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# Choroidopathy with serous retinal detachment in a patient with polymyositis

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PURPOSE. *The authors report a unique case of multifocal choroidopathy in association with polymyositis.*
METHODS. *The clinical presentation, laser treatment, and subsequent changes in ocular findings are described.*
RESULTS. *A 65-year-old Japanese man with polymyositis experienced mild acute vision impairment in both eyes during systemic corticosteroid treatment for interstitial pneumonia. Fundus examination revealed a gray-white subretinal exudate with serous retinal detachment. Angiographic examination disclosed partial dilatation of the choroidal vein at the posterior pole and dye leakage points in the retinal pigment epithelium. The patient's left eye developed bullous retinal detachment and multifocal choroidal nodules at the posterior pole during the period of tapering of oral corticosteroid administration. Retinal burns were applied to the dye leakage points in the right eye using an argon laser. Vision in the right eye showed rapid recovery due to regression of the serous retinal detachment. However, in the left eye without retinal burns visual acuity decreased to light perception only even after regression of the bullous retinal detachment and choroidopathy.*
CONCLUSIONS. *In serous retinal detachment with choroidopathy associated with polymyositis, laser photocoagulation at leakage points may be useful for rapid restoration of vision and for the prevention of permanent severe vision loss. (Eur J Ophthalmol 2006; 16:318-25)*

KEY WORDS. *Choroidopathy, Corticosteroid, Laser photocoagulation, Polymyositis*

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## INTRODUCTION

Polymyositis is a collagen vascular disease with an autoimmune background and involves striated muscles. When skin lesions are found in addition to muscle involvement, the condition is called dermatomyositis. Posterior ocular manifestations, such as cotton wool spots, occur occasionally in patients with either polymyositis or dermatomyositis (1-6), whereas choroidopathy with serous detachment of the retina has not been reported previously.

Choroidopathy with serous retinal detachment was seen in a patient with polymyositis and laser treatment resulted in successful resolution of the serous detachment and recovery of visual acuity in one eye. In the other eye without laser treatment, however, bullous exudative retinal detachment and multifocal choroidal nodules developed and resulted in severe vision impairment.

## Case report

A 65-year-old Japanese man with a 2-year history of polymyositis was admitted to the rheumatic disease unit of our hospital because of deterioration of interstitial pneumonia on December 21, 2002.

The patient presented with proximal muscle weakness, arthralgia of the wrists and knees, and lung fibrosis on

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plain chest x-ray film. Serologic examination showed the patient to be positive for antinuclear antibody. Creatine phosphokinase (CPK) level was markedly elevated at 2658 IU/L. The patient was treated with 300 mg/day of cyclosporin A, pulsed intravenous methylprednisolone (1000 mg for 3 days), and subsequent oral prednisolone at a dose of 55 mg/day to be reduced gradually by 5 mg every 4 weeks. During the subsequent 4 weeks of observation, the patient's symptoms improved with a reduction in CPK level to 273 IU/L.

However, the patient reported blurred vision in both eyes for 1 week at referral to our clinic on January 18, 2003. On ophthalmologic testing, his best-corrected visual acuity was 20/20 and intraocular pressure was 12 mm Hg in both eyes. The anterior segments were unremarkable. Both eyes had focal shallow serous elevations of the sensory retina without cotton wool spots or hemorrhages, and gray-white subretinal exudates were observed in the serous detachment (Fig. 1, A and B). Fluorescein angiography (FA) indicated early leakages at the subretinal exudates in both eyes and a faint granular hyperfluorescent lesion in the left eye (Fig. 1, C and D).

Indocyanine green angiography (ICGA) of the right eye showed partial dilatation of the choroidal vein underlying the site of dye leakage revealed by FA and showed extended patchy choroidal hypofluorescent lesions underlying the faint granular yperfluorescent lesions seen by FA in the left eye (Fig. 1, E and F).

Considering his general condition, cyclosporin A was continued and oral prednisolone treatment was reduced to 35 mg/day on April 22, 2003.

The areas of serous detachment increased, while the visual acuity decreased to 20/25 in the right and 20/40 in the left. Gray-white lesions were enlarged and newly appeared in the right eye, and bullous exudative retinal detachment and multifocal disciform subretinal nodules that looked like accumulation of fibrinous material appeared (Fig. 2, A and B).

