Management of Bilateral Exudative Retinal Detachment: Case Report & Small Review

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I. Case Report

A 34 year old male presented with chief complaints of sudden diminution of vision in left eye followed by right eye since 1 day which was progressive unassociated with pain. There was neither any history of surgery or trauma nor there was headache, tinnitus, floaters, hearing loss, vertigo, meningismus. No history of fever, giddiness, respiratory illness (TB), STD’s, alopecia, vitiligo, poliosis. Past history was insignificant.

Slit-lamp examination revealed occasional cells in anterior chamber both eyes. Visual acuity was finger counting ½ meter both eyes. Fundus examination detected bilateral multifocal retinal detachments which was also supported by OCT and FFA.

**OCT AT PRESENTATION** - multifocal neurosensory detachments in both eyes.

Right eye Left eye
FFA AT PRESENTATION -

Right eye –

Left eye –

FFA of both eyes revealed hyperfluorescent dots in early phase followed by pooling of dye in sub-retinal space in the late phase.

**B-SCAN** of both eyes evidenced multifocal retinal detachments, with no evidence of any intraocular malignancy.

A systemic workup was initiated. The patient tested negative for tuberculosis, syphilis, toxoplasmosis, and Lyme disease. The patient's tests revealed normal ESR, CRP, ANA, RF, C-ANCA, P-ANCA, serum viscosity. The complete blood count was normal. The comprehensive metabolic panel was within normal limits. No abnormality detected on Chest X-ray.

**DIAGNOSIS** -

Based on history, clinical examination and investigations a diagnosis of **Probable VKH syndrome (Harada disease)** was made.

**TREATMENT** –

In view of patient’s very low vision, patient was put on intravenous pulse therapy for 5 consecutive days followed by oral steroids that were tapered gradually and topical steroid was given qid.

**RECOVERY** –

After 5 consecutive pulse therapy vision in both eyes improved to 6/18.
FFA AT DAY 7–
Right eye -

![Image of FFA at day 7 for right eye]

Left eye -

![Image of FFA at day 7 for left eye]

FFA shows resolution of exudative retinal detachment at day 7.

OCT AT DAY 7-
Right eye –

![Image of OCT for right eye]

Left eye -

![Image of OCT for left eye]
OCT at day 7 revealed maximal resolution of neurosensory detachments with improvement in macular thickness.

**B-SCAN AT DAY 7**
Right eye –

Left eye -

B-SCAN evidenced almost completely reattached retina.

**AT 2 WEEKS** –
Right eye - 6/12
Left eye - 6/12
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FFA & OCT showing completely reattached retina with normal macular thickness.

**AT 4 WEEKS** – after 4 weeks posttreatment, vision improved to 6/9 and 6/6p in right and left eyes respectively with complete resolution of RD both clinically and on OCT.

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<th>Right eye</th>
<th>Left eye</th>
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<td>FFA &amp; OCT showing completely reattached retina and average macular thickness at 4 weeks.</td>
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**II. Discussion**

**Exudative RD** is defined as an elevation of the neural retina caused by the accumulation of subretinal fluid in the absence of a retinal break or significant preretinal traction.

**Characteristics features are** • Subretinal fluid that shifts with postural changes. • Lack of rhegmatogenous or tractional component. • Occurs secondary to local ocular or systemic etiology.
PATHOPHYSIOLOGY-

The choroid, which is both highly vascular and highly permeable, employs both hydrostatic and osmotic forces to “pull” fluid through the RPE and out through the venous drainage, effectively creating a vacuum to pull the retina into apposition with the RPE. Finally, any fluid that cannot be removed by way of the choroid escapes through the sclera, completing internal to external fluidic movement.\(^1\)

Changes in the scleral thickness, as can occur in nanophthalmos, decrease fluid outflow. Further, any disruption of the RPE, or its active transport, will lead to fluid accumulation in the subretinal space. Thus serous detachments can be studied under three mechanisms- alterations in choroidal flow, poor scleral outflow, and breakdown of the RPE and retina.

A. ALTERATIONS IN CHOROIDAL FLOW

Idiopathic Central Serous Retinopathy – Often affects young healthy individuals in third-fourth decade of life with male predominance. Type A personalities and patients using exogenous corticosteroids and pregnancy also carries an increased risk of ICSC.\(^2\) In ICSC, fluorescein angiography (FA) reveals defects in the integrity of the RPE, most commonly as an “expansile dot” of leakage or, classically, a “smokestack” pattern of focal leakage. Bullous formoften manifests in the inferior retina.\(^3\)

Most cases resolve spontaneously. However, in atypical or recurrent cases, or in rare cases with associated choroidal neovascularization, one can consider focal laser photocoagulation or anti–vascular endothelial growth factor (VEGF). In addition, reduced fluence photodynamic therapy (PDT) with verteporfin has been employed, most commonly for the chronic form of ICSC.\(^4\) Finally, oral agents that block aldosterone, primarily spironolactone and eplerenone, have demonstrated reductions in subretinal fluid and improved visual acuity.

