Case Report

Surgical Treatment of Bullous Exudative Retinal Detachment Secondary to Atypical Bilateral Central Serous Chorioretinopathy

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Abstract

This study aimed to report the diagnostic process, treatment, and follow-up of a patient with bullous exudative retinal detachment (RD) associated with an atypical variant of bilateral central serous chorioretinopathy (CSCR). A 28-year-old woman was referred to our clinic for total bullous RD in the right eye with a vision level of light perception only. She had been previously diagnosed with idiopathic uveal effusion syndrome and treated with systemic corticosteroid therapy with no response, and was referred to us for scleral window surgery. Four-quadrant scleral window surgery with external drainage of the subretinal fluid was performed, resulting in a transient partial attachment of the retina. RD started to progress again within 3 weeks, which prompted comprehensive imaging together with more advanced systemic workup for systemic lupus erythematosus and other rheumatological and immunological diseases. Systemic corticosteroid therapy was initiated during this period but did not stop the progression and was discontinued after a short time. Fluorescein angiography and indocyanine green angiography revealed multifocal choroidal leakage foci and large choroidal vessels without any intraocular inflammation findings and led to the diagnosis of atypical CSCR. Pars plana vitrectomy (PPV), internal drainage of the subretinal fluid, endolaser to the focal leakage areas, and intravitreal aflibercept injection were performed. Visual acuity increased to 0.8 within 8 months after the surgery with no recurrence. Bullous exudative RD is a very rare and atypical form of CSCR, and a favorable outcome can be obtained with PPV and surgical drainage of subretinal fluid followed by laser photocoagulation.

Keywords: Atypical central serous chorioretinopathy, bullous exudative retinal detachment, bilateral involvement, corticosteroid therapy, drainage of subretinal fluid, laser photocoagulation, pars plana vitrectomy

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Introduction

Central serous chorioretinopathy (CSCR) is a disorder characterized by serous macular detachment and/or focal changes in the retinal pigment epithelium (RPE), frequently limited to the macula and associated with fluid leakage in the subretinal space.[1] The main pathogenesis of CSCR has not been clearly defined but some theories focus on the role of the choroid and RPE. Choroidal hyperpermeability and RPE dysfunction caused by stasis, ischemia, or inflammation lead to vascular dilatation and leakage into the interstitial or stromal space.[1]

An atypical CSCR variant with exudative bullous retinal detachment (RD) is observed very rarely in comparison with acute and chronic CSCR. The differential diagnosis of bullous exudative RD includes Vogt-Koyanagi-Harada (VKH) disease, posterior scleritis, choroidal tumors (malignant melanoma, hemangioma, metastasis), uveal effusion syndrome, nanophthalmos, retinal vasculitis, lupus choroidopathy, multifocal choroiditis, Coats' disease, retinal hemangioblastoma, vasoproliferative tumors, malignant hypertension, and even rhegmatogenous RD.[2] Corticosteroid therapy administered as a result of misdiagnosis causes worsening of CSCR findings and delay of appropriate treatment.[3]

In the present case, we report the diagnostic steps, treatment, and follow-up of a patient with atypical CSCR manifesting with massive bullous exudative RD.

Case Report

A 28-year-old woman with no known systemic disease presented 1 year earlier to another center with sudden vision loss in her left eye (LE). She was diagnosed with RD and underwent pars plana vitrectomy (PPV). Visual acuity (VA) in the LE was no light perception postoperatively. She presented again to the same clinic with sudden vision loss in the right eye (RE) that started 2 weeks earlier. She was diagnosed with idiopathic uveal effusion syndrome following a limited systemic workup and received intravenous pulse steroid therapy for 3 days, followed by oral steroids. When no response was observed, she was referred to our center for possible scleral window surgery.

On admission to our clinic, VA was light perception only in the RE and no light perception in the LE. Intraocular pressures were 16 mmHg (RE) and 18 mmHg (LE). Anterior segment examination of the RE revealed leukocoria due to the bullous detachment of the retina coming to the back of the clear lens (Figure 1A). There was no flare or cells in the anterior chamber. The LE was aphakic with total fibrotic RD associated with subretinal fibrosis (Figure 1B).

Ultrasonography was done to rule out a tumor or posterior scleritis. Systemic steroid therapy was rapidly tapered and a systemic workup was started for rheumatologic and inflammatory diseases. However, laboratory tests were inconclusive. A 4-quadrant scleral window surgery with external drainage of subretinal fluid was performed under general anesthesia and resulted in almost complete reattachment of the retina at the end of surgery. At postoperative 2 weeks, BCVA had increased to counting fingers at 10 cm with shallow inferior detachment and subretinal yellow-white fibrin deposits. There was no vitritis or vitreous haze in the RE. Optical coherence tomography (OCT) images showed shallow foveal detachment with subretinal hyperreflective material suggestive of fibrin. The patient was followed up without additional treatment.

