Surgical Management of Bilateral Exudative Retinal Detachment Associated with Central Serous Chorioretinopathy

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Purpose: To report a case of bilateral bullous exudative retinal detachment in central serous chorioretinopathy (CSC) which was attached by vitrectomy and internal drainage of the subretinal fluid.
Methods: A 47-year-old man affected by bilateral atypical CSC with a bullous retinal detachment with subretinal exudate. A fluorescein angiogram (FAG) showed multiple points of leakage and staining of subretinal fibrosis. A tentative diagnosis of Vogt-Koyanagi-Harada (VKH) syndrome was made and the patient was treated with systemic corticosteroids and immunosuppressive agents. However, the subretinal fluid was not absorbed. He was then treated with vitrectomy and internal drainage of subretinal fluid.
Results: The retina was attached successfully in both eyes. Visual acuity improved to 20/50 in his left eye but did not improve in the right eye due to subretinal fibrotic scarring and atropic changes on the macula.
Conclusions: Our case suggests that the surgical management of bullous exudative retinal detachment is safe and necessary. *Korean Journal of Ophthalmology 20(2):131-138, 2006*

Key Words: Bullous retinal detachment, Central serous chorioretinopathy, Subretinal fluid drainage, Vitrectomy

In typical central serous chorioretinopathy (CSC), serous retinal detachment usually resolves spontaneously with the return of visual function. If it persists, laser treatment to focal leaks outside the foveolar area is effective in resolving the detachment.[1-8] On rare occasions, a bullous exudative RD, usually associated with multiple large retinal pigment epithelial detachments there are often hidden beneath a cloudy subretinal serofibirnous exudate, may occur in patients with CSC.[4-20] This atypical CSC may be misdiagnosed as Vogt-Koyanagi-Harada (VKH) syndrome, uveal effusion, multifocal choroiditis, metastatic carcinoma, or lymphoma because of the combined bullous retinal detachment (RD).

It is very unusual for the deposition of fibrin to be followed by fibrotic scar formation.[4,9-12,15-17,19-20] So far, 25 cases have been reported in the Korean literature in which bullous exudative RD developed spontaneously in association with CSC; only one of them presented with subretinal fibrotic band formation with permanent visual loss.[4-6,19]

Here, we report a case of progressive bilateral bullous RD and subretinal fibrosis associated with atypical CSC which had been treated by vitrectomy and internal drainage of subretinal fluid.

Case Report

A 47-year-old man who had experienced a decrease in visual acuity and a visual field defect at the superotemporal side of his right eye visited our clinic. He did not complain of neurologic symptoms, such as headaches, tinnitus, or dysacusia. His family history and past history were unremarkable. His visual acuity was 20/40 in the right eye and 20/20 in the left eye. While the left anterior segment was normal, there were few pigmented cells in the right anterior chamber and vitreous cavity. Fundus examination of the right eye revealed subretinal fibrosis, a retinal fold, a pigmented lesion on the posterior pole (Fig. 1A), and a bullous RD which shifted to the most dependent location. There were multiple alterations of the RPE in the left eye (Fig. 1B). A fluorescein angiogram (FAG) of the right eye revealed multiple points of dye leakage and pooling into the subretinal space, as well as variable blockage and staining of the areas of subretinal fibrosis (Fig. 1C). In addition, several pinpoint leakages were found in his left eye: this was not accompanied