FA examination showed multiple leakages and hypofluorescent lesions in the right eye and multiple disciform hypofluorescent lesions corresponding to the nodules and a large non-perfusion area in the inferior equatorial area of the fundus in the left eye (Figs. 2, C and D, and 3, A and B). ICGA examination also revealed hyper- or hypofluorescent lesions of the choriocapillaris in the right eye and disciform hypofluorescent lesions corresponding to the multiple nodules, indicating that these nodules in the left eye were choroidal lesions (Fig. 2, E and F).

Due to the decreases in visual acuity of 20/30 in the right eye and 20/400 with total bullous retinal detachment in the left eye and apparent lack of effect of systemic corticosteroid therapy on the retina, laser photocoagulation (argon laser 150 mW, 0.2 seconds, 150 μm spot size, 50 spots) was performed on the point of dye leakage in the right eye to prevent the development of bullous retinal detachment on June 9, 2003.

Laser photocoagulation could not be applied to the left eye due to total retinal detachment. Two weeks later, serous detachment had regressed and the corrected visual acuity of the right eye was restored to 20/20, whereas the vision of the left eye decreased to light perception with no change of bullous retinal detachment.

Six months later, the systemic condition was controlled with oral prednisolone (30 mg/day) and cyclosporin A (200 mg/day). Visual acuity remained at 20/20 in the right eye and light perception only in the left eye.

Fundus examination revealed that the gray-white lesions and serous detachment had disappeared in the right eye (Fig. 4A), and the bullous exudative retinal detachment and multifocal disciform subretinal nodules were resolved gradually, whereas subretinal fibrosis had newly appeared in the left eye (Fig. 4B).

FA showed small lesions of granular faint hyperfluorescence indicating retinal pigment epithelium (RPE) impairment in the right eye (Fig. 4C) and extended granular hyperfluorescence and faint dye leakage from the optic nerve head in the left eye (Figs. 3, C and D, and 4D). ICGA revealed patchy hypofluorescent lesions of the choriocapillaris in both eyes and disciform hypofluorescent lesions had diminished in the left eye (Fig. 4, E and F).

The results of computed tomography of the head were normal. T1-weighted magnetic resonance imaging (MRI) in the coronal plane showed a reduction in size of the left side of the optic chiasm as compared to the right side (Fig. 5).

## DISCUSSION

Choroidopathy, with or without serous retinal detachment in association with systemic lupus erythematosus (SLE), is a rare collagen vascular disease (7-15). To our knowledge, this is the first report of a patient with polymyositis developing choroidopathy and bullous exudative retinal detachment resulting in severe visual loss.

