Post-vitrectomy Observation of Coat's Disease Associated with Exudative Retinal Detachment, Successfully Treated with Long-term Silicone Oil Tamponade

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(Received September 11, 2013; Accepted January 24, 2014)

Background: Treating Coat's disease with exudative retinal detachment remains a challenge, since vitreous surgery is frequently accompanied by serious complications such as secondary glaucoma.
Case: A 25-year-old woman with Coat's disease of the right eye had cystic exudative retinal detachment along the naso-inferior vessels at the peripheral retina, due to 5-o'clock telangiectasia. Right visual acuity was finger counting. This patient underwent vitrectomy simultaneously with cataract surgery, completed with silicone oil replacement, with thorough drainage of subretinal exudates through the artificial break. Although the complication of secondary glaucoma developed as a result of using silicone oil, approximately three years after the operation, this condition was ameliorated by removing the silicone oil. Thus, blindness was prevented in this patient.
Conclusion: Long-term tamponade with silicone oil facilitates performing vitreous surgery in this patient suffering from Coat's disease associated from exudative retinal detachment.

Key words: Coat's disease, exudative retinal detachment, vitrectomy, long-term silicone oil tamponade,

INTRODUCTION

Coat's disease is an uncommon form of retinal telangiectasia, characterized by venous dilatation in the peripheral retina, and massive subretinal exudates resulting from excessive permeability of vessels [1, 2]. As for the outcomes of patients with Coat's disease, the simultaneous development of secondary retinal detachment results in a generally incurable state, although minimal involvement of abnormal vessels occasionally allows spontaneous recovery. Direct coagulation of the abnormal vessels has been established as the main therapeutic strategy for Coat's disease, regardless of whether photocoagulation or cryocoagulation is selected [3, 4]. However, direct coagulation eventually increases the subretinal exudates, and secondary retinal detachment develops [5]. Herein, we report that we successfully treated a Coat's disease case despite the complications of exudative retinal detachment. In this case, we attempted vitrectomy followed by silicone-oil tamponade. We also present fundus photographs taken before and after surgery, to illustrate findings at the onset of Coat's disease through seven years postoperatively.

CASE

A 25-year old woman complained of sudden visual loss in the left eye in June 2004, and was initially diagnosed as having acute retinal necrosis in a municipal hospital in Kanagawa. She was then referred to the different university hospital, which was unable to provide an adequate explanation for her visual disturbance, before coming our university hospital. Based upon the referral information, visual acuity was initially 50/50 bilaterally, with normal intraocular pressure. Right fundus examination revealed, extensive exudates, with associated retinal detachment, and capillary aneurysms. The exudates extended into the fovea in July 2004, when right visual acuity diminished to finger counting. The patient visited our clinic for another consultation on May 24, 2005. Her family and prior medical histories were unremarkable.

Best-corrected visual acuity in the right eye at the first visit to our hospital was finger counting, but was 60/50 in the left eye. Slit-lamp examination of the anterior segments yielded normal findings. Indirect ophthalmoscopy revealed not only scattered capillary microaneurysms, but also extensive yellow-white, subretinal exudates extending to the fovea, in the right eye (Fig. 1). We finally diagnosed as Coat's disease. Immediately after the examination, we performed photocoagulation repeatedly, but exudative retinal detachment developed in the naso-inferior peripheral retina. On September 21, 2005, phacoemulsification and aspiration, intraocular lens implantation, vitrectomy, endolaser therapy, and silicone-oil tamponade were carried out. Through an intentional break, we thoroughly removed the subretinal exudates with a back-flush needle, until the retina was completely attached, and the patient was then discharged on October 1, 2005 (Fig. 2).