Tumors of the Choroid and Retina - Benign lesions, such as nevi and hemangiomas, can also cause serous retinal detachments, seen both clinically and by optical coherence tomography (OCT).\(^5\)

Choroidal melanomas are often dome-shaped subretinallesions. On FA, these tumors appear hyperfluorescent because of their dilated, highly permeable vascular networks. In addition, they may also show a distinct “double circulation” pattern. Choroidal Metastases often show late hyperfluorescence on FA, often with pooling of dye into overlying serous detachments.\(^6\) Treatment is aimed at the underlying primary malignancy and should be coordinated with an oncologist.

Retinoblastomas, often exophytic in growth, are also highly vascularized tumors and can be seen to produce large serous detachments of overlying and surrounding retina. Treatment traditionally consisted of enucleation, but significant advances in globe- and vision-sparing therapy have been made through the use of localized chemotherapy.\(^7\)

Systemic Disease With Disrupted Choroidal Blood Flow - Other, more systemic processes which disrupt normal choroidal flow and lead to overlying serous retinal detachments include malignant hypertension, DIC, renal failure, and pre-eclampsia. FA reveals areas of patchy hypofluorescence, demonstrating focal areas of occlusion and disrupted choroidal blood flow, and these choroidal lesions are often associated with overlying serous retinal detachments. Treatment is directed at the underlying pathology.\(^8\)

Vasculitis and Autoimmune Disease- Systemic lupus erythematosus (SLE), Wegener’s granulomatosis, polyarteritis nodosa, relapsing polychondritis, dermatomyositis, and Goodpasture’s disease produce vascular leakage and/or acute occlusion of the choriocapillaris, or precapillary choroidal arterioles, resulting in necrosis of the overlying RPE and retinal detachment by the mechanism described above. Diagnosis of SLE, and other inflammatory diseases, is best done in conjunction with a rheumatologist, and treatment is directed at systemic control.\(^9\)

B. POOR SCLERAL OUTFLOW

Nanophthalmos and Uveal Effusion Syndrome– Nanophthalmos patients universally have abnormally thickened sclera and can exhibit serous retinal detachments in the absence of other inciting factors.\(^10\) On FA, these patients often demonstrate a diffuse “leopard spot” pattern that notably lacks focal points of leakage, and ICGA shows diffuse hyperfluorescence. Although the exact mechanism of impedance is uncertain, decreased fluid movement through the sclera is the cause of fluid accumulation in the subretinal space, and treatments improving scleral outflow—such as the creation of scleral “windows”—successfully treat the detachments.\(^11\)

Posterior Scleritis - Posterior scleral inflammation and edema, as seen in posterior scleritis and orbital cellulitis, have been reported with serous detachments. Posterior scleritis presents with severe retrobulbar pain, decreased vision, and posterior segment inflammation. Ultrasoundography is useful for diagnosis and reveals
diffusely thickened posterior sclera. Treatment, similar to other forms of sclerosis, consists of high-dose corticosteroids. Orbital cellulitis, rarely exhibits serous retinal detachment. Other infectious etiologies affecting the posterior globe, such as Lyme disease and cat scratch disease, can exhibit small peripapillary serous detachments. Treatment consists of antibiotic therapy tailored to the particular infectious agent.

C. BREAKDOWN OF THE RPE AND RETINA

Vogt–Koyanagi–Harada Disease - VKH is an inflammatory disease found more commonly in Asian and Native American populations, defined principally by intraocular inflammation (panuveitis) associated with exudative retinal detachments. Vision loss is nearly universal, with associated headache, meningismus, poliosis, vitiligo, and hearing loss. The pathological mechanism is an autoimmune response to melanin-containing tissues, which explains the predilection for the RPE and the highly pigmented choroid. In patients with VKH disease, FA reveals patchy filling of the choroid, followed by multiple pinpoints of hyperfluorescence and eventual pooling of dye in areas of exudative detachment. ICGA reveals diffuse hyperpermeability of the choroidal vasculature.

Sympathetic ophthalmia (SO) - It is characterized by an immune reaction to ocular tissue in the wake of a penetrating ocular injury or intraocular surgery is rarer, but can present a similar pathological picture.

Sarcoidosis - Sarcoidosis can also present with associated serous retinal detachment. It is a systemic granulomatous disease of unknown etiology that affects the eye in approximately 25% of patients. Association with RPE detachments has been described in patients with sarcoidosis, presumably from inflammation leading to RPE breakdown and subretinal fluid accumulation.

Infectious Diseases - Toxoplasmosis, a protozoal infection and the most common cause of posterior uveitis, causes a necrotizing chorioretinopathy that leads to characteristic scarring. The detachments were, in the majority of cases, associated with areas of retinal and vascular damage, as well as likely RPE breakdown, although some patients showed associated areas of choroidal ischemia, suggesting a mixed mechanism for serous retinal detachment. Treatment is aimed at the underlying infectious process and leads to resolution of the associated retinal detachment.

