

## **Abstract**

**Introduction:** Exudative retinal detachment (ERD) occurs due to accumulation of subretinal fluid in the absence of a retinal break or traction because of disruption in anatomical arrangement of retina. Management of ERD is usually not surgical. Surgical intervention should be done after a failure of conventional therapy to avoid complications of a chronic ERD.

**Purpose:** To report a case of bilateral ERD unresponsive to steroid therapy which was treated with pars plana vitrectomy, internal drainage of subretinal fluid, endolaser photocoagulation, and silicone oil tamponade in a 21-year-old male patient.

**Methods:** A case report.

**Case Report:** A 21-year-old male patient came to Vitreoretina Clinic with bilateral blurry vision and phoptopsia since 7 years ago and progressively worsen. The patient has no history of ocular trauma and related systemic disease. The patient was treated with oral methylprednisolone and oral methotrexate but showed no improvement. Visual acuity was light perception in the right eye and hand movement in the left eye. Slit-lamp biomicroscopy examination revealed a pseudophakic right eye and a phakic left eye. Funduscopy examination found a retinal detachment with hole and subretinal fibrosis in the right eye and a total bullous retinal detachment with subretinal fibrosis in the left eye. The patient was diagnosed as ERD OS, chronic retinal detachment OD, and pseudophakia OD. Pars plana vitrectomy with internal drainage of subretinal fluid, endolaser photocoagulation, and silicone oil tamponade was performed in the left eye.

**Conclusion:** Pars plana vitrectomy with internal drainage of subretinal fluid, endolaser photocoagulation, and silicone oil tamponade is a viable treatment option in ERD unresponsive to steroid therapy.

## **I. Introduction**

Exudative retinal detachment (ERD) occurs when there is an elevation of neurosensory retina due to accumulation of subretinal fluid in the absence of a retinal break or significant pre-retinal traction. Any disruption in anatomical arrangement between neurosensory retina and RPE can cause accumulation of fluid in subretinal space. Neoplasia and inflammatory diseases are the leading causes of ERD.<sup>1,2</sup>

Unlike rhegmatogenous or tractional retinal detachment, the management of ERD is usually not surgical. The treatment of ERD must address the underlying

disease which includes antibiotic, immunosuppression, chemotherapy, or regulating blood sugar and blood pressure. Permanent damage to RPE as well as outer retinal structures occurs when the fluid is non-resolving and retinal detachment is chronic which may lead to subretinal fibrosis formation. To avoid these complications, surgical intervention should be done after a failure of conventional treatment.<sup>1-3</sup>

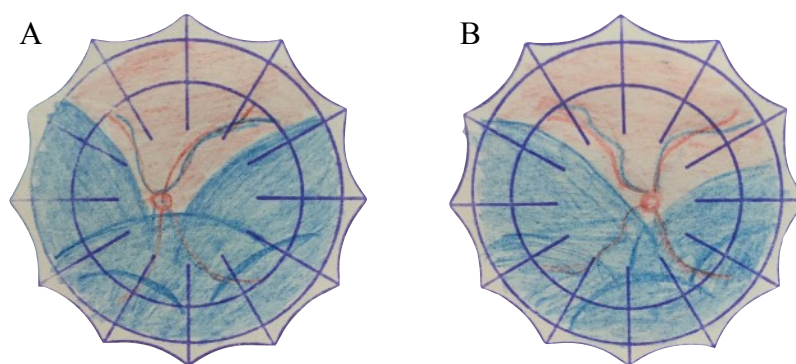
The documentation of surgical treatment for ERD is still lacking. This case report will present a case of bilateral ERD unresponsive to steroid therapy in which had been treated with pars plana vitrectomy, internal drainage of subretinal fluid, endolaser photocoagulation, and silicone oil tamponade in a 21-year-old male patient with probable Vogt-Koyanagi-Harada (VKH) syndrome.

