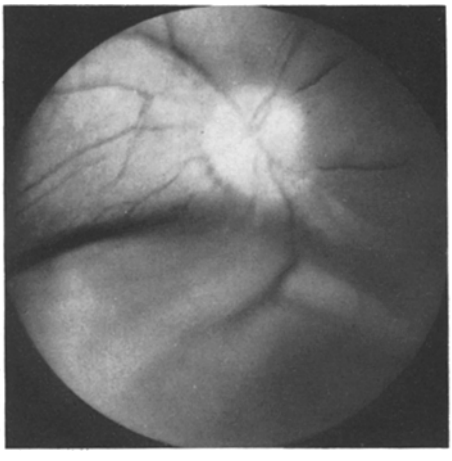

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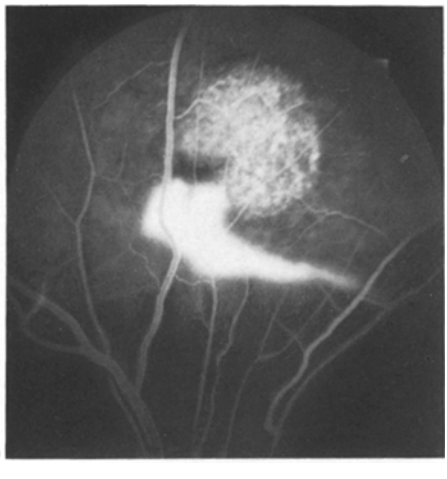

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Fig. 8. Fibroblastic macular scar

Fig. 9. A highly ballooning inferior retinal detachment in the right eye of a 47-year-old woman in the sitting position. In the supine position, the detachment rapidly changed to total ballooning detachment

Fig. 10. In the same case as in Figure 9, fluorescein angiography revealed a single prominent leakage of the dye and atrophy of the retinal pigment epithelium

color in the central area of the lesion fades away and the lesion then shows a doughnut-like appearance (Figs. 1 and 6).

Fluorescein angiography reveals prominent leakage of dye from the choroid into the subretinal space at the sites of primary lesions (Figs. 2, 7 and 10). The dye spreads into the subretinal space and diffuses into the retina. A nonperfused area or retardation of choroidal circulation cannot be confirmed.

The severity of retinal detachment is often similar, but not necessarily simultaneous in both eyes. In some patients, one eye has a bullous detachment while the other eye has serous detachments of the retinal pigment epithelium without subretinal leakage, as seen in case 2, or only flat and localized macular elevation, as observed in case 1.

3. General Findings. Extensive medical evaluation including a general physical examination, hemogram, blood chemistry, urinanalysis, and chest x-ray studies reveal data to be within normal limits. Elevation of cerebrospinal protein was not confirmed.

4. Therapy and Visual Outcome. The efficacy of any form of medical treatment including antibiotics and corticosteroid was not established. Photocoagulation of leakage sites

of fluorescein cause rapid resolution of retinal detachment, although additional coagulation of newly developed leakage sites were frequently necessary. In cases where there was no photocoagulation, a spontaneous healing of bullous detachment resulted 7–9 months after the onset of the disease, leaving fibroblastic macular scars (Fig. 8). In one eye, neovascularization developed on the optic disc and caused recurrent vitreous hemorrhage, which was prevented by photocoagulation. Visual outcome is summarized in Tables 2–4.