At the next visit 6 weeks after surgery, VA in the RE had decreased to hand motions and subretinal fluid had increased to become bullous again in the inferior hemisphere (Figure 2A, B, C). Ocular ultrasound revealed RD and a thick choroid (Figure 2B). Choroidal thickness was measured as 397 µm with enhanced depth image-OCT. The patient was hospitalized to conduct further laboratory testing for systemic lupus erythematosus and other causes of systemic vasculitis. Intravenous 1000 mg pulse methylprednisolone (Prednol, Mustafa Nevzat İlaç Sanayi, Türkiye) was given for 3 days and azathioprine (Imuran, ASPEN Europe GmbH, South Africa) 25 mg twice daily was initiated while preparing for fluorescein angiography (FA) and indocyanine green angiography (ICGA). ICG dye is not readily available in our country and must be imported from abroad. However, RD continued to progress during the 3-day period after steroid treatment. FA demonstrated multifocal hyperfluorescence in the early phase and multifocal staining (multiple hot spots) in the late phase in all quadrants (Figure 2D). There was no leakage from the retinal vessels or staining in the optic nerve head on FA. ICGA revealed widespread diffuse dilated choroidal vessels with no evidence of choroiditis (Figure 2E). Disease progression after corticosteroid treatment, absence of intraocular inflammation, thick choroid, and the FA and ICGA findings led us to the diagnosis of atypical CSCR with bullous exudative RD. Corticosteroid and azathioprine were stopped. A psychiatry consultation was arranged to start antidepressants. A second surgery was planned for the bullous RD, which again reached the back of the lens during this period.

PPV with internal drainage through a superior small retinotomy was performed and endolaser was applied to the staining foci by looking at the FA images during the surgery. Sulfur hexafluoride 20% was used as a tamponade and intravitreal aflibercept 2 mg/0.05 mL was injected at the end of the surgery.

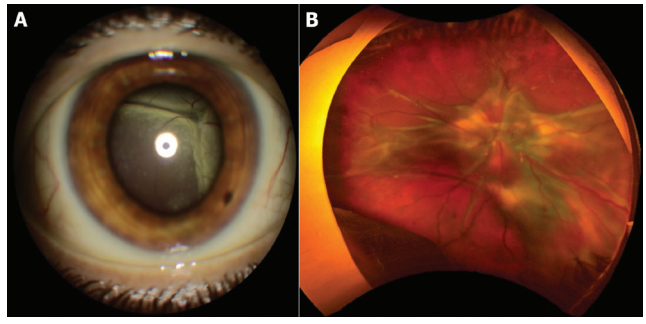

Figure 1. Anterior segment image of the right eye and fundus photograph of the left eye at first admission. (A) Total bullous retinal detachment was seen behind the clear lens in the right eye. (B) Wide-angle fundus photograph of the left eye showed subretinal fibrotic bands and detached retina at the posterior pole

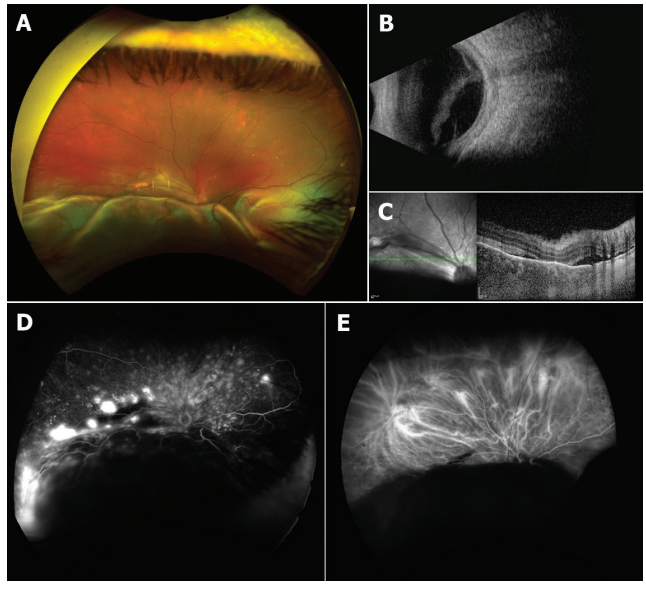

Figure 2. Multimodal imaging of the right eye at 6 weeks after the first surgery. (A) Inferior bullous retinal detachment. Ultrasonography (B) and enhanced depth imaging optical coherence tomography (C) revealed a thick choroid. (D) Fluorescein angiography demonstrated widespread leakage from multiple choroidal foci in the late phase at 4:15 min. Note the absence of retinal vascular leakage or staining of the optic nerve head. (E) Diffuse dilated choroidal vessels were observed on indocyanine green angiography