All of the cases of SLE reported previously had been re-

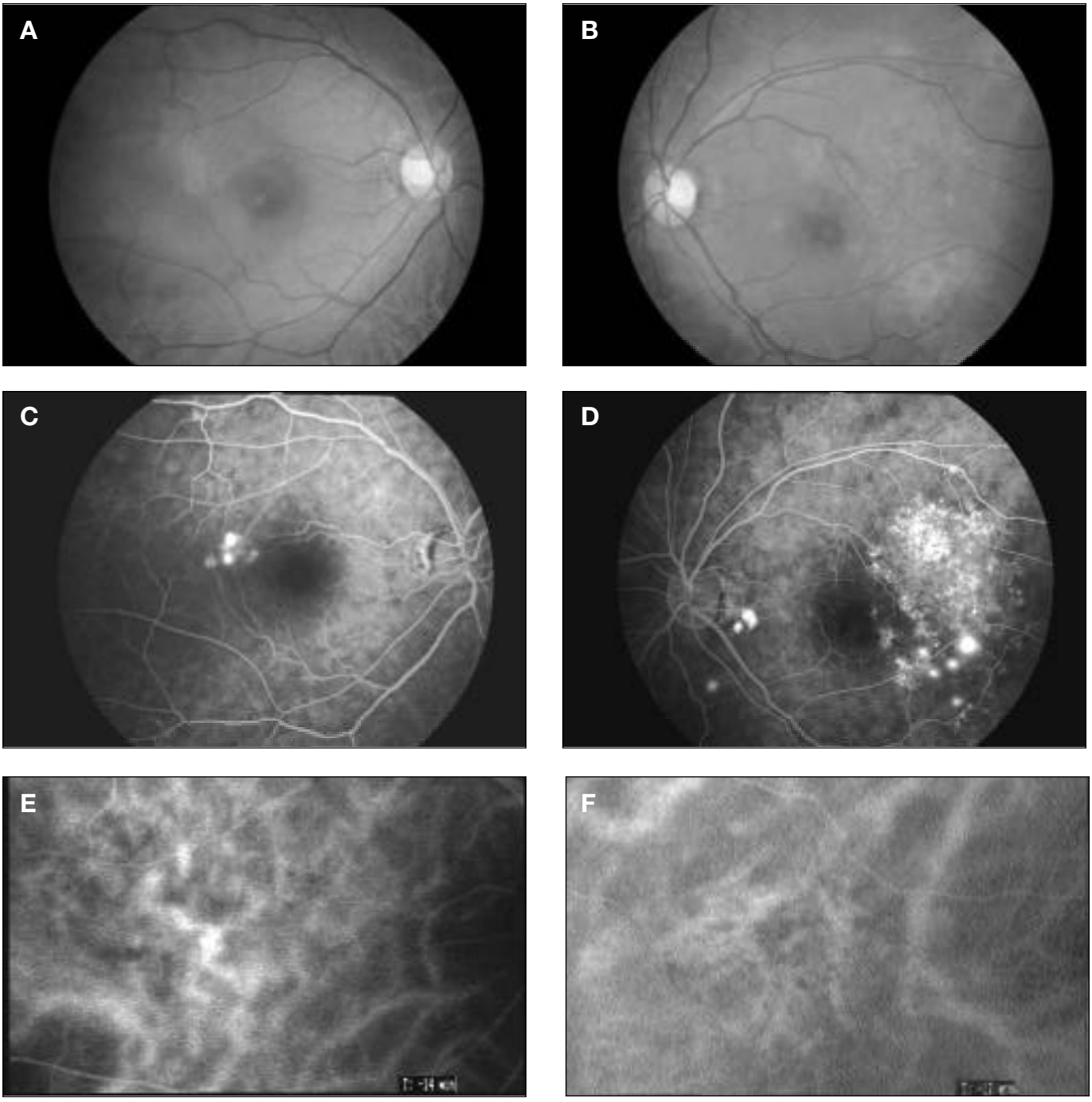


**Fig. 1 -** *Fundus photographs of the left eye (***A***) and right eye (***B***) of a 65-year-old man show a gray-white subretinal exudate. Fluorescein angiography (FA) showed multiple leakages from the gray-white subretinal exudate in both eyes and faint granular hyperfluorescence beneath the large retinal serous detachment in the left eye (***C***, **D***). Indocyanine green angiography of the right eye showed partial dilatation of the choroidal vein underlying the sites of dye leakage observed by FA (***E***) and showed extended patchy choroidal hypofluorescent lesions underlying faint granular hyperfluorescent lesions detected by FA in the left eye (***F***).*

ceiving treatment with systemic steroid therapy for systemic conditions. The case of polymyositis reported here had also been receiving systemic corticosteroid therapy for interstitial pneumonia. There is evidence that the systemic application of corticosteroids can result in choroidal hyperpermeability, which may lead to a breakdown of the blood-retinal barrier or alter RPE ion transport capabilities, and cause serous retinal detachment (16-18). How-