## **II. Case Report**

A-21-year-old male patient came to Vitreoretina Clinic at Cicendo Eye Hospital on September 15, 2016 with a chief complaint of blurry vision in both eyes since 7 years ago and progressively worsen. The patient also complained photopsia. There was no eye pain, red eye, and headache. There was no history of ocular trauma, high myopia, hypertension, diabetes mellitus, and tuberculosis. No sign of vitiligo, poliosis, alopecia, meningismus, tinnitus, and neurological deficits were experienced by the patient.

On February 17, 2009, visual acuity was 0,15 in both eyes. Intraocular pressure (IOP) was 10 mmHg in the right eye and 17 mmHg in the left eye. Anterior segment were normal in both eyes, vitreous cells were 1+. On funduscopy examination, bilateral inferior retinal detachment with macular involvement were found in both eyes. The patient was consulted to External Eye Disease, Neuro-ophthalmology, and Internal Medicine for the underlying disease but no evidence of vasculitis, autoimmune, and tuberculosis was found in this patient. The patient was diagnosed as bilateral ERD and probable VKH syndrome. The differential diagnosis for VKH syndrome was central serous

chorioretinopathy (CSC) and idiopathic uveal effusion syndrome. The patient was treated with oral methylprednisolone 1 mg/kg per day for 3 months and tapered off slowly and oral methotrexate 7,5 mg once a day 3 times a week for 1 month and tapered off slowly. After the therapy, visual acuity was improved in the left eye to 0,5 but showed no improvement in the right eye. On July 6, 2009, Intravitreal triamcinolone acetonide (IVTA) injection was done in both eyes but no resolution of ERD was observed.

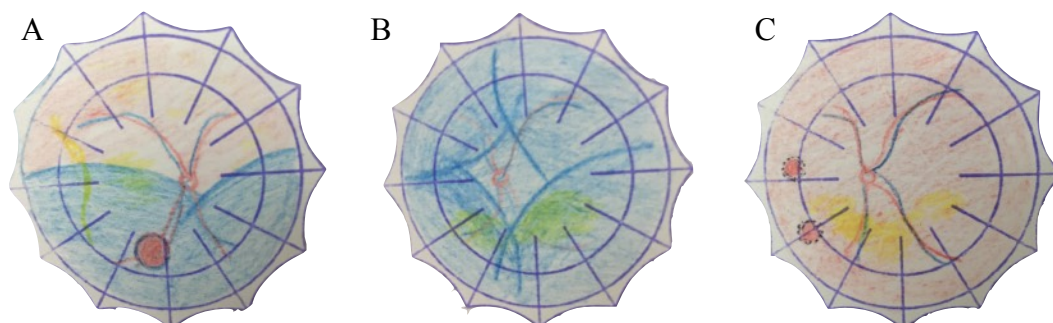


**Figure 1.** Fundus drawing of the right eye (A) and the left eye (B) on first visit showed a bilateral inferior bullous ERD.

On December 26, 2012, visual acuity was light perception in the right eye and 0,2 in the left eye. A dense complicated cataract was formed in the right eye. Exudative retinal detachment was found in the left eye. Phacoemulsification with posterior chamber intraocular lens (IOL) was done in this patient. Visual acuity in the right eye was improved to hand movement after 1 month post-surgery.

On September 15, 2016, visual acuity was light perception in the right eye and hand movement in the left eye. Intraocular pressure was 16 mmHg in the right eye and 10 mmHg in the left eye. The right eye was pseudophakic and anterior segment was normal. The retina was detached on inferior region with a macular hole in inferotemporal region. Subretinal fibrosis was extensive and optic disc was slightly hyperemic. The left eye was phakic and no abnormalities was found on anterior segment. On funduscopy examination, a total bullous retinal detachment