The retina was totally attached but VA remained at the level of counting fingers with resorption of the gas at the end of the first postoperative month. OCT revealed severe damage to the outer retinal layers. Follow-up FA revealed almost total regression of the leaking foci. The patient was followed up without further intervention. At the final examination 8 months after surgery, the retina remained attached and VA had increased to 20/25. OCT demonstrated restoration of the outer retinal layers starting from the fovea (Figure 3).

Discussion

An atypical form of CSCR, severe exudative bullous RD is a very rare clinical entity characterized by bilateral occurrence, multiple leakage foci, and severe vision loss.[4,5,6] This clinical presentation may be misdiagnosed as inflammatory diseases such as VKH disease, posterior scleritis, multifocal choroiditis, idiopathic posterior uveitis, lupus choroidopathy, or uveal effusion syndrome.[2] Multimodal imaging is crucial for the differential diagnosis, which is key in the management of these cases.

Several risk factors have been described, including male sex, type A personality, conditions that cause high corticosteroid levels (i.e., Cushing syndrome, pregnancy), systemic or local corticosteroid use, and psychological stress. The exact pathogenesis is still unclear. Some authors have suggested that the effects of steroids on the blood-retina barrier, choriocapillaris, and RPE lead to hyperpermeability and subretinal fluid accumulation.[3] This atypical severe CSCR associated with bullous exudative RD may be an exacerbated form of CSCR with possible risk factors such as corticosteroids. Sharma et al.[7] reported that 23 of 29 CSCR patients with exudative RD were using systemic corticosteroids. Gass and Little[6] reported a case of bilateral bullous RD that occurred after the administration of systemic and sub-Tenon corticosteroid in a patient with CSCR who was misdiagnosed as having choroiditis. Similarly, Cebeci et al.[5] reported bilateral bullous RD in a patient with CSCR who was given systemic and sub-Tenon corticosteroids with the misdiagnosis of VKH disease.

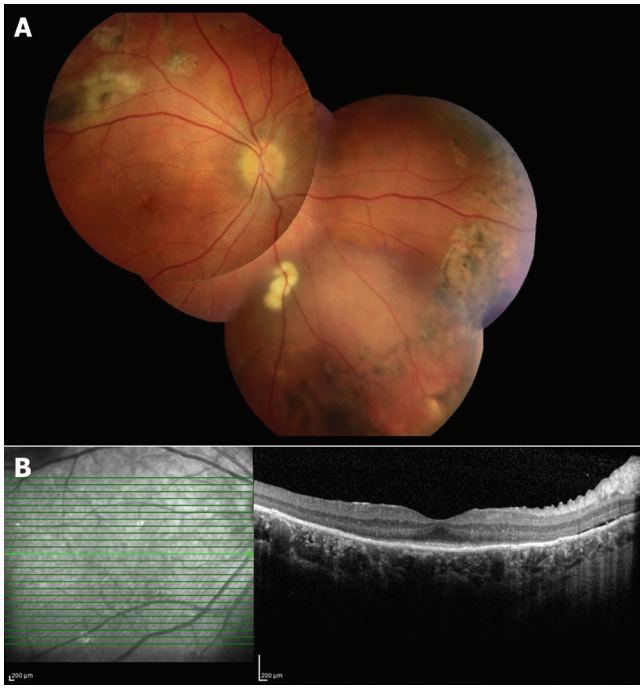

Figure 3. Imaging of the right eye at postoperative 8 months. (A) Composite fundus photograph showed the retina remained attached, with laser scars and some subretinal fibrin deposits. There was only a small area of shallow detachment in the inferonasal retina. (B) Optical coherence tomography showed macular attachment and marked improvement of the outer retinal layers, with reformation of the ellipsoid zone

Our patient was referred to our clinic for scleral window surgery with a possible diagnosis of uveal effusion syndrome. Although she received a course of steroid treatment with no response, it was not the steroid that initially triggered the formation of bullous exudative RD. There was also no history of steroid therapy during the loss of vision in the previously affected LE. In addition, recurrence of bullous RD after the first surgery was not related to steroid use in the present case. However, continued progression of exudative RD despite intravenous steroid therapy, the presence of pachychoroid, and the findings of FA (multifocal choroidal leakage without leakage from the retinal vessels or staining of the optic nerve head) and ICGA (dilated choroidal vessels) led us to the diagnosis of atypical CSCR.