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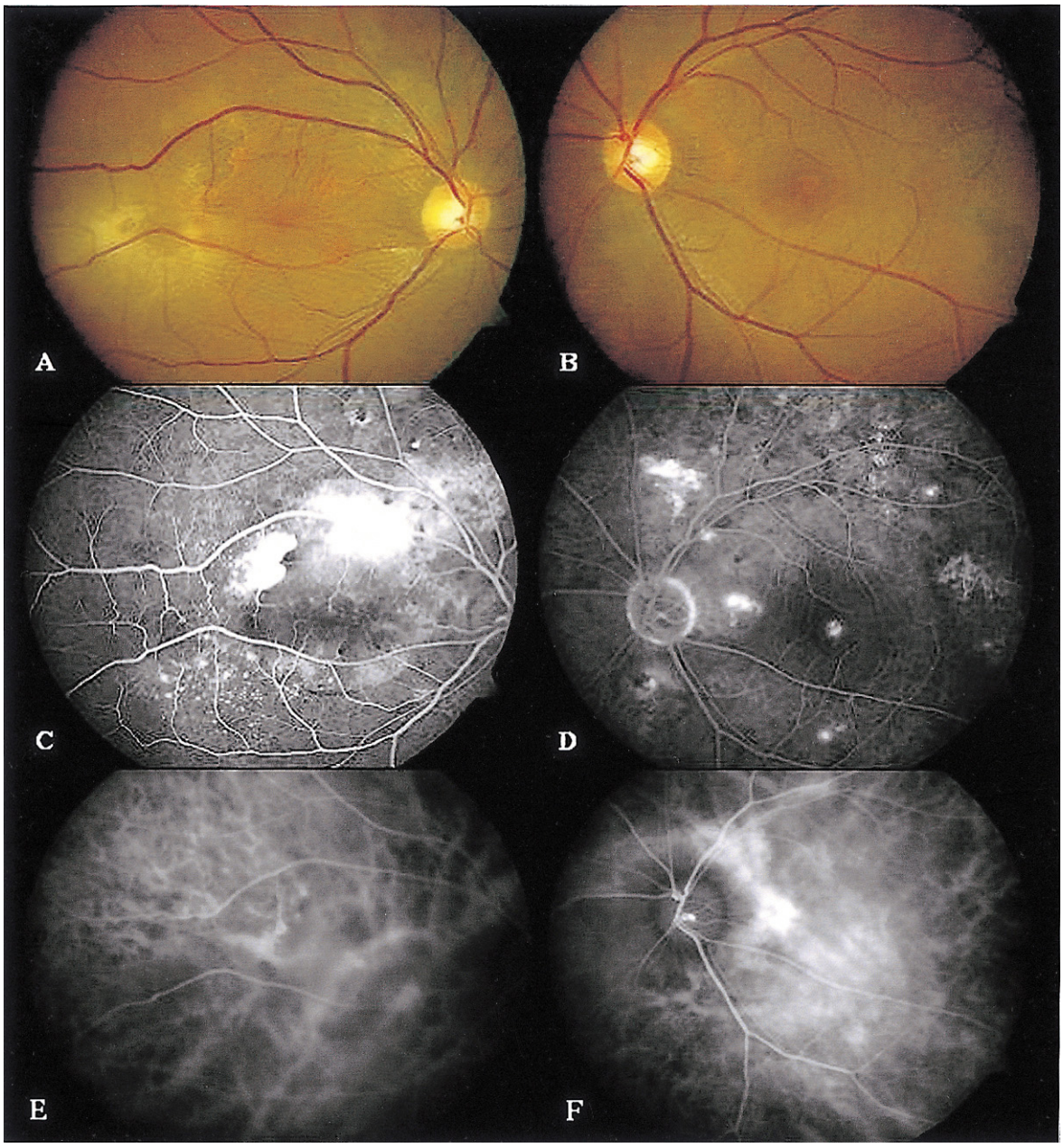

Fig. 1. Color fundus photographs (A, B), fluorescein angiograms (C, D), and indocyanine green angiogram (E, F) before systemic steroid therapy. (A) Right eye. Subretinal fibrosis, retinal fold and pigmented lesion on the posterior pole. (B) Left eye. RPE changes in the macular region and around the vascular arcade. (C) Right eye. Multiple points of dye leakage and pooling into the subretinal space, and variable blockage and staining of areas of subretinal fibrosis. (D) Left eye. Multiple pinpoint leakages. (E) Right eye and (F) left eye, showing multiple choroidal vascular hyperpermeability and focal staining of pigment epithelium.

by exudative detachment (Fig. 1D). An indocyanine green angiogram (ICGA) of both eyes revealed multiple areas of choroidal vascular hyperpermeability and pigment epithelial focal staining (Fig. 1E, F). Laboratory evaluations were unremarkable. A tentative diagnosis of VKH syndrome was made, and we prescribed oral prednisolone (60 mg daily). During the 2-week follow-up, the visual acuity in his right eye decreased to 20/200, and repeated examinations showed worsening of the exudative retinal detachment and subretinal fibrosis, as well as development of a pigment epithelial detachment (PED) in the macular region (Fig. 2A, C, E). In addition to that, visual acuity in the left eye also decreased to 20/30, and multiple localized PEDs were detected in the left eye by fundus examination (Fig. 2B, D, F). At the same time, human leukocyte antigen (HLA) DQB1*0303 (DQ9) was detected as a result of HLA-DQ serologic subtypings. As such, we were more convinced regarding the diagnosis of VKH syndrome from this. Therefore, oral cyclosporine A