Syphilis and cytomegalovirus have also been reported in association with serous retinal detachments. Additionally, Lyme disease (caused by Borrelia burgdorferi), tuberculosis, histoplasmosis, coccidiomycosis, and cryptococcosis have all been associated with serous retinal detachment, and cat scratch disease (caused by Bartonella henselae) and other neuroretinitides can cause peripapillary serous detachments.

Retinal Vascular Diseases - Coats’ disease, a rare disease of unknown inheritance with a 3 : 1 male predominance, presents at an early age and is classically thought to be a unilateral disease. The characteristic presentation is capillary nonperfusion and telangiectasias, with massive exudation of fluid and cholesterol. FA demonstrates “light bulb” aneurysms of the congenitally weakened vessels, and arterioles, capillaries, and venules are all affected. The leakage and exudation leads to serous retinal detachment. Treatment usually involves laser photocoagulation, although recent studies have evaluated the use of anti-VEGF therapy.

Acquired vascular damage can also lead to serous retinal detachment, as has been reported in association with retinal vein occlusions, diabetic macular edema, retinal macroaneurysms, and exudative age-related macular degeneration (AMD).

III. Diagnostic And Ancillary Testing

The presence of a serous retinal detachment is most readily established by clinical examination using indirect ophthalmoscopy, unless precluded by hazy ocular media. Initial examination should include careful evaluation for retinal breaks or tractional membranes. Observation of fluid shifting with changes in headposition, as well as the absence of retinal corrugations or tractional membranes, is an important diagnostic clue.

Complete ocular examination for structural abnormalities, inflammatory manifestations, tumors, or vascular disease helps narrow the diagnosis, and a thorough review of systems is critical. Ancillary imaging serves two purposes: first, evaluation in the setting of hazy ocular media or retrolubular pathology, as with B-scan ultrasonography and computed tomography/magnetic resonance imaging (CT/ MRI); second, demonstration of various pathological patterns, assisting in diagnosis. Additionally, blood testing (as with infectious and autoimmune etiologies) and cerebrospinal fluid sampling (as with VKH disease) can be useful.

Diagnostic Ultrasonography - Serous retinal detachments usually appear as smooth, dome-shaped collections of subretinal fluid. Fluid shifting can also be appreciated by ultrasonography. Associated macrostructural pathology, such as choroidal tumors or thickening of the posterior sclera (e.g., the “T” sign in posterior scleritis; )can be seen. Diffuse choroidal thickening can be seen in both VKH disease and SO.
Optical Coherence Tomography - OCT can differentiate between detachment of the retina and that of the RPE, distinguish retinal detachments from schisis cavities, establish the presence of subretinal exudation or neovascular membranes, identify discontinuities in the RPE, and monitor the resolution of subretinal fluid during treatment. Recently, modification of this imaging technique through enhanced depth imaging–OCT (EDI-OCT) has been used to evaluate choroidal changes in various diseases, such as ICSC and VKH disease.

Fluorescein Angiography and Indocyanine Green Angiography - Critical in elucidating the different patterns of posterior segment disease, as well as in providing in vivo evidence of the likely pathological mechanisms, as seen in recent ICGA studies of ICSC showing choroidal hyperpermeability underlying areas of serous detachment. These imaging techniques help distinguishing various diseases, identify specific areas of leakage, and can direct focal laser treatment and PDT, if necessary.

Optical Coherence Tomography Angiography - OCTA can generate a rapid, noninvasive angiographic picture of the retinal vessels and the underlying choroidal vasculature, as well as abnormal vascular patterns, such as neovascular membranes.

Computed Tomography and Magnetic Resonance Imaging - The broader imaging modalities of CT and MRI can also be used to evaluate posterior ocular and retrobulbar pathology associated with serous retinal detachment, such as with orbital cellulitis. Appropriate use of contrast in these studies can improve evaluation of inflammatory and neoplastic disease.

TREATMENT
Treatment of serous retinal detachments must address the underlying local or systemic disease, which includes - Antibiotic therapy (retinal and choroidal infections) like pyrimethamine and sulphanilamide for toxoplasmosis. For CMV, oral valganciclovir 900 mg twice daily for 3 weeks for induction and 900 mg daily for maintenance and intravitreal injection of ganciclovir. Ganciclovir slow release intravitreal implants are less commonly used and has efficacy of 8 months.

ATT to be used in tubercular cases, topical and systemic steroids may be used concomitantly to reduce inflammation induced damage and laser may be applied for ischemic areas. Penicillin is used in syphilitic cases under the supervision of infectious disease specialist and topical and systemic steroids may be given in conjunction to ameliorate inflammation. Oral doxycycline, amoxycillin or erythromycin is useful for lyme disease and steroids should be given undercut of antibiotics.

Cat scratch fever requires oral cotrimoxazole, azithromycin, rifampicin or ciprofloxacine. Steroids may be used in some cases. Antifungal agents like amphotericin B, vorconazole are used for fungal infections like Cryptococcus and coccidiomycosis, intravitreal agents