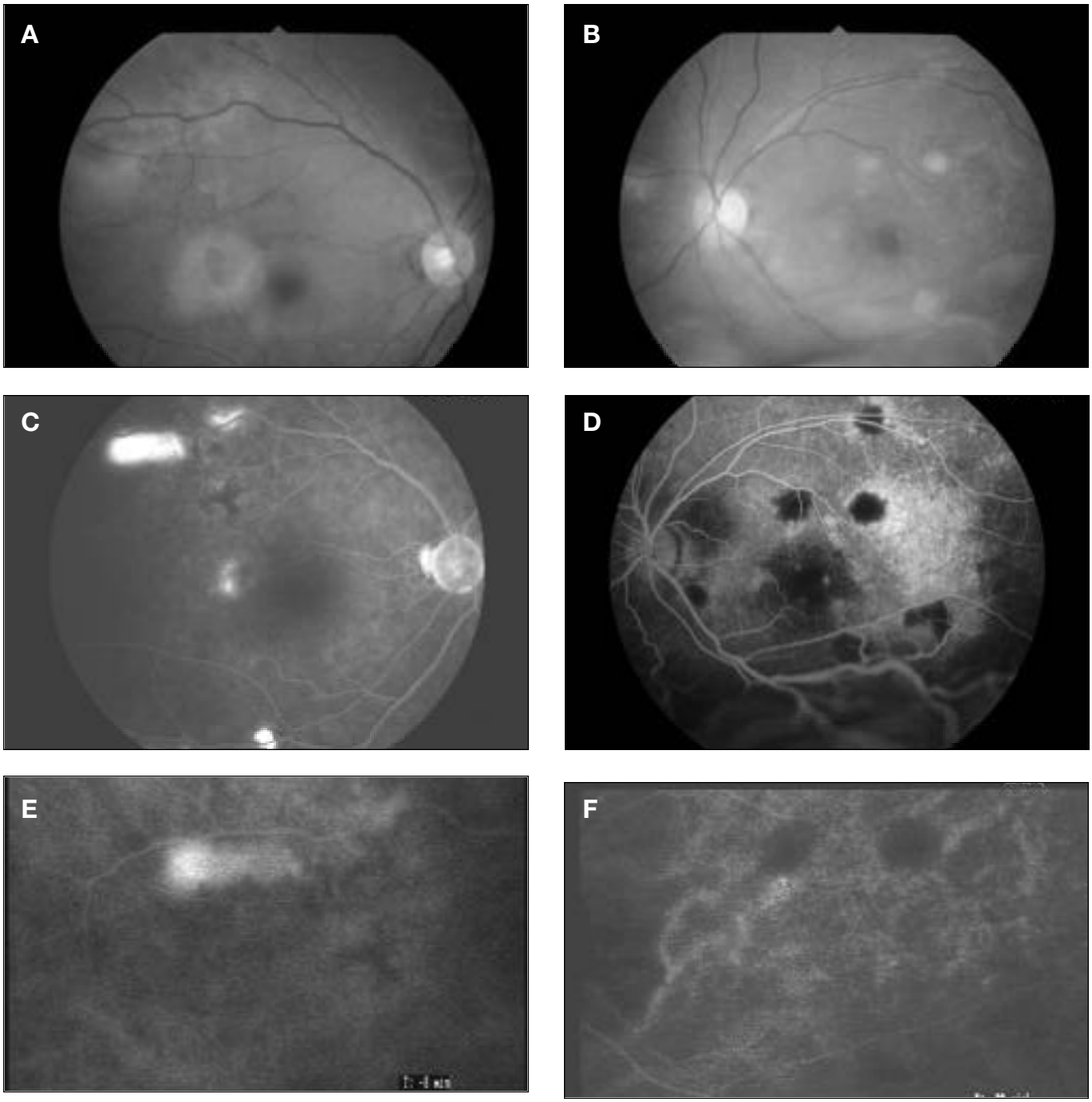


**Fig. 2 -** *Three months after initial presentation, gray-white lesions were enlarged and newly appeared in the right eye (**A**). Fundus photograph of the left eye shows bullous exudative retinal detachment and multifocal disciform subretinal nodules (**B**). Fluorescein angiography showed multiple leakages and small hypofluorescent lesions in the right eye (**C**) and multiple disciform hypofluorescent lesions corresponding to the nodules and non-perfusion area in the inferior equatorial area of the fundus in the left eye (**D**). Indocyanine green angiography also revealed hyper- or hypofluorescent lesions of the choriocapillaris in the right eye (**E**) and disciform hypofluorescent lesions corresponding to multiple nodules indicating that the nodules were choroidal lesions in the left eye (**F**).*

ever, there have been few reports regarding steroid-induced chorioretinopathy during the clinical course of collagen vascular diseases (15). Some of the cases of choroidopathy reported previously could have been associated with systemic steroid therapy as well as the collagen vascular disease itself, and steroid-induced choroidopathy must also be taken into consideration in the differential diagnosis in our case.