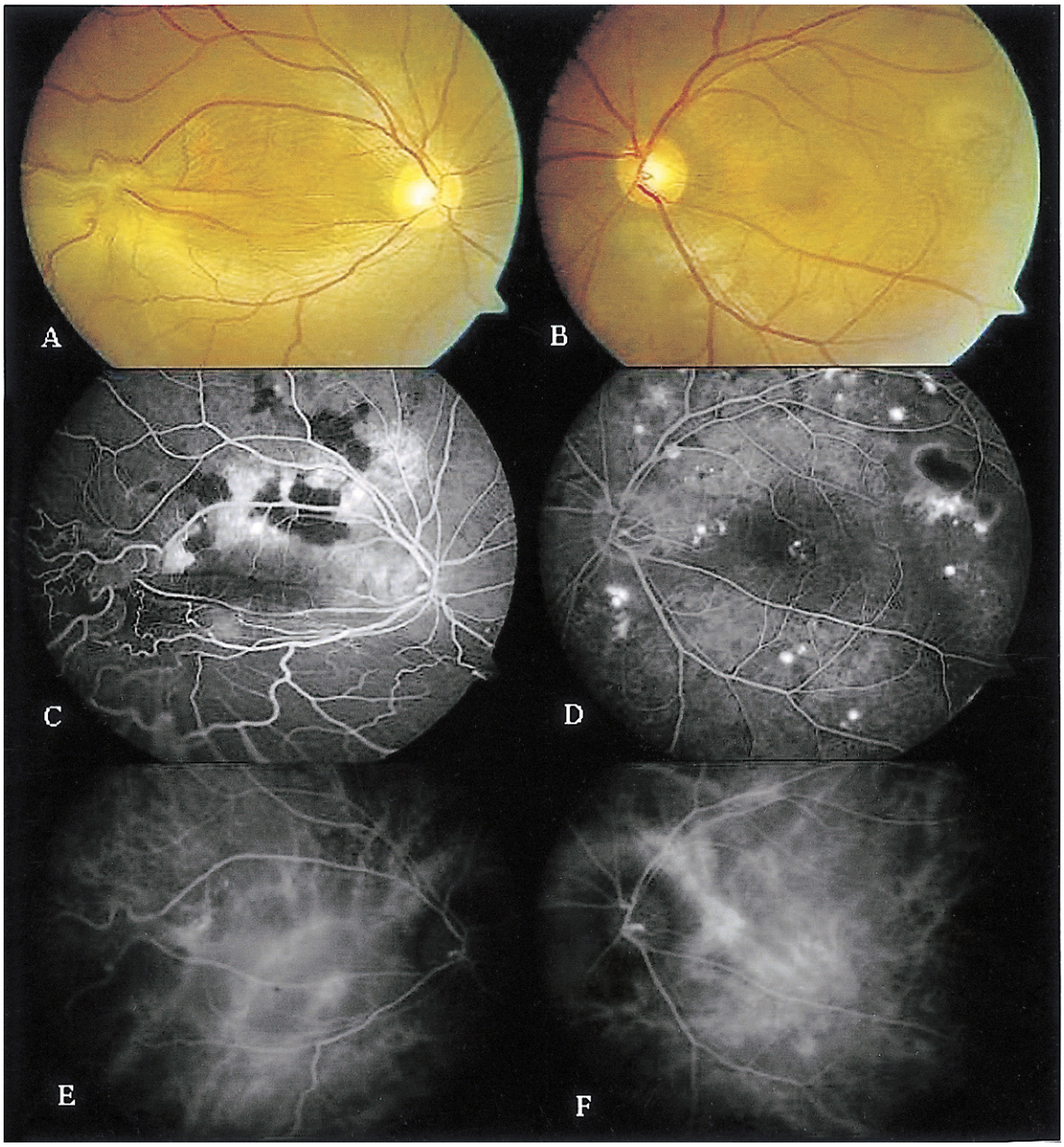

Fig. 2. Color fundus photographs (A, B), fluorescein angiograms (C, D), and indocyanine green angiogram (E, F) two weeks after systemic steroid therapy. (A) Right eye. Progressed exudative retinal detachment ,subretinal fibrosis, retinal fold and pigmented lesion on the posterior pole. (B) Left eye. Multiple localized PEDs. (C) Right eye. Multiple points of dye leakage and pooling into the subretinal space, and variable blockage and staining of areas of subretinal fibrosis. (D) Left eye. Multiple pinpoint leakages. (E) Right eye. (F) Left eye, multiple choroidal vascular hyperpermeability and focal staining of the pigment epithelium.

(100 mg daily) was added to the prednisolone.

On the second day of combination therapy, inferior RD extended to the inside of the inferior vascular arcade in the right eye and the disease seemed to have progressed in both eyes. Therefore, laser photocoagulation treatment to the leakage points was done on two occasions in both eyes. During the next 2 weeks, his visual acuity stabilized at 20/125 in the right eye and 20/25 in the left, and the fundus in both eyes remained stable with focal pigmentary abnormalities at the sites of the previous PED.

During next 2 weeks, the visual acuity in the right eye deteriorated to hand motions, but that of the left eye was unchanged. In the right eye, the exudative RD extended to the fovea and a subretinal fibrotic band grew across the macula (Fig. 3A, C, E). In the left eye, multiple exudative flecks appeared on the posterior pole (Fig. 3B, D, F). B-scan ultrasonography of the right eye revealed an inferior bullous RD with a shifting of subretinal fluid (Fig. 4). We stopped

