

Anterior proliferative vitreoretinopathy in a patient with Coats disease

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Dear Editor,

I am Satoru Kase, from the Department of Ophthalmology, Faculty of Medicine and Graduate School of Medicine, Hokkaido University, Sapporo, Japan. I write to present a case of Coats disease showing anterior proliferative vitreoretinopathy (PVR) and neovascular glaucoma.

Coats disease is characterized by exudative retinopathy with unknown etiology, which manifests microvascular abnormalities, retinal exudation, exudative retinal detachment and subsequent neovascular glaucoma (NVG). Patients with Coats disease are treated with preservative ways at a relatively early stage; however, surgical interventions are required in the advanced stage of the disease^[1]. Indeed, eventual enucleation or evisceration may be performed in patients with end-stage Coats disease to avoid ocular pain resulting from NVG. Coats disease can rarely complicate PVR^[2], which is a serious phenomenon causing inhibition of retinal attachments and blindness. However, little is known about the pathogenesis of PVR in Coats disease. We herein report a case of end-stage Coats disease presenting with anterior PVR and NVG who was treated with vitrectomy and excision of anterior PVR membrane that was submitted for histological examinations.

A 12-year-old boy complained of blurred vision and sudden ocular pain in his left eye (OS) for a couple of days. He was healthy otherwise without any medical history. He was referred to Hokkaido University Hospital because of retinal detachment and glaucoma. He had no medical history of trauma. His visual acuity was 20/10 right eye (OD) and hand motion OS. Intraocular pressure was 40 mm Hg OS. Slit-lamp examination

displayed corneal epithelial edema with rubeosis iridis and the detached retina behind the lens (Figure 1A). B-mode echography showed lobulated presentation of highly elevated bullous retinal detachment and hyperreflective subretinal fluid (Figure 1B). It was difficult to determine clinical diagnosis at this stage. Because the patient and his family refused to receive evisceration/enucleation as an initial treatment, the patient underwent 25-gauge pars plana vitrectomy with phacoemulsification and intravitreal anti-vascular endothelial growth factor (VEGF) antibody injection. After removal of the lens, posterior capsule incision and anterior vitrectomy, there were the total exudative retinal detachment with its poor mobility and a plenty of microangiopathy in the peripheral retina (Figure 1D). The proliferative membranes existed from the ciliary body to the retina at 360 degrees, which were peeled off using bimanual technique (Figure 1E; arrows), and a part of membrane was submitted for histopathological analyses. Posterior hyaloid was detached from the surface of an elevated and fragile retina as much as possible following visualization by intravitreal triamcinolone acetonide injection. Telangiectatic vessels were treated using endodiathermy. Perfluoron liquid was injected upon the optic disc to stabilize posterior pole and assess the retinal mobility. After aspiration of perfluoron liquids, a scleral buckle at 360 degrees of eyeball was placed. Subretinal fluids were subsequently removed by internal drainage. After retinal attachment and laser photocoagulation, silicon oil tamponade and inferior iridectomy were performed. Anti-VEGF treatment was performed at the end of the initial surgery. Oral 10 mg prednisolone was administered to control the post-operative inflammation and to prevent a recurrence of PVR. Subsequently, because retinal detachment recurred together with dense fibrin membrane formation in the anterior chamber 3mo after the initial surgery, additional surgical interventions were required. The final visual acuity was hand motion with normal intraocular pressure OS 8mo after the initial surgery. The retina revealed marked exudation under silicon oil tamponade without bullous retinal detachment (Figure 1C).

The anterior PVR membranes obtained during the initial surgery were submitted for histological examination. At a low magnification of the membrane, there was some oval/slit-like defects of the tissue admixed with cellular components (Figure 2A). At a high magnification, several slit-like tissue

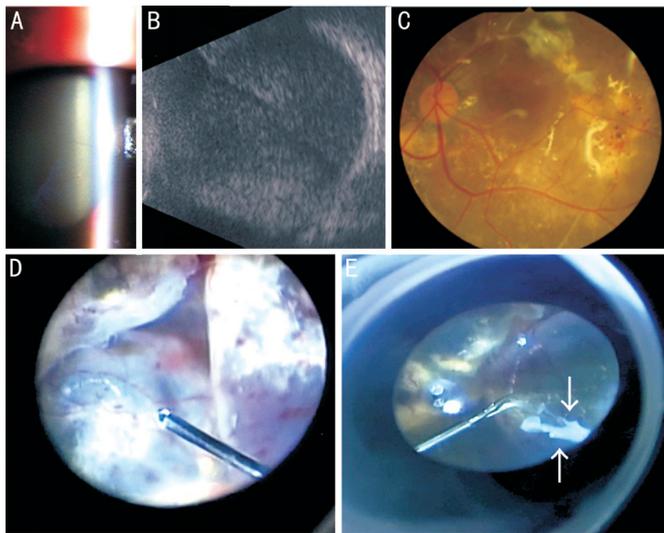


Figure 1 Clinical photographs before (A, B) and 6mo after the initial surgery (C), and intraoperative finding (D, E) in the initial surgery A: The slit-lamp examination displays detached retina adjacent to the lens with iris neovascularization; B: B-mode echography indicates closed funnel suggesting severe total retinal detachment; C: In the final visit of the patient, marked retinal exudation is noted under silicon oil tamponade; D: After posterior capsule incision in the initial surgery, the retina is totally detached with poor mobility and a plenty of telangiectasia vessels; E: The proliferative membrane was peeled of using bimanual technique (arrows), which was submitted for histopathological analyses.