In January 2007, right intraocular pressure rose to more than 25 mmHg. This was recognized a complication often observed in the vitreous surgery using silicone oil. This hypertony could not be overcome by combined instillation of both latanoprost and carte-

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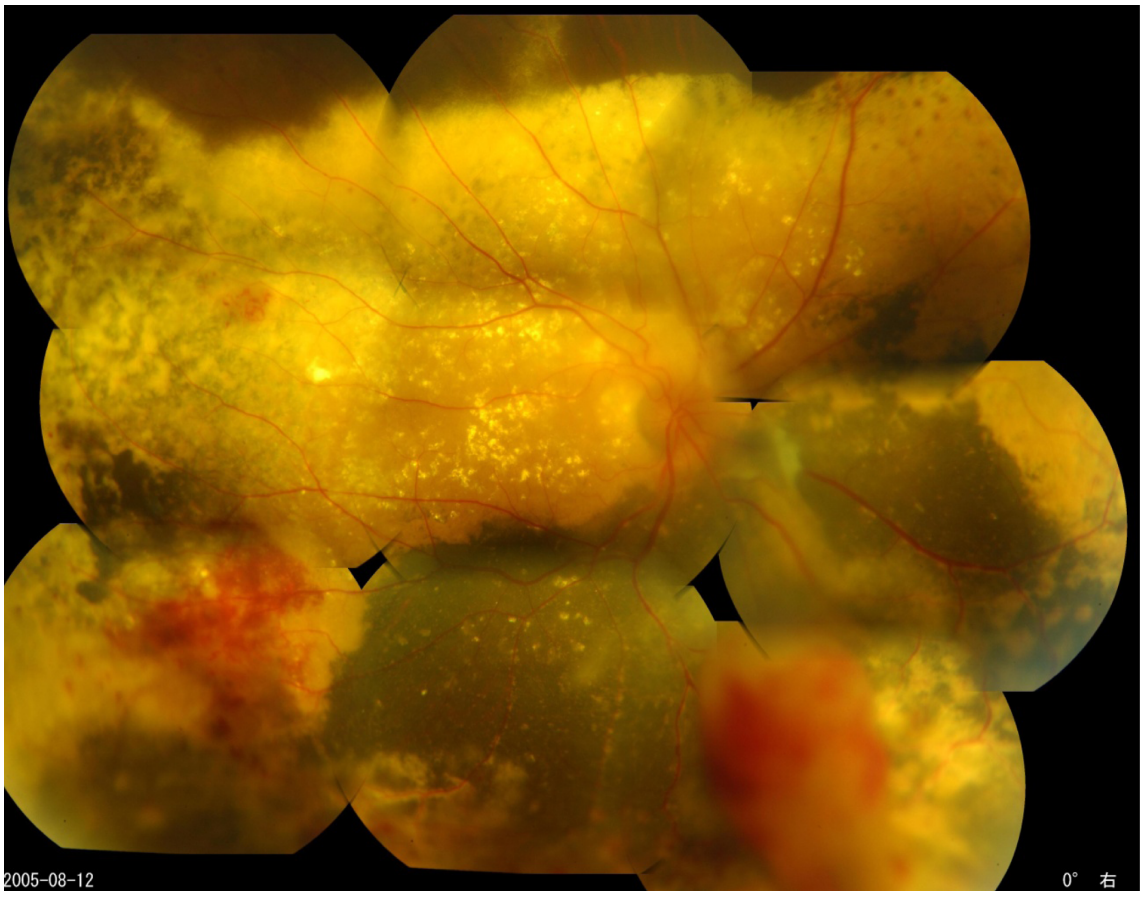

Fig. 1 Massive exudates appeared almost the entire retina. Most notably, exudative retinal detachment was observed at the infero-nasal periphery of the retina