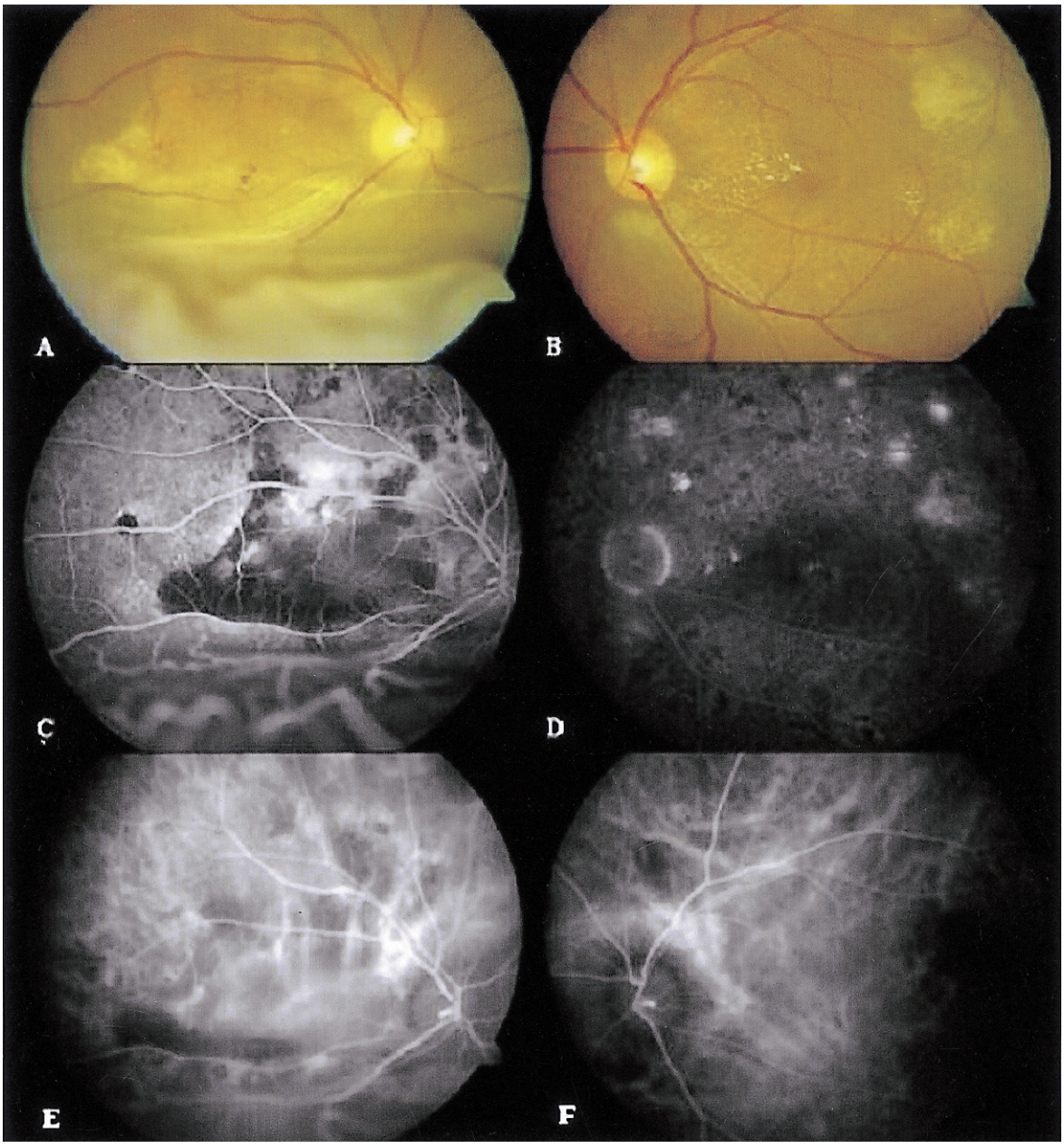

Fig. 3. Color fundus photographs (A, B), fluorescein angiograms (C, D), and indocyanine green angiogram (E, F) at the time that oral steroid and immunosuppresive treatment were stopped. (A) Right eye. Inferior bullous exudative RD, subretinal fibrotic band, fixed retinal fold, and subtle intraretinal hemorrhages. (B) Left eye. Multiple PEDs and exudative flecks. (C) Right eye. Large blockage of an area of exudative RD, variable blockage and staining of areas of subretinal fibrosis, and leakage and pooling into the subretinal space. (D) Left eye. Multiple pinpoint leakages and poolings. (E) Right eye. (F) left eye. Multiple choroidal vascular hyperpermeability and focal staining of the pigment epithelium.

oral corticosteroid and cyclosporine treatment and considered surgical management for the right eye. Two months after his first visit to our clinic, transscleral