defects were consistent with cholesterol clefts (Figure 2B). Several CD34-positive endothelial cells infiltrated the tissue (Figure 2C). Moreover, CD3-positive T cells were observed in the PVR membrane (Figure 2D), where CD20-positive B cells and CD68-positive macrophages were not intermingled (data not shown). Differential diagnosis of juvenile patients posing retinal detachments with NVG includes Coats disease, long-lasting rhegmatogenous retinal detachment, familial exudative vitreoretinopathy, retinopathy of prematurity, and Norrie disease. The diagnosis was not easily made in this case before surgery, since the retina was not fully evaluated because of corneal epithelial edema and the closed funnel. There is a possibility that evisceration/enucleation can be primarily chosen as an initial treatment if such patients suffered from ocular pain and severe vision loss. Yamashita *et al*^[1] reported a 12-year-old boy having severe retinal detachment due to Coats disease who showed no light perception; however, the visual acuity successfully recovered after vitrectomy. On the other hand, even if active treatments such as retinal reattachment surgeries were conducted in Coats disease complicating PVR as well as NVG, the patients' vision may not be well preserved. This case was treated with vitreoretinal surgeries after the informed consent was obtained. A formal review and approval were waived by institutional review board in Hokkaido University. The principles outlined in the Declaration of

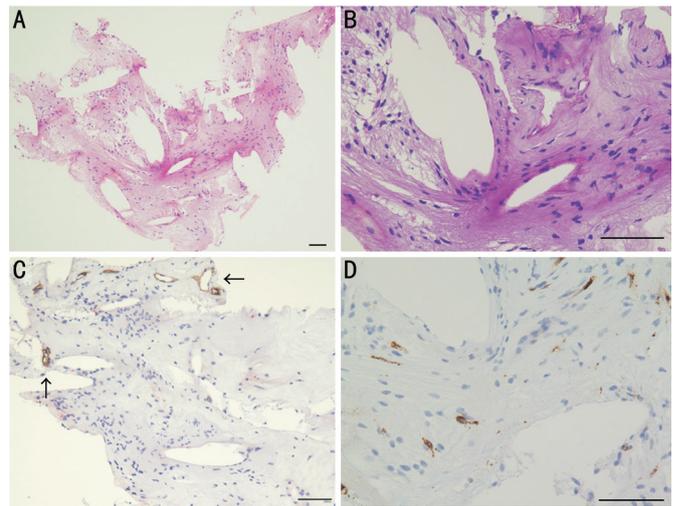


Figure 2 HE staining (A, B), and immunohistochemistry for CD34, a marker for endothelial cells (C), and CD3, a marker for T cells (D), in the excised anterior proliferative vitreoretinal membrane A: At a low magnification of the membrane, there are some oval defects of the tissue together with cellular components (H&E staining); B: At a high magnification, several slit-like tissue defects are noted which are consistent with cholesterol clefts (H&E staining); C: There are several CD34-positive endothelial cells located in the tissue (arrows); D: CD3-positive T cells infiltrate the proliferative membrane. Bars indicate 50 μ m.

Helsinki were followed. The diagnosis with Coats disease could be made during the initial surgery.

Kubota *et al*^[2] reported that pathology of PVR membrane revealed glial proliferation and lipid-laden macrophages in a patient with Coats disease. The case showed no retinal neovascularization using fluorescein angiography^[2]. Cholesterol clefts are characterized by accumulation of lipids in the human tissues that can be identified as clefts in the histology in a process of making specimens. We have reported that cholesterol clefts could be observed in the subretinal space of eyes with Coats disease^[3]. Surprisingly, this study proved the cholesterol clefts in the PVR membrane located upon the retina. In addition, CD34-positive endothelial cells were noted in the PVR membrane proving retinal neovascularization, which was not consistent with the previous report^[2]. We have also demonstrated that VEGF protein expression was strongly detected in the macrophages and detached retina in eyes with the advanced Coats disease^[3]. These results suggest that glial cells and lipid-laden macrophages mainly infiltrate the retina without neovascularization as relative early PVR in Coats disease; however, in the further advanced stage of PVR, VEGF expression originated from macrophages and/or the detached retina led to neovascularization, increased vascular permeability, and enhanced leakage of the lipid from the neovessels, which developed a variety of cholesterol clefts in the PVR membrane.

Lim *et al*^[4] reported that macrophage and perivascular T cell infiltration was noted in the retina of an enucleated eye with Coats disease. We have shown that chronic inflammation takes place on not only the retina but also the choroid in eyes with Coats disease^[3]. This pathological study demonstrated CD3-positive T cell, but not B cell, infiltration in the PVR membrane in addition to the cholesterol clefts. These results suggest that cellular immunity plays an important role in the pathogenesis of PVR in Coats disease as well.

ACKNOWLEDGEMENTS

Conflicts of Interest: Kase S, None; Mori S, None; Noda K, None; Ishida S, None.

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CORRIGENDUM

Retinal ganglion cell-inner plexiform and nerve fiberlayers in neuromyelitis optica

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Correction to: First author affiliation

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The first author's affiliation of Suzhou University was spelling mistake by the authors at acceptance, so hereby certify Soochow University is the corrected affiliation in this paper.

The authors apologize for any inconvenience caused by this error.