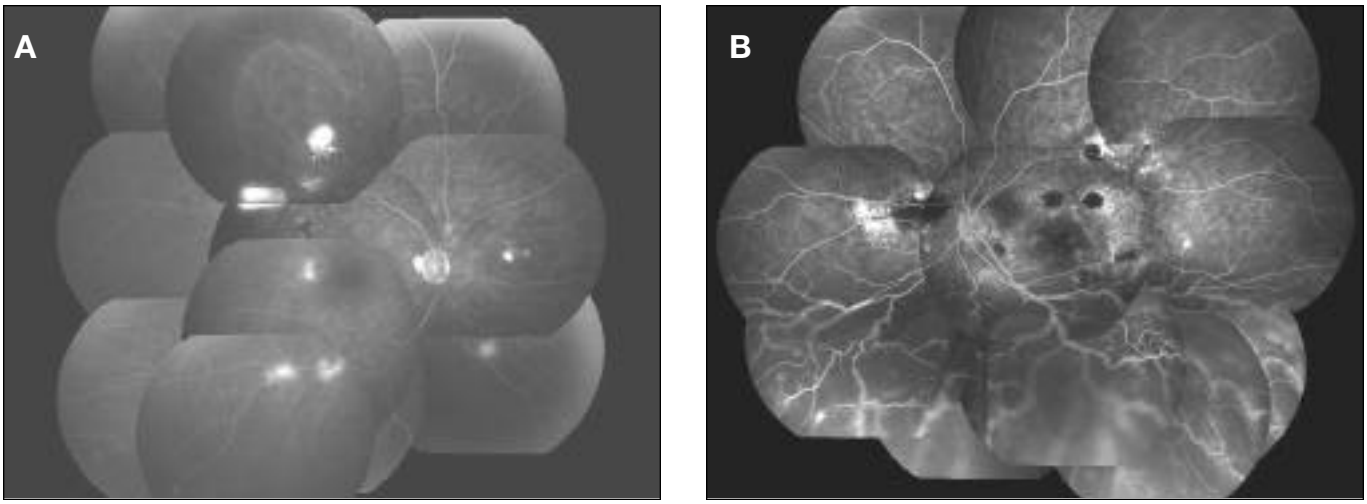


**Fig. 3 -** *Three months after initial presentation, fluorescein angiography (FA) showed many active leakage points at midperipheral fundus without bullous retinal detachment in the right eye (***A)***, whereas FA did not show obviously an active dye leakage point that could explain the cause of progressive bullous retinal detachment in the left eye (***B)***. Only faint granular hyperfluorescence around the nodules was shown in the left eye.*

On first presentation, we believed that the ocular manifestations in our patient might have been a side effect of systemic corticosteroid therapy due to a history of high-dose systemic corticosteroid therapy, as reported previously (16-21), rather than being induced by choroidopathy associated with polymyositis. The ocular manifestations, i.e., serous retinal detachment with gray-white subretinal exudate, multiple fluorescein dye leakage, dilatation of the choroidal vein underlying fluorescein dye leakage sites, mimicking multifocal posterior pigment epitheliopathy (MPPE) (22) or bullous retinal detachment as unusual manifestations of central serous chorioretinopathy (CSC) (23), were consistent with previous reports (16-21). The granular hyperfluorescence seen by FA in the left eye at the first presentation, however, may represent atrophic changes of the RPE due to previous recurrent-chronic CSC. Thus, the chronic choroidopathy associated with polymyositis prior to the systemic corticosteroid therapy should be also considered as another interpretation of the fundus manifestation.

Initially, we could only observe the fundus of the patient during the period of tapering of systemic corticosteroid therapy according to the patient's systemic condition in the rheumatic disease unit of our hospital. Systemic steroid therapy is thought to have two opposite effects during the clinical course of collagen vascular diseases. On the one hand, it aids in the resolution of choroidopathy by reducing the inflammation of systemic vasculitis. However, it also accelerates breakdown of the blood-retinal barrier and leads to serous retinal detachment. In the present case, during the reduction of oral prednisolone from 55 to 35 mg/day, the ocular manifestations showed further deterioration. At this time, it was not possible to determine whether the corticosteroid was beneficial in this case because it could not be discontinued completely because of the patient's systemic condition.

However, it is clear that the decrease in visual acuity was mainly due to serous retinal detachment in the right eye, because laser treatment, which is effective in some cases of steroid-induced CSC (19), resulted in rapid regression of the serous detachment and subsequent improvement of visual acuity. Therefore, it was possible that MPPE or unusual CSC induced by systemic corticosteroid therapy was the cause of the retinal detachment in the right eye. Bullous retinal detachment in the left eye could also be explained as a complication of systemic corticosteroid therapy, because there is evidence that bullous exudative retinal detachment may complicate idiopathic CSC during systemic corticosteroid therapy (16). However, the gray-white retinal exudation and fluorescein leakage point disappeared in the left eye at that time, whereas multifocal choroidal nodules with disciform hypofluorescence appeared. In addition to total bullous retinal detachment, this made us hesitant to apply laser treatment to the left eye.