drainage of subretinal fluid, gas injection through the pars plana, radial scleral buckling, and focal laser photocoagulation were performed on the right eye. One month after the operation, fundus examination showed diminution of the exudative RD in the right eye and the new development of an exudative RD inferiorly in the left eye. His visual acuity was hand motions in the right eye and 20/40 in the left.

Six weeks after surgery, the retina was attached with subretinal proliferation in the fovea of the right eye (Fig. 5A, C); in the left eye, the inferior exudative RD, multiple serous PEDs, and exudative flecks had all worsened (Fig. 4B, D). The visual acuity in the left eye decreased gradually, eventually becoming 20/200. Two months after his first visit

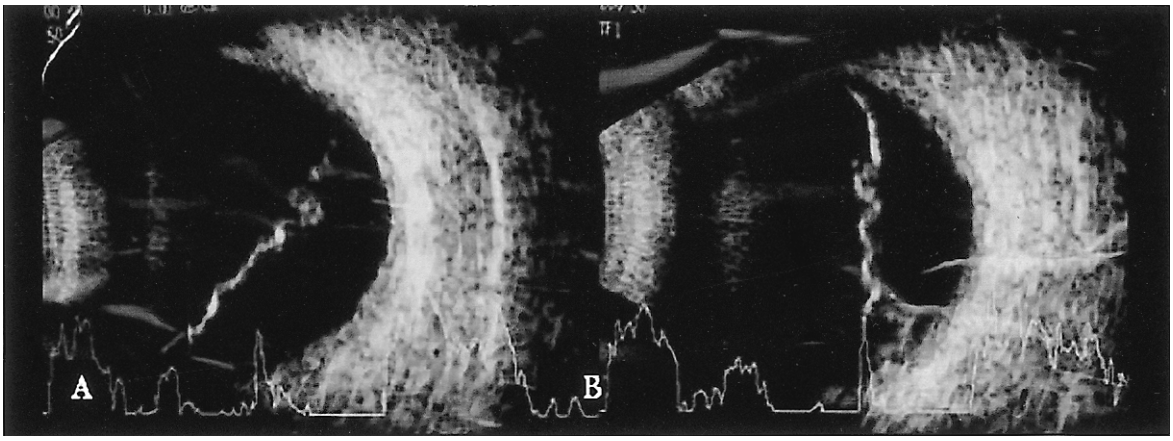

Fig. 4. B-scan (A, B). showing shifting of subretinal fluid. (A) sitting position. (B) supine position.