5. Electrophysiologic Tests. Electrophysiologic tests of retinal and pigment epithelium were not performed.

Table 2. Visual acuity in evolutional and final stage of the disease

Visual acuity	Evolutional stage (eyes)	Final stage (eyes)
1.0 or better	2 (6 %)	8 (24 %)
0.6–0.9	2 (6 %)	2 (6 %)
0.2–0.5	7 (21 %)	9 (27 %)
Under 0.1	22 (67 %)	14 (42 %)
Total	33	33

Table 3. Visual acuity in nonphotocoagulated eyes

Visual acuity	Evolutional stage (eyes)	Final stage (eyes)
1.0 or better	2 (10 %)	3 (15 %)
0.6–0.9	1 (5 %)	0
0.2–0.5	2 (10 %)	6 (30 %)
Under 0.1	15 (75 %)	11 (55 %)
Total	20	20

Table 4. Visual acuity in photocoagulated eyes

Visual acuity	Before photocoagulation (eyes)	Final
1.0 or better	0	5 (39 %)
0.6–0.9	1 (7 %)	2 (15 %)
0.2–0.5	4 (31 %)	2 (15 %)
Under 0.1	8 (62 %)	4 (31 %)
Total	13	13

Discussion

In our experience, the most characteristic objective features of this disease are retinal detachment without tear of variable severity and multiple, round exudative focal areas at and near the posterior pole of the fundus. The severity of retinal detachment is often similar, but not always simultaneous in both eyes. As shown in cases 1 and 2, a bullous detachment is present in one eye, while localized serous detachments of the retinal pigment epithelium or central serous retinopathylike macular elevation is observed in the other eye. Gass reported 5 patients in whom rapid visual loss secondary to bullous retinal detachment developed in the posterior pole and inferior fundus. In our experience, case histories revealed that patients were frequently treated as cases of central serous retinopathy for many years. It is likely that a bullous detachment in the evolutional stage of the disease is preceded by a flat, macular elevation in the prodromal stage. The detachment fails to respond to steroid therapy, but photocoagulation applied to the leakage sites of the dye, as detected by fluorescein angiography, produces a resolution of detachment, although frequent additional photocoagulation should be applied to newly developed leakage sites. However, in cases in which the leakage sites are present in the central macular area and photocoagulation cannot be performed, spontaneous healing of the detachment results 7–9 months after onset, frequently leaving fibroblastic macular scars. Prognosis of central visual acuity is frequently poor, but spontaneous reattachment with minimal visual loss may occur in some cases.

Etiology of the detachment is obscure. Extensive medical evaluation fails to reveal common findings that might be of etiologic significance. Gass (1973) and Tsukahara (1973, 1974) considered this condition to be a severe and an unusual form of central serous retinopathy. These are different clinical expressions of the same condition, with the former being multifocal and the latter focal. Both central serous retinopathy and this type of detachment have the following in common: (1) age distribution and sex of the patients, (2) presence of leakage areas of the dye revealed by fluorescein angiography, (3) failure of the retinal detachment to respond to steroid therapy, and (4) resolution of the detachment following photocoagulation of leakage sites. However, clinical features of both conditions are different in (1) severity of retinal detachment, (2) multiplicity of primary focal areas at or near the macula, (3) final visual acuity, (4) fibroblastic macular scars as a frequent end stage of the disease, and (5) atrophy of the retinal pigment epithelium over a broad area. A breakdown of function of the retinal pigment epithelium as a barrier between the choroid and retina induces both disorders. At the present time, however, it is difficult to determine whether the retinal pigment epithelium is a primary cause of this disorder or whether it is affected by disorders in the adjacent choroid. At any rate, the retinal pigment epithelium becomes a primary source of complication. The authors tentatively propose the term central serious choroidopathy (CSC) with bullous retinal detachment to describe the newly recognized secondary retinal detachment, and consider this to be a new clinical entity.

In 1963, Schepens and Brockhurst reported a serous retinal detachment as uveal effusion. Most of their patients were middle-aged. A serous retinal detachment with shifting of the subretinal fluid and without retinal break was evident. Another consistent

finding was elevation of the cerebrospinal protein without pleocytosis. Fluorescein angiography was not available at this time. In our patients, elevation of cerebrospinal protein was not confirmed and most of their cases differed from ours.

Acknowledgements. Thanks are due to M. Ohara, Kyoto University, for assistance with the manuscript.

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Received November 8, 1977