This unique disciform choroidal nodule has not been documented previously in a patient with either CSC or SLE choroidopathy. Multiple disciform choroidal nodules, possibly due to multifocal choroidopathy associated with polymyositis, were resolved with regression of the bullous

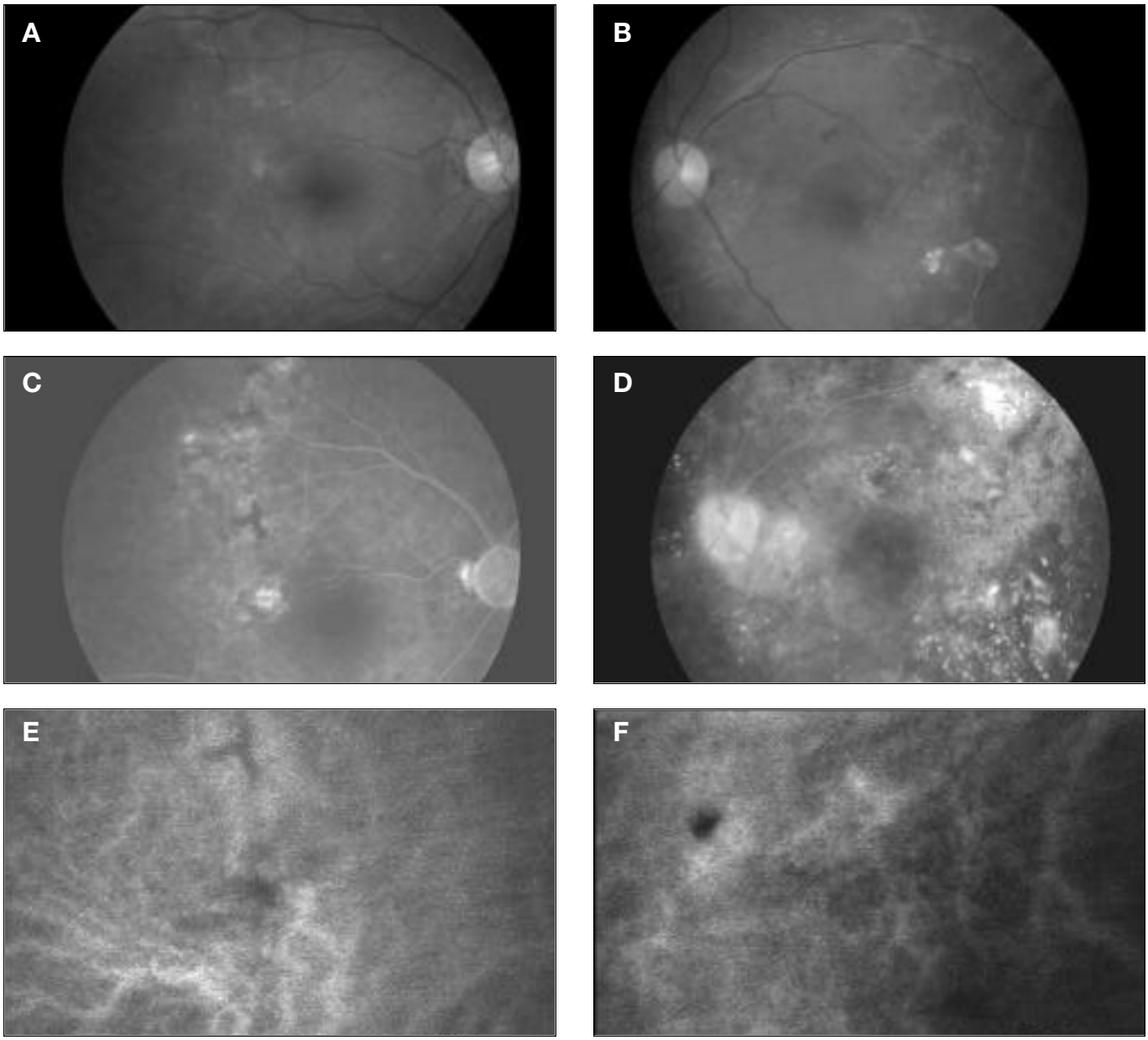


**Fig. 4 -** *Six months after laser treatment, gray-white lesions and serous detachment disappeared in the right eye (***A***), and the bullous exudative retinal detachment and multifocal disciform subretinal nodules were also resolved, whereas subretinal fibrosis appeared in the left eye (***B***). Fluorescein angiography showed small lesions with granular faint hyperfluorescence indicating deterioration of the retinal pigment epithelium in the right eye (***C***) and extended granular hyperfluorescence and faint dye leakage from the optic nerve head in the left eye (***D***). Indocyanine green angiography revealed patchy hypofluorescent lesions of choriocapillaris in both eyes and disciform hypofluorescent lesions had diminished in the left eye (***E, F***).*

retinal detachment even with continuing systemic corticosteroid therapy. As suggested previously by Wakakura et al (19), corticosteroid administration was a predisposing factor in our case, rather than a direct cause, for the disruption of the outer blood-retinal barrier. Therefore, we suspected that the focal inflammatory lesion itself was the cause of the bullous retinal detachment in the left eye, and not widespread choroidal vasculopathy induced by systemic corticosteroid therapy.

Finally, even after resolution of bullous retinal detachment, the visual acuity in the left eye has remained as on-

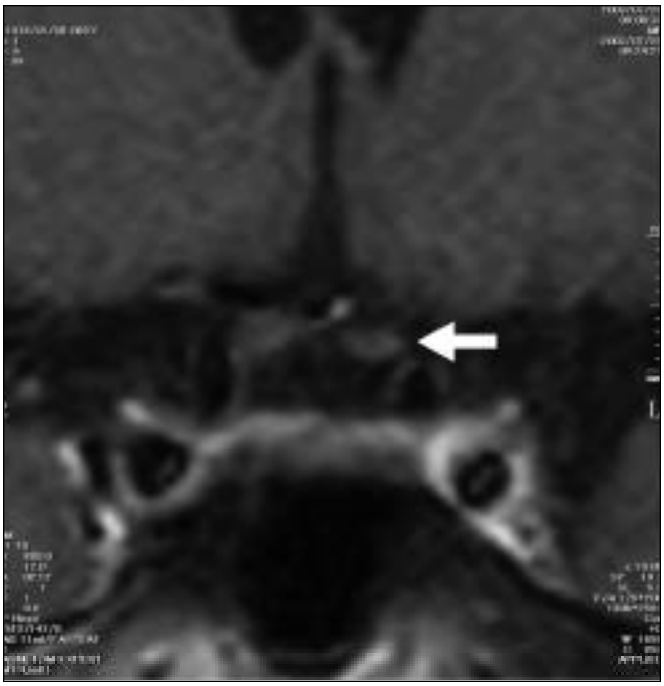