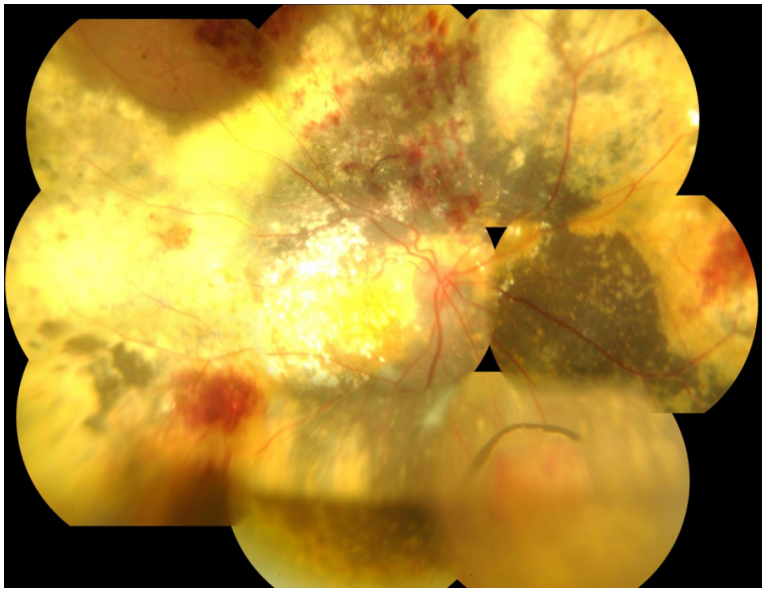

Fig. 2 The fundus picture of our case after the first surgery shows extrusion of the subretinal exudates through the artificial break at the 5 o'clock site

olol hydrochloride, necessitating surgical removal of the silicone oil on August 22, 2008. Her intraocular pressure has remained below 10 mmHg by applying both carteolol hydrochloride and carbonic anhydrase inhibitor, although transiently elevated upto nearly 30 mmHg for a few months after silicone oil removal. A fibrous scar has been formed beneath the posterior pole of the retina, but without foveal involvement. Fortunately, there were no ischemic findings such as iris rubeosis which could eventually have led to a poor outcome (Fig. 3).

DISCUSSION

The first choice for Coat's disease treatment should be direct coagulation of the abnormal vessels, employing either photocoagulation or cryocoagulation [3,

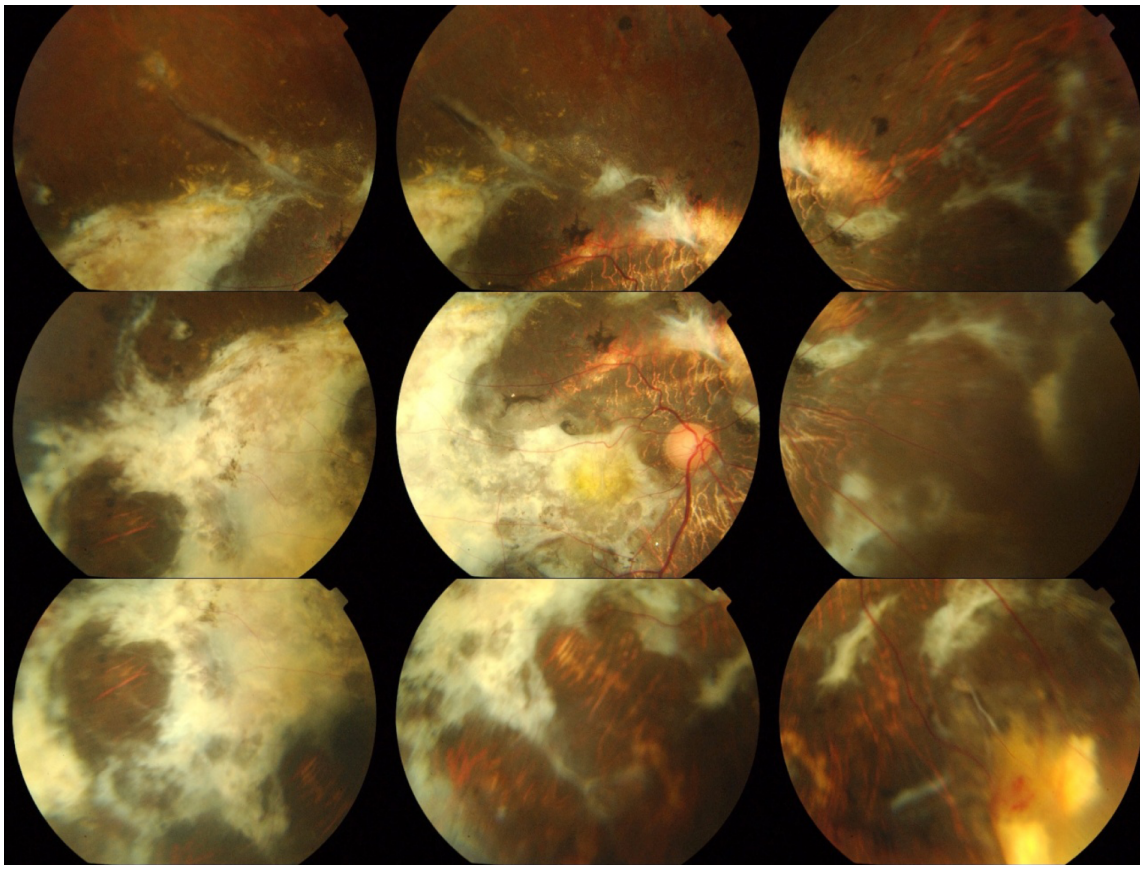

Fig. 3 The exudates disappeared, and had been replaced by a fibrous scar five years after the first operation

4]. The efficacy of these therapies depends upon how large the lesion with abnormal vessel involvement is before the initiation of direct coagulation. If the exudates extend over three quarters of the entire retina, we may have difficulty completely treating this disease with success rates of only 20–75% versus nearly 100% resolution in cases with damage to less than half of the retina [1]. This patient was initially managed at another university hospital, without discussing whether vitreous surgery or observation would have been optimal strategy at that time. We ultimately encouraged this patient to have the operation, because the subretinal exudates had become so massive that no other options were feasible.

The development of secondary retinal detachment may account for the poor visual outcomes of Coat's disease patients with subretinal exudates. Whether or not treating Coat's disease might be effective should be evaluated from two perspectives. First, we should pay attention to fundus improvement. We must also measure the visual acuity of the damaged eye. Without amelioration of the exudative retinal condition, we cannot expect satisfactory improvement of visual acuity. However, photocoagulation in Coat's disease has resulted in only 20–50% of cases achieving better visual acuity, despite ameliorating fundus abnormalities in 60–90% of cases [6]. Preventing organization of exudative changes involving the macula, to the maximum extent possible, allows the gradual restoration of the retina.

Since more than three quarters of the entire retina showed exudative abnormalities in the present case, we immediately selected vitreous surgery to achieve recovery. Through an intentional tear, the subretinal exudate was thoroughly removed prior to photocoagulations. Finally, the operation was completed by placing silicone oil in the vitreous cavity, thereby preventing the exudates at the posterior pole of the retina from reorganizing (Fig. 2). For the purpose of avoiding recurrent exudates, we had left the silicon oil in the vitreous space, which worked the permanent tamponade effect unlike gas replacement. Three years post-vitrectomy, secondary glaucoma developed as a complication of the silicone oil tamponade, and subsequent removal of the oil allowed intraocular pressure to be maintained with eye drops alone. On fundus examination at three years postoperatively, a diffuse inactive and fibrous subretinal scar was present and the exudates had disappeared. Therefore, disease activity was apparently suppressed. Nevertheless, the patient's visual acuity remained the same as at the initial visit to our clinic, probably because the architecture of the retina was irreversibly damaged.

Other than cases with severe proliferative vitreoretinopathy (PVR), vitreous surgery for the treatment of Coat's disease is controversial [7], since serious postoperative complications such as rubeotic glaucoma, and phthisis bulbi may develop [4]. This case had an exudative retinal detachment and we thus selected the vitreous approach, despite with mild PVR. The vitreous surgical approach enabled us to reduce the exudative area, which resulted in the cessation of disease activity. Consequently, vitreous surgery should be considered among the first treatment options for managing Coat's

disease.

As the secondary glaucoma frequently develops as a complication in the silicone-oil infected eye, the present patient continued to suffer from temporal ocular hypertension, despite the silicone oil having been surgically removed, due to irreversible formation of peripheral anterior synechia in response to the silicone oil. Ultimately, this patient's ocular pressure was brought under control with a topical anti-glaucoma drug. Machemer et.al formerly stressed that not only retinal exudation, but also vitreous exudation can induce a sequential cascade leading to PVR in Coat's disease cases [8]. The proteinous exudates spreading into the vitreous cavity through the retina might lead to vitreous contraction, followed by retinal traction, and eventually to exudative retinal detachment. This hypothesis of Coat's disease involving both the retina and the vitreous constitutes the rationale for our long-term tamponade using silicone oil.

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