Subretinal fibrosis and scar formation seem to be associated with severe CSCR when treated with corticosteroids. Hooymans[8] reported a patient with CSCR who developed subretinal fibrotic scar formation during systemic corticosteroid therapy. Sharma et al.[7] reported 29 multifocal CSCR patients with subretinal fibrosis and exudative RD, most of whom were given systemic steroids. The formation of subretinal bands and scarring at the posterior pole causes severe vision loss.[4,5] Our patient had subretinal fibrosis leading to tractional RD and no light perception in the fellow eye. Although she had a history of PPV in this eye a year earlier in a different clinic, we do not know the details of that surgery.

Differential diagnosis is essential for the treatment of this variant of CSCR. Cessation of corticosteroid therapy (if started) should be the first step of treatment. If there is no regression of the RD after steroid cessation, treatment should be considered. Non-surgical treatments like photodynamic therapy (PDT) or focal argon laser photocoagulation may be an option in cases with limited exudative RD.[3,5] However, cases with extensive bullous RD should be treated with surgical techniques such as external drainage or PPV with internal drainage to prepare for the application of any laser treatment. In the present case, scleral window surgery with external drainage of the subretinal fluid resulted in partial reattachment of the retina in the early period but could not prevent RD recurrence in a very short time. The second surgery included PPV with internal drainage of subretinal fluid, as well as endolaser to the leakage foci observed on FA, which we believe was the "sine qua non" of the surgery to

obtain the successful outcome. Anti-vascular endothelial growth factors (VEGFs) are thought to have beneficial effects in chronic CSCR and cases with subretinal fibrin exudates.[9] Yannuzzi[9] suggested that subretinal fibrin in CSCR is a result of leakage from abnormal choroidal vessels and recommended intravitreal anti-VEGF injections in cases with fibrin exudates, as we did in the present case.

The systemic mineralocorticoids eplerenone and spironolactone have been used with limited success for the treatment of this variant of CSCR.[1] Cebeci et al.[5] used eplerenone in combination with laser photocoagulation and PDT on an eye with bullous RD in a patient with atypical CSCR and asymmetric bilateral exudative RD, also with limited success. Kang et al.[4] performed a surgical technique similar to ours in their bilateral case but reported increased VA in only one eye because of the development of subretinal fibrosis in the fellow eye, which exhibited more severe involvement. Ng et al.[10] administered a half dose of verteporfin PDT to a patient with inferior exudative RD, which resulted in complete resolution of the RD within 3 months. Both laser photocoagulation and PDT seem to be effective for the treatment of CSCR with limited exudative RD.

In retrospect, we criticize ourselves for starting a second course of steroids for the recurrent RD following the first surgery, which resulted in more rapid progression. During that time, the RD was already progressing without steroids and we had to wait 3 more days to get ICG because it is not readily available in our country. We should not have challenged with steroids during this short time period just to be sure that it did not respond to steroids.

The present case may be unique in demonstrating a quick response to PPV and internal drainage, which enabled evacuation of the subretinal fibrin deposits and thereby prevented late subretinal fibrosis. Intravitreal anti-VEGF injection at the end of the surgery may have provided an additional benefit to prevent subretinal fibrosis. Intraoperative FA-guided endolaser application to the choroidal leakage foci was another important feature of the surgery. The restoration of the outer retinal layers and very satisfactory increase in VA within months after the surgery were also unique to this case.

In conclusion, the differential diagnosis of exudative RD should be done with multimodal imaging and careful systemic investigation to exclude malignancies and inflammatory and vascular diseases. An atypical variant of CSCR should be considered, especially when there is progression in response to steroid therapy. Steroid cessation, mineralocorticoid therapy, PDT, and laser photocoagulation therapy can be tried in moderate cases. However, surgical treatment with PPV, internal drainage, and endolaser photocoagulation to the leaking choroidal foci may be useful in the severe form of the disease, offering rapid recovery and good visual improvement. Anti-VEGFs injected at the end of surgery may be beneficial in reducing fibrin accumulation.

Ethics

Informed Consent: Obtained.

Peer-review: Externally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: Ş.Ö., H.B.Ö., M.H., Concept: Ş.Ö., H.B.Ö., G.G., İ.T.T., Design: Ş.Ö., H.B.Ö., M.Y., Data Collection or Processing: H.B.Ö., M.Y., Ş.Ö., A.M.S., Analysis or Interpretation: Ş.Ö., G.G., A.M.S., M.H., İ.T.T., Literature Search: M.Y., H.B.Ö., Writing: H.B.Ö., M.Y., Ş.Ö.

Conflict of Interest: No conflict of interest was declared by the authors.

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