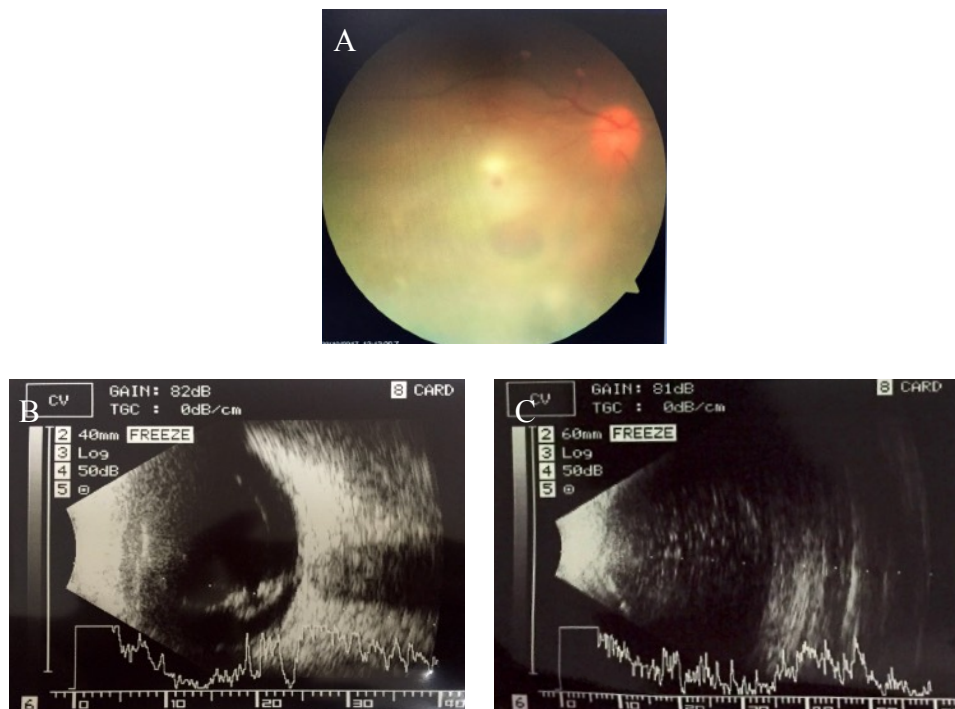
was found. This patient was diagnosed as ERD OS, chronic retinal detachment with macular hole OD, probable VKH syndrome and pseudophakia OD.



**Figure 2.** Fundus drawing showed a chronic retinal detachment with hole and extensive subretinal fibrosis in the right eye (A) and a total bullous ERD with subretinal fibrosis in the left eye (B). Postoperatively (C), the left eye was completely attached with subretinal fibrosis and iatrogenic breaks surrounded by photocoagulation scar.

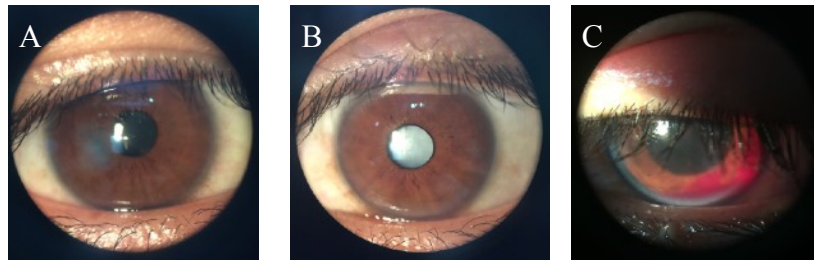
This patient underwent pars plana vitrectomy, endrodrainage, endolaser photocoagulation, and silicone oil 1300 tamponade. Two Iatrogenic breaks were made in the nasal region to perform internal drainage of subretinal fluid. Retina was completely attached at the end of the procedure. Postoperatively, the patient was treated with oral ciprofloxacin 500 mg twice a day, oral paracetamol 500 mg 3 times a day. OS, ofloxacin eye drop 6 times a day, prednisolone acetate eye drop 6 times a day OS, cyclopentolate 1% 3 times a day. Post-surgery day 7, visual acuity was hand movement in the left eye and IOP was 12 mmHg. Retina was completely attached with subretinal fibrosis.