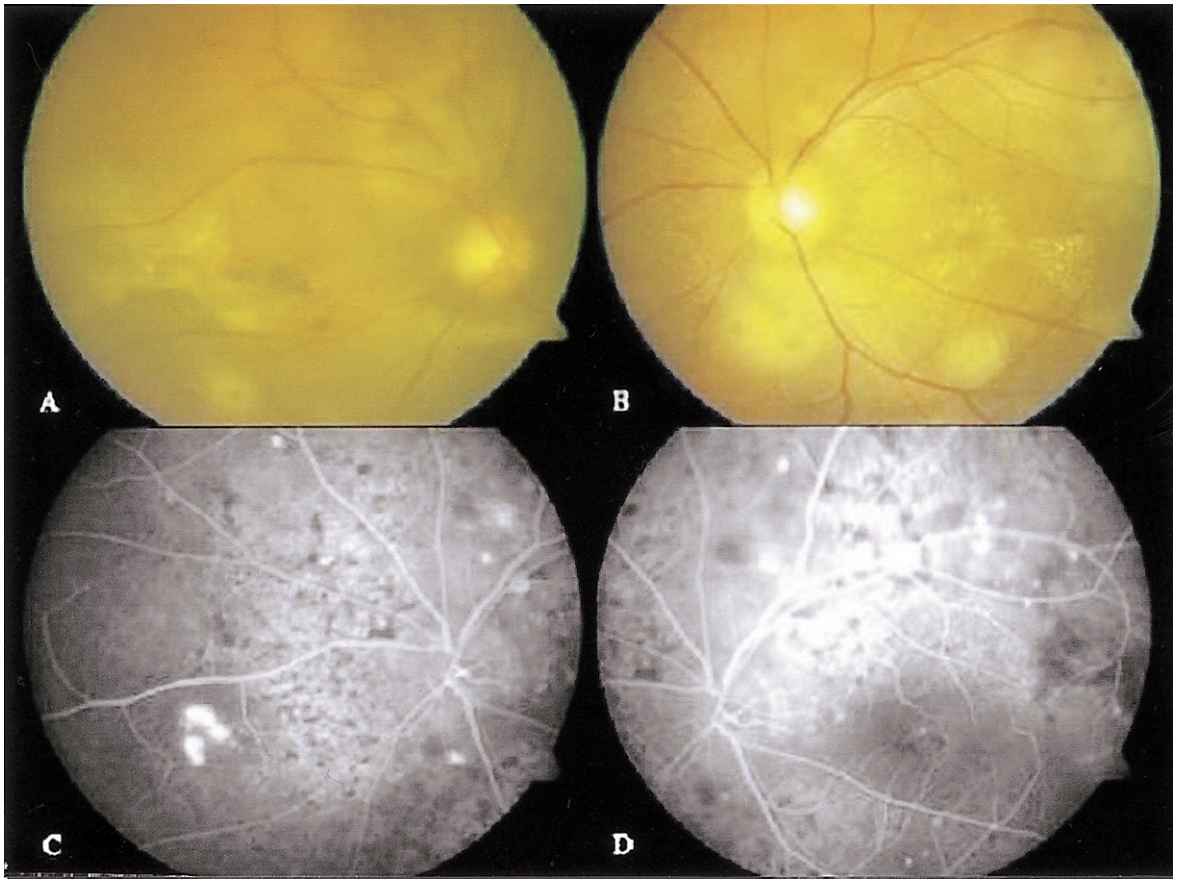

Fig. 5. Color fundus photographs (A, B) and fluorescein angiograms (C, D) six weeks after external drainage of subretinal fluid in the right eye. (A) Right eye. Attached retina with subretinal proliferation. (B) Left eye. Multiple PEDs and exudative flecks. (C) Right eye, Eye absorbed subretinal fluid. (D) Left eye. Multiple points of dye leakage and pooling into the subretinal space.