**Fig. 5 -** *T1-weighted magnetic resonance imaging in the coronal plane showed that the left side of the optic chiasm (arrow) was reduced in size as compared to the right.*

ly light perception, whereas that in the right eye was restored. There have been several reports of patients with CSC associated with or without SLE who developed subretinal fibrosis similar to that seen in the left eye in our case (12, 24-26). Schatz et al (25) reported six cases of subretinal fibrosis in CSC. Final visual acuities of three out of the six patients were less than counting fingers with some deterioration of the macula (i.e., submacular fibrosis, subretinal neovascularization, telangiectatic changes). From the results of fundus examination of our case, it was difficult to explain the severe vision impairment in the left eye. However, FA showed faint dye leakage from the optic nerve head in the left eye.

From this finding, optic neuritis, which is rare but could be associated with collagen vascular diseases, such as SLE (27-29), should also be taken into consideration in diagnosis. MRI findings also indicated optic chiasm atrophy in the left side, although the optic nerve head still appeared healthy in both eyes.

Systemic corticosteroid administration might play a significant role in the development of serous retinal detachment in choroidal inflammatory background, although the precise pathogenetic mechanism of choroidopathy associated with collagen vascular diseases remains unclear. However, the present case of polymyositis, showing multifocal choroidopathy in the left eye, indicated the presence of focal inflammation at the choriocapillaris. Therefore, the usefulness of focal laser treatment, which would be applied to sites of choroidal inflammation, awaits further investigation.

*The author has no proprietary interest in any aspect of this article.*

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