On January 31, 2017, funduscopy examination found emulsified silicone oil. Silicone oil evacuation and exchange to silicone oil 5700 were performed in this patient. Retina was completely attached with hand movement visual acuity in the left eye after 1 week post-surgery.



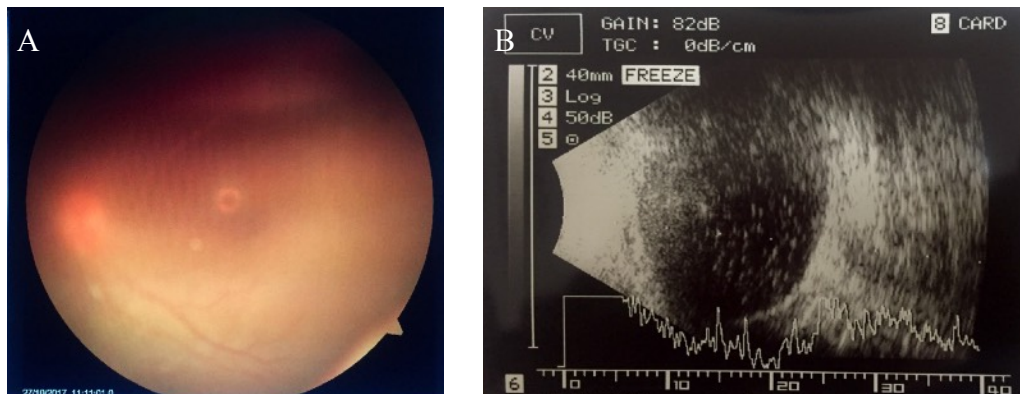
**Figure 3.** A chronic retinal detachment in the right eye (A) as seen on USG (B) and an attached retina in the left eye on USG examination (B).

On September 14, 2017, the patient complained of worsening vision in the left eye. Visual acuity was hand movement in the left eye and IOP was 19 mmHg. On slit-lamp biomicroscopy examination, emulsified silicone oil was found in the anterior chamber, and the cataract was dense. The retina was attached on ultrasonography (USG) examination. This patient was diagnosed as complicated cataract OS, attached retina OS, silicone oil-filled eye OS, chronic retinal detachment with macular hole OD, probable VKH syndrome, and pseudophakia OD. Small incision cataract surgery (SICS) combined with evacuation of silicone oil and exchange to sulfur hexafluoride (SF<sub>6</sub>) tamponade were performed in this patient. Intraoperatively, the retina was attached and subretinal fibrosis was observed.



**Figure 4.** Pseudophakic right eye (A) and complicated cataract in the left eye (B). Anterior segment of the left eye after silicone oil evacuation and exchange to SF6

Post-surgery day 1, Visual acuity was light perception in the left eye., IOP was 9 mmHg. The left eye was aphakic with 3+ aqueous flare and cells. Funduscopy examination revealed a hazy posterior segment. The patient was treated with oral ciprofloxacin 500 mg twice a day, oral paracetamol 500 mg 3 times a day. OS, ofloxacin eye drop 6 times a day, prednisolone acetate eye drop 6 times a day OS, cyclopentolate 1% 3 times a day. Post-surgery day 7, visual acuity was hand movement, IOP was 10 mmHg. Aqueous flare and cells were 1+. An attached retina was seen in funduscopy examination. Prednisolone acetate eye drop was tapered off slowly.



**Figure 5.** Fundus photograph (A) and USG (B) of the left eye showed an attached retina after SICS, silicone oil evacuation, and exchange to SF6.

### III. Discussion

Exudative retinal detachment will be formed when there are abnormalities that drive fluid into the subretinal space and that limit its subsequent removal by active transport, hydrostatic pressure, and plasma oncotic pressure. Outer retina

becomes ischemic due to loss of its blood supply from the choroid. Photoreceptor cell degeneration has been shown to increase as the distance between RPE layer and photoreceptor layer increases.<sup>2,4,5</sup>

Etiology of ERD are due to alterations in choroidal flow, poor scleral outflow, and breakdown of the RPE and retina which can arise in different clinical situations, such as VKH syndrome, CSC, choroidal melanoma, Coats' disease, uveal effusion syndrome, exudative age-related macular degeneration, sarcoidosis, and infections. Exudative retinal detachment is also seen in association with systemic diseases such as pregnancy-induced hypertension, malignant hypertension, connective tissue disorders, vascular disorders, and disseminated intravascular coagulation.<sup>2,3,6</sup>

Vogt-Koyanagi-Harada syndrome is an inflammatory disease defined principally by panuveitis associated with ERD. Vision loss is nearly universal, with associated headache, meningismus, poliosis, vitiligo, and tinnitus. The pathologic mechanism is an autoimmune response to melanin containing tissues, explaining the predilection for the RPE and the highly pigmented choroid. Retinal pigment epithelium breakdown, formation of sub-RPE Dalen-Fuchs nodules, and choroidal inflammation involving the choriocapillaris are key features of this damage, which leads to subretinal fluid accumulation and ERD. Diagnostic criteria for VKH syndrome divides the disease into 3 category: complete, incomplete, and probable VKH syndrome. Regardless of the form of the disease, essential features for the diagnosis of VKH syndrome include bilateral involvement, no history of penetrating ocular trauma, and no evidence of other ocular or systemic disease.<sup>7,8</sup>

One of the more commonly seen etiologies of ERD is idiopathic CSC, which often affects young, otherwise healthy individuals in the third to fourth decade of life. A significant male predominance exists, and there is likely an association with both type A personalities and patients using exogenous corticosteroids. Additionally, atypical forms of CSC have been reported, which

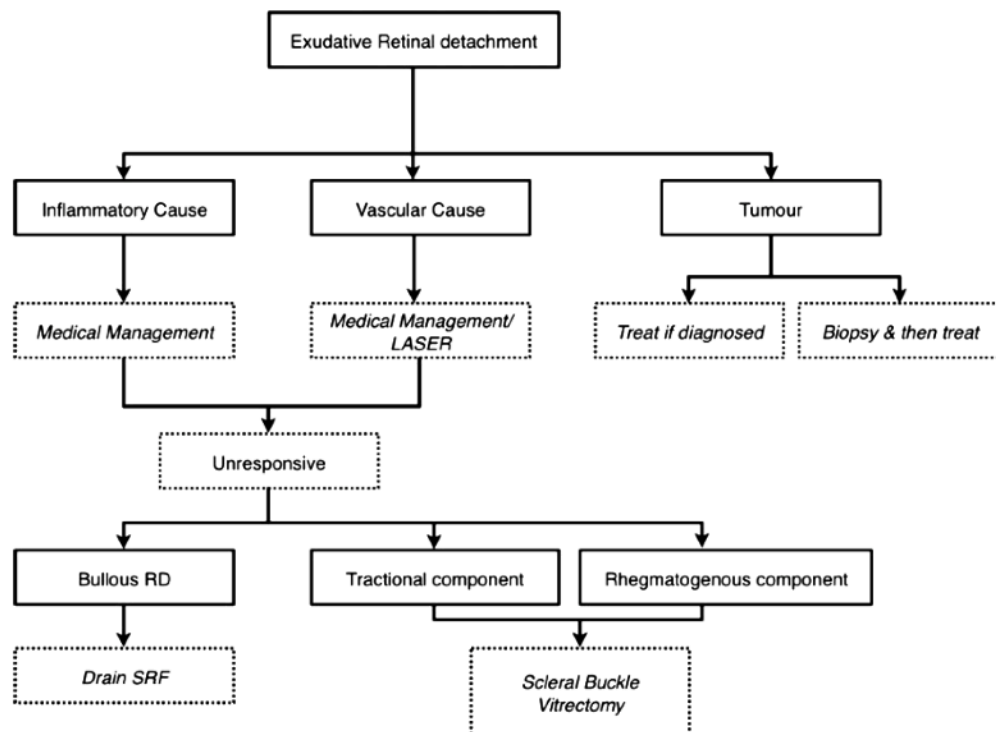
include a chronic form and an acute bullous form which often manifests in the inferior retina. In comparison with the typical form of CSC, the atypical form is characterized by older age, the presence of multiple and bilateral foci, a higher tendency for recurrence, and greater accentuation of functional damage.<sup>1,2,9</sup>

Uveal effusion syndrome caused by alteration of net water movement across the vitreous cavity and posterior eye wall due to reduced trans-scleral aqueous outflow. These patients exhibit dilated conjunctival vessels, shallow anterior chambers, and ciliochoroidal effusions. On fluorescein angiography, these patients often demonstrate a diffuse “leopard spot” pattern which is notably lacking in focal points of leakage, and indocyanine green angiography shows diffuse hyperfluorescence. Uveal effusion syndrome is divided into 3 types. Type 1 is nanophthalmic eye with high hyperopia, type 2 is non-nanophthalmic eye with clinically abnormal sclera, and type 3 is idiopathic non-nanophthalmic eye with clinically normal sclera. Idiopathic uveal effusion syndrome occurs predominantly in men and 10 years earlier than CSC. It is known that idiopathic uveal effusion has a poorer prognosis than other types, and that surgery or systemic steroid therapy is not effective.<sup>2,10</sup>

Vogt-Koyanagi-Harada syndrome, CSC, and idiopathic uveal effusion syndrome were the differential diagnosis in this case. It was difficult to conclude the cause of ERD in this patient due to limited data of ancillary tests. The presenting signs in this patient was only bilateral ERD with 1+ vitreous cells, neurological, auditory, integumentary, and other VKH characteristics were not present. Idiopathic CSC could mimic an early stage of VKH syndrome. While accurate diagnosis can be established in most cases on the basis of clinical examination and ancillary imaging, few cases may have overlapping features making the diagnosis and initiation of therapy difficult. The use of high dose corticosteroids may systemically be a risk factor for the development of a bullous ERD in association with CSC. Exudative retinal detachment and disc hyperemia were the most common posterior segment findings in the acute cases, while sunset



glow fundus was the most common finding in chronic or convalescent cases. This patient did not show any sign of dilated conjunctival vessels, shallow anterior chamber, and ciliochoroidal effusions. Scleral and choroidal thickness were normal in USG examination, therefore it is unlikely that the patient had idiopathic uveal effusion syndrome although this disease is a diagnosis of exclusion. Based on the clinical findings in this patient, the most likely cause of ERD was probable VKH syndrome.<sup>9,11</sup>



**Figure 6.** Management algorithm of ERD

The primary treatment for ERD is to target the underlying disease. Infections are controlled with antibiotics and inflammation with anti-inflammatory therapies. Most of the time, ERD resolves with medication. However, ERD occasionally persists for several months or years, despite the resolution of the inflammation. When this occurs, it can induce the formation of a subretinal fibrosis, and the retina may fail to permanently reattach. Specifically,

ERD associated with uveitis is treated using oral anti-inflammatory agents, subtenon triamcinolone acetonide injection, and IVTA. Moreker and Lodhi, from their study on the use of IVTA in VKH syndrome, have concluded that IVTA, when used as an adjuvant, can induce remission in the acute stage of VKH and help avoid the long-term use of systemic corticosteroids. Aggressive treatment may result in fewer complications and less recurrence. Paredes et al. described immunosuppressive therapy (IMT) given within 6 months of diagnosis with or without steroid was associated with a superior visual outcome when compared to steroid as monotherapy or with delayed additional of IMT. Methotrexate has been used to control pediatric or adult VKH syndrome that inhibit nucleotide synthesis and proliferation of inflammatory cells. Tentative diagnosis of VKH syndrome was made on the first visit. Aggressive therapy with oral corticosteroid, oral methotrexate, and IVTA were given in this patient and improved his vision, but retinal detachment was not resolved. There is no sign of VKH syndrome late manifestation in this patient after 8 years, this could be indicative of a successful immunosuppressive therapy.<sup>3,7,11</sup>

Surgical options in ERD are internal and external subretinal fluid drainage. External drainage can be performed in bullous retinal detachment in periphery. Needle drainage is a relative safe procedure with lower rate of complications such as subretinal hemorrhage, vitreous loss, and retinal incarceration. Internal approach can be done via pars plana vitrectomy. Subretinal fluid may play a role in prohibiting resorption by creating high oncotic pressure that can lead to fluid leakage. The surgical intervention was considered to offer a normal trans-retinal pressure gradient, reduced damage of retinal pigmentary epithelium and photoreceptors, and a tamponade effect from the silicone oil. These characteristics may induce visual improvement through ocular stabilization and retinal reattachment.<sup>3,6</sup>

John et al. reported pars plana vitrectomy and scleral buckle had a long-term success in reattachment of retina in patient with chronic ERD associated with

CSC. A successful case of bilateral ERD with CSC treated with pars plana vitrectomy and internal drainage of subretinal fluid in both eyes was reported by Kang et al. A successful pars plana vitrectomy for bilateral bullous retinal detachment in a patient with VKH syndrome was reported. Gaun et al. described pars plana lensectomy and vitrectomy being done on a 78-year-old female with severe reduction of visual acuity in both eyes because of an extremely bullous ERD accompanied by VKH syndrome. In uveal effusion syndrome, the most common treatment performed is full-thickness sclerectomies to provide an exit for choroidal fluid. Internal drainage of subretinal fluid performed in conjunction with partial thickness sclerectomies may be a preferred method of treating ERD in idiopathic uveal effusion syndrome. In cases of uveal effusion without nanophthalmic, vitrectomy hastens reattachment of the retina and may result in better visual outcome. But in cases of nanophthalmic eyes, it would be better to perform sclerostomy first. Nicholson et al. described the use of pars plana vitrectomy in the management of a patient with an ERD secondary to lupus choroidopathy. Surgery was performed after anti-inflammatory treatments and laser photocoagulation failed to resolve the condition. The patient's vision improved, and the subretinal fluid has not reaccumulated at 5 years postoperative.  
3,9,12-7

In this patient, pars plana vitrectomy, internal drainage of subretinal fluid, endolaser photocoagulation, with silicone oil tamponade was done in the left eye. The surgery was done after initial therapy with methylprednisolone unsuccessfully resolve the bullous detachment. Silicone oil was evacuated 1 year later in combination with SICS when emulsified silicone oil and dense complicated cataract was formed. Surgery was done to prevent secondary glaucoma to occur. Sulfur hexafluoride in order to give a temporary tamponade.

The visual prognosis was *ad malam* due to worsening visual acuity, although we successfully achieve anatomical attachment of the retina in the left eye. Permanent damage of the retina and formation of subretinal fibrosis lead to a

permanent visual impairment. Kang et al. concluded that surgical treatment should be done earlier to prevent the formation of subretinal fibrosis.<sup>3,9</sup>

#### **IV. Conclusion**

Exudative retinal detachment can occur in many clinical scenarios. Inflammatory, vascular, and tumor are the leading cause of ERD. Comprehensive history taking, physical examinations, ophthalmologic examinations, and ancillary tests should be done in order to diagnose the cause of ERD. The management of ERD are based on the underlying disease and rarely surgical. In ERD unresponsive to initial medical treatment, surgical approach should be performed to prevent late complications of ERD such as subretinal fibrosis to preserve a better visual acuity.

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