to our clinic, reoperation was performed on the right eye (including pars plana vitrectomy, lensectomy, membranectomy, internal drainage of subretinal fluid, fluid-gas exchange, and endolaser photocoagulation to leakage points). One week later, the left eye was also treated with the same operation maneuvers. Postoperatively, the retina was well attached in both eyes (Fig. 5), but the final visual acuity of the right eye was unchanged due to a remaining subretinal fibrotic scar and atrophic change of the macula. Fortunately, visual acuity in the left eye improved gradually to 20/200 with correction. Four months after surgery in the left eye, the exudative detachment had not developed further, and he was

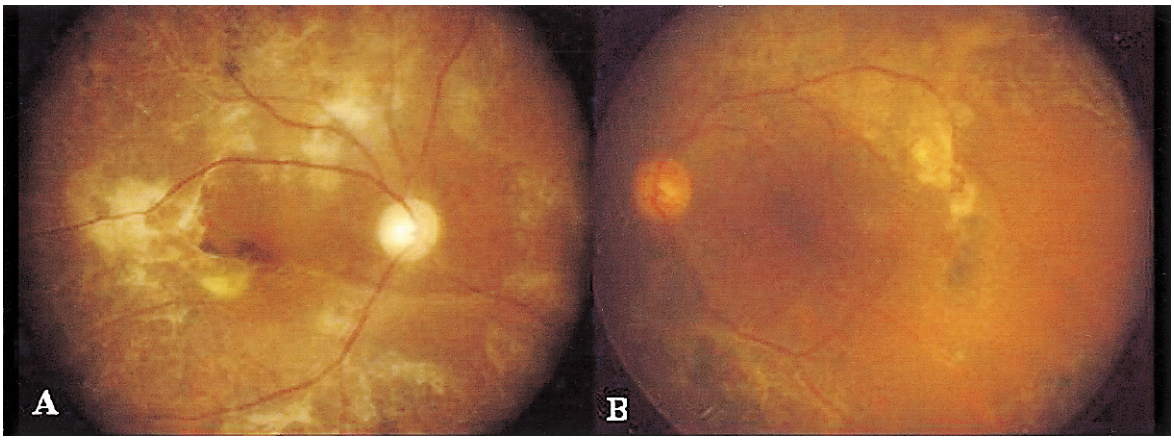

Fig. 6. Color fundus photographs (A, B) after vitrectomy and internal drainage of subretinal fluid of both eyes. (A) Right eye and (B) left eye. Demonstrating an attached retina with atrophic change and subretinal proliferation.

given a secondary intraocular lens insertion in the sulcus. Final visual acuity of the left eye was 20/50.

Discussion

A variant of CSC characterized by a bullous inferior RD with shifting subreitnal fluid and multifocal posterior exudations may predominantly affect healthy male patients. In comparison with the typical form of CSC, the atypical form is characterized by older age, the presence of multiple and bilateral foci, a higher tendency for recurrence and greater accentuation of functional damage.[5-6,8] A bullous exudative RD can arise in different clinical situations for different reasons, such as postsurgical, inflammatory, infectious, or in autoimmune uveitis, vasculitis, and neoplastic causes.[21] Therefore, from a diagnostic and management standpoint, atypical forms of CSC must be distinguished from VKH syndrome, posterior scleritis, multifocal chorioretinitis, uveal effusion syndrome, and choroidal tumors, all of which can also result in exudative RD. In particular, exact differential diagnosis is important in order to avoid the inappropriate use of systemic corticosteroids. On most occasions, diagnoses are made on the basis of the clinical findings and ophthalmoscopic examination. VKH syndrome is considered to cover a wide spectrum of diseases affecting the eyes, meninges, skin, and audiovestibular system that manifest in symptoms of dysacusia, tinnitus, headache, vomiting, and flu-like symptoms for several weeks before the onset of ocular symptoms, vitiligo, poliosis, and alopecia during the convalescent stage. Pleocytosis in the cerebrospinal fluid, granulomatous uveitis in the anterior segment, and trace cells in the vitreous are also characteristic signs of VKH syndrome which help discriminate from atypical CSC. Posterior scleritis occurs predominantly in women, and does not show the yellow-white lesion to the outer retinal layers with single or multiple PEDs. Multiple choroiditis is more common in females, and is accompanied by panuveitis. Idiopathic uveal effusion occurs predominantly in men and usually 10 years earlier than CSC; also, the presence of ciliochoroidal edema, annular peripheral choroidal detachment, scleral thickening, and minimal fluorescein angiographic evidence of permeability alteration at the level of the RPE are distinguishable from atypical CSC. Choroidal tumors (including choroidal hemangioma and choroidal melanoma) can be differentiated by standardized echography according to a characteristic shape and internal reflectivity.[4-6,10,17,21-22] Treatment is directed at the cause of the RD, reabsorbing the subretinal fluid over a variable time interval.

