Graefe's Arch Clin Exp Ophthalmol (1998)
236:876–879

SHORT COMMUNICATION

Johanna M.M. Hooymans

Fibrotic scar formation in central serous chorioretinopathy developed during systemic treatment with corticosteroids

Received: 8 September 1997
Revised version received: 10 February 1998
Accepted: 18 February 1998

J.M.M. Hooymans (✉)
University Hospital of Groningen,
Department of Ophthalmology,
P.O. Box 30.001, 9700 RB Groningen,
The Netherlands, Fax +31-50-3611709

Abstract ● Background: The purpose of the study is to demonstrate the development of subretinal fibrotic scar formation in central serous chorioretinopathy (CSCR) that developed during systemic corticosteroid treatment. ● Methods: The clinical and photographic records of a patient in whom an unusual manifestation of severe central serous chorioretinopathy developed during systemic corticosteroid treatment were reviewed. Results: A 34-year-old man received high dosages of corticosteroids for several years because of severe asthma. There was no previous history of idiopathic CSCR. He developed three successive attacks of CSCR. On the third occasion, an unusual variant of severe CSCR was observed which presented as a bullous sensory retinal detachment with subretinal serofibrinous exudate followed by subretinal fibrosis and fibrotic scar formation with contracting bands. After tapering of the corticosteroid dosage to a low level the retinal detachment resolved without laser treatment. ● Conclusion: The finding suggests that during systemic corticosteroid treatment atypical manifestations of CSCR may develop with subretinal fibrosis and scar formation.

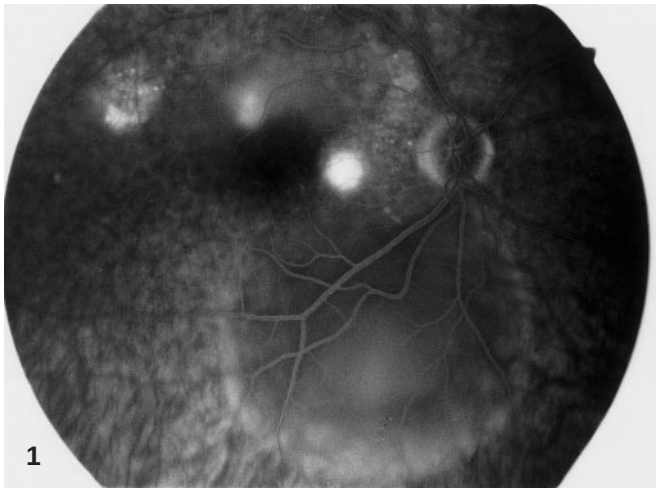
Introduction

Central serous chorioretinopathy (CSCR) is a disease that typically affects healthy adult males between 30 and 50 years of age [13]. It is characterized by the development of serous detachment of the sensory retina and of the retinal pigment epithelium (RPE), often during periods of emotional stress. In most cases spontaneous reattachment occurs in 3–6 months. The disorder recurs, however, in 20–30% of the patients [1]. An unusual variant of CSCR presents as a large bullous inferior retinal detachment with shifting fluid [3, 11]. Occasionally fibrin can be found in the subretinal space [5, 9]. Eventually, the fibrin plaque disappears. It is very unusual for the deposition of fibrin to be followed by fibrotic scar formation [3, 5, 9]. CSCR has been reported in association with corticosteroid use [2, 6, 7, 8, 12]. Treatment of CSCR with systemic corticosteroids is ineffective and in some cases causes severe exacerbation of the disease [6].

This report describes a patient who developed a severe and unusual form of CSCR while receiving high doses of corticosteroids systematically. During this third exacerbation a bullous retinal detachment with subretinal serofibrinous exudate developed. This was followed by fibrotic scar formation.

Case report

In June 1994, a 34-year-old man experienced decreased vision in both eyes. Since 1990 he had taken systemic steroids periodically because of very severe asthma. During the last year he had used 60 mg of prednisolone per day. During periods of emotional stress he was taking fluoxetine Prozac. His past ophthalmological history was unremarkable. At the time of his initial examination, approximately 3 weeks after the onset of the symptoms, his visual acuity was 0.2 in the right and 0.4 in the left eye. The aqueous humor and vitreous were clear in both eyes. The intraocular pressure was 14 mm Hg bilaterally. In the fundus of the patient's right eye there were four pigment epithelial detachments in the posterior pole and a serous retinal detachment adjacent to the optic disc. In the left



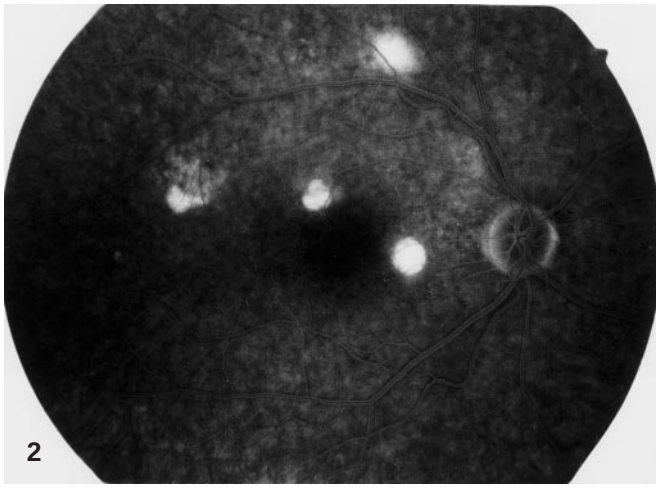
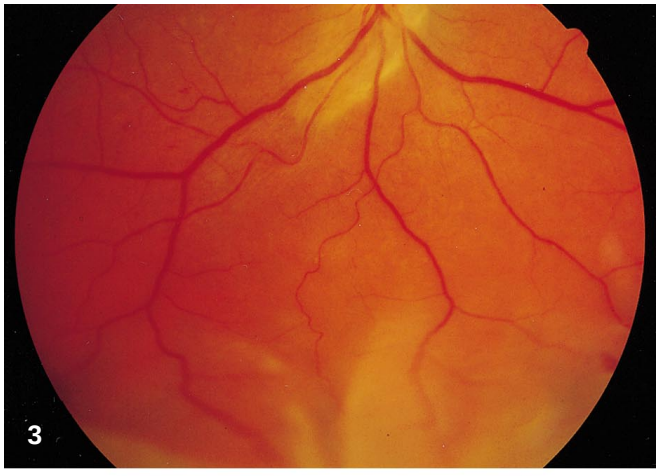


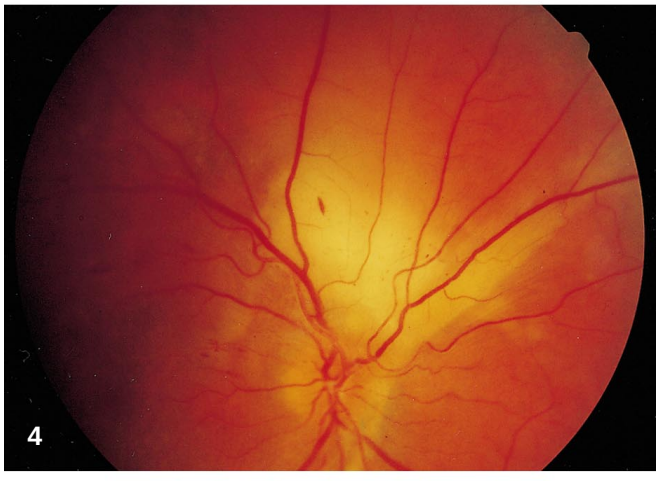
Fig. 1 Fluorescein angiogram of the right fundus demonstrates hyperfluorescence of retinal pigment epithelial detachments and a serous retinal detachment

Fig. 2 Fluorescein angiogram of the right eye 4 weeks after tapering the corticosteroid dosage to 35 mg. Note that the retinal detachment has resolved completely. There is still hyperfluorescence in the areas of pigment epithelial detachments

fundus he had a pigment epithelial detachment superior to the fovea. Fluorescein angiography confirmed the presence of serous retinal and pigment epithelial detachments (Fig. 1). Areas of RPE alteration that might have represented the sequela of previous episodes of the disease were not found. The prednisolone dosage was lowered to 35 mg/day, and 1 month later the serous retinal detachment in the right eye was resolved completely. Repeated fluorescein angiography revealed hyperfluorescence from the area previously affected by pigment epithelial detachments (Fig. 2). The patient's visual acuity had improved to 0.5 in the right and 0.8 in the left eye. His asthma did not allow further reduction of the prednisolone dosage. Four months later, his visual acuity had improved to 0.9 in the right and 1.0 in the left eye.

The patient returned 8 months later with recurrent metamorphopsia in the right eye. At that time he had been using 75 mg of prednisolone per day for 3 weeks because of an exacerbation of his asthma. His visual acuity was 0.6 in the right eye and 1.0 in the left eye. Upon biomicroscopy with a fundus contact lens, a





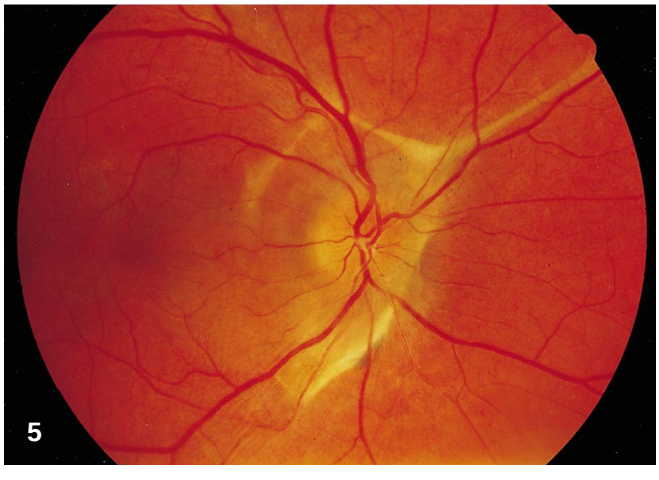


Fig. 3 Fundus of the right eye. During treatment with 60 mg prednisolone per day for 4 months a bullous serous retinal detachment had developed inferiorly

Fig. 4 Fundus of the right eye shows an exudative retinal detachment with yellowish subretinal serofibrinous exudate around the optic disc

Fig. 5 Right fundus 5 months after tapering the corticosteroid dosage to 20 mg. A narrow strand of subretinal fibrous tissue around the optic disc has developed. Note that the serous retinal detachment has flattened completely

slight increase in granularity of the pigment epithelium and yellow subretinal deposits were noted in the area of previous detachments. Fluorescein angiography of the right eye showed recurrent leakage of the four original pigment epithelial detachments and window defect-type hyperfluorescence above the optic disc. The fundus of the left eye was unchanged in the area of previous involvement. No new pigment epithelial detachments had developed. The prednisolone dosage was lowered to 40 mg/day. As a compensation, cyclosporin was administered. The patient's visual acuity remained unchanged. His asthma stabilized and cyclosporin was discontinued after 4 weeks. Three months later there was still some leakage of two pigment epithelial defects close to the macula of the right eye. The patient refused laser treatment of the leakage sites. Visual acuity stabilized at 0.6 in the right and 1.0 in the left eye. Two years after his initial complaints, the patient again noted decreased vision in his right eye. Four months previously the prednisolone dosage had been increased to 60 mg/day because of his asthma. Visual acuity was 0.2 in the right and 1.0 in the left eye. The anterior segments were unremarkable. In the fundus of the right eye there were multiple pigment epithelial detachments in the posterior pole and a bullous serous detachment of the retina inferiorly (Fig. 3). The subretinal fluid shifted and no retinal break was found. Subretinal fibrin was present around the optic disc. The left fundus remained stable with focal pigmentary abnormalities at the site of previous pigment epithelial detachment. The prednisolone dosage was tapered to 30 mg/day. Four weeks later there was a total exudative retinal detachment with partly organized, yellowish subretinal serofibrinous exudate around the optic disc (Fig. 4).

Subtle intraretinal hemorrhages were seen in the posterior pole. Maintenance treatment of 20 mg prednisolone per day was started. For the second time the patient refused laser treatment. During the next 5 months, the serous detachment flattened completely. The deposition of subretinal fibrin was followed by fibrotic scar formation (Fig. 5). A dense, narrow strand of subretinal fibrotic tissue tented up the retina slightly. Visual acuity improved to 0.4. Six months later the ocular findings were unchanged. Visual acuity has remained 1.0 in the left eye.

Discussion

The patient reported here developed CSCR while receiving high doses of corticosteroids systemically for severe asthma. The detachments resolved concurrent with tapering the dosage. Neither discontinuation of steroid therapy nor photocoagulation of leaking points were necessary to flatten the retina. The patient developed three successive attacks of CSCR. The first occurred after he had received 60 mg of prednisolone daily for 1 year. The second and third attacks came after he had received 75 mg of prednisolone daily for 3 weeks and 60 mg of prednisolone daily for 16 weeks respectively. In between times he received maintenance treatment of 35–40 mg prednisolone daily. After the third attack the prednisolone dosage had to be tapered to 20 mg to flatten the retina. This attack was much more severe than is typical for CSCR. A large bullous retinal detachment occurred with subretinal fibrin. The deposition of fibrin was followed by subretinal fibrosis and fibrotic scar formation.

Several reports have indicated that the use of high-dose corticosteroids systemically may be a risk factor for the development of CSCR. Wakakura and Ishikara reported two patients in whom CSCR developed unilaterally while they were receiving systemic corticosteroids for retrobulbar neuritis [12]. Both patients had a history of CSCR in the affected eye. A reduction in the steroid dosage resulted in spontaneous recovery. Polak and colleagues reported the development of CSCR in 34 patients from 10 European eye clinics during systemic treatment with corticosteroids because of several different systemic disorders or after allotransplantation [8]. The serous detachments resolved after discontinuation of the treatment or decrease of the daily steroid dosage. Bullous inferior retinal detachment and subretinal fibrotic scar formation were not observed in this series.

The cause of idiopathic CSCR is still unknown. The disease is probably linked to a breakdown in the permeability of the choriocapillaris in one or more focal areas of the posterior fundus. This allows for passage of leaked choroidal fluid across the RPE, producing retinal elevations [4]. The apparent adverse reaction to systemic corticosteroid therapy by some patients with idiopathic CSCR suggests that increased plasma levels of cortisol may be responsible for aggravating the hyperpermeability of the choriocapillaris. It is understandable that steroids can prolong or worsen the disease, because cortisol delays and inhibits endothelial repair [14]. A possible effect of elevated plasma cortisol levels is suggested in reports describing the development of CSCR during pregnancy with a spontaneous recovery after delivery or abortion [5, 10]. Experimental CSCR with injections of steroids in monkeys supports this approach [15].

In the present case without a history of previous serous retinal detachments, the most severe form of CSCR with subretinal fibrotic scar formation developed. This is a very unusual manifestation in CSCR. When fibrin is present it disappears in nearly all cases. Gass and Little described the development of subretinal fibrosis in two men treated with high-dose corticosteroids systemically for atypical manifestations of CSCR [6]. Fibrotic scar formation has not been described in CSCR developing during corticosteroid treatment. Recognition of the atypical manifestations of CSCR, which may masquerade as rhegmatogenous retinal detachment, Harada disease, uveal effusion, multifocal choroiditis, metastatic carcinoma or lymphoma, is important in order to avoid the use of systemic corticosteroid treatment. Several systemic disease, however, require high-dose corticosteroids, and this treatment may be life saving. In many patients, therefore, like the man reported here, discontinuation or reduction of corticosteroid treatment is contraindicated. These patients run the risk of developing CSCR, even a severe form with permanent loss of vision.

References

1. Brancato R, Scialdone A, Pece A, Coscas G, Binaghi M (1987) Eight-year follow-up of central serous chorioretinopathy with and without laser treatment. Graefe's Arch Clin Exp Ophthalmol 225:166–168
2. Friberg TR, Eller AW (1990) Serous retinal detachment resembling central serous chorioretinopathy following organ transplantation. Graefe's Arch Clin Exp Ophthalmol 228:305–309
3. Gass JDM (1973) Bullous retinal detachment. An unusual manifestation of idiopathic serous choroidopathy. Am J Ophthalmol 75:810–821
4. Gass JDM (1986) Stereoscopic atlas of macular diseases: diagnosis and treatment, 3rd edn, vol 1, Mosby, St Louis, pp 45–59
5. Gass JDM (1991) Central serous chorioretinopathy and white subretinal exudation during pregnancy. Arch Ophthalmol 109:677–681
6. Gass JDM, Little H (1995) Bilateral bullous exudative retinal detachment complicating idiopathic central serous chorioretinopathy during systemic corticosteroid therapy. Ophthalmology 102:737–747
7. Haimovici R, Gragoudas ES, Duker JS, Sjaarda RN, Eliott D (1997) Central serous chorioretinopathy associated with inhaled or intranasal corticosteroids. Ophthalmology 104:1653–1660
8. Polak BCP, Baarsma GS, Snyers B (1995) Diffuse retinal pigment epitheliopathy complicating systemic corticosteroid treatment. Br J Ophthalmol 79:922–925
9. Schatz H, McDonald R, Johnson RN, Chan CK, Irivine AR, Berger AR, Folk JC, Robertson DM (1995) Subretinal fibrosis in central serous chorioretinopathy. Ophthalmology 102:1077–1088
10. Sunnes JS, Haller JA, Fine SL (1993) Central serous chorioretinopathy and pregnancy. Arch Ophthalmol 111:360–364
11. Tsukahara I, Uyama M (1978) Central serous choroidopathy with bullous retinal detachment. Graefe's Arch Clin Exp Ophthalmol 206:169–178
12. Wakakura M, Ishikawa S (1984) Central serous chorioretinopathy complicating systemic corticosteroid treatment. Br J Ophthalmol 68:329–331
13. Yannuzzi LA (1986) Type A behavior and central serous chorioretinopathy. Trans Am Ophthalmol Soc 84:799–845
14. Yoshioka H, Katsume Y, Akune H (1981) Experimental central serous chorioretinopathy. I. Clinical findings. Jpn J Ophthalmol 25:112–118
15. Yoshioka H, Katsume Y, Akume H (1982) Experimental central serous chorioretinopathy in monkey eyes: fluorescein angiographic findings. Ophthalmologica 185:168–178