In atypical CSC, laser photocoagulation therapy on area of the pigment epithelial detachment reduces the duration of the disease.[3-6,8] Most of the previously reported cases have indicated that the use of high-dose corticosteroids may systemically be a risk factor for the development of a bullous exudative RD in association with CSC.[8,10-16] However, the atypical form of CSC can occur spontaneously without previous history of corticosteroid therapy.[17,18]

At present, the only treatment available for patients affected by CSC is laser photocoagulation to the leakage points identified during fluorescein angiography.[1-7] However, when the RD is bullous (as was this case), the subretinal fluid can interrupt the available treatment, meaning that the subretinal fluid should be drained in order to carry out the appropriate therapy. Previously reported cases introduced different techniques for subretinal fluid drainage.[12,20] The simplest technique is an external drainage through the sclera in the inferior area. However, when there is a large amount of dense subretinal fluid and fluid-shifting to a dependent position (as in this case), complete transscleral drainage is very difficult. In 1980, Benson et al.[12] performed fluid or air injections through the pars plana to complete this drainage procedure. This approach may pose a problem in postoperative visualization by making the use of a laser difficult in the immediate postoperative period. In 2003, Kang et al.[23] introduced the subretinal aspiration and injection device

(SR-AID), an instrument designed to facilitate controlled external drainage. This device provides a safe approach to the subretinal space because it penetrates the eye wall obliquely and allows a changeable, predetermined length of the needle tip. The surgeon can also directly observe the retina with an indirect ophthalmoscope during the drainage. However, it is not commercially available and requires skillful handling.

Another technique involves carrying out an internal drainage, using pars plana vitrectomy, drainage retinotomy and fluid-air exchange with endolaser photocoagulation during the surgery or an external laser in the postoperative period. Several problems also have been pointed out concerning this method, such as reopening of the retinotomy itself, as well as the difficulties already mentioned with visualization with air in the vitreous cavity. Adán and Corcóstegui[20] presented a new surgical technique for the treatment of exudative retinal detachment associated with CSC; this includes pars plana vitrectomy, injection of perfluorocarbon liquid (PFCL) with transscleral drainage, and endolaser photocoagulation to leakage points. According to their report, this approach has some advantages, such as complete intraoperative reapplication of the retina and intraoperative laser photocoagulation due to excellent visualization through the PFCL without requiring a drainage retinotomy. In 2003, Chen et al.[24] reported that they had successful results with the same method.

There have been no published reports regarding a surgical approach in bullous exudative RD which develops in association with atypical CSC, although 25 cases have been previously reported in the Korean literature.[4-6,19] We first tried to perform transscleral drainage of subretinal fluid in the right eye; as a result, the retina was attached. At a later time, however, exudative RD recurred in the right eye and developed newly in the left eye, and an internal drainage of subretinal fluid for both eyes had to be performed. The surgery consisted (in order) of pars plana vitrectomy, lensectomy, membranectomy, drainage retinotomy, fluid-gas exchange and endolaser photocoagulation to leakage points.

Postoperatively, the retina was well attached in his right eye, but his final visual acuity was not improved. The long-term existence of subretinal fluid followed by irreversible atropy of retinal pigment epithelium and neuroretina results in poor visual outcome.[6] In addition, subretinal fibrotic scarring is a risk factor for permanent loss of vision.[11] Although we cannot completely exclude the effect of the initially administrated corticosteroid on the disease progression, the poor final visual outcome in his right eye is probably because of the subretinal fibrotic scar and the folds on the macula which had been present at his first visit. His left eye had received surgical treatment before subretinal fibrosis progressed, and visual acuity was improved to 20/50.

In conclusion, we consider this surgical management of bullous exudative RD associated with atypical CSC to be safe and useful. However, the surgical treatment should be performed early to prevent subretinal proliferation, which has a negative influence on the final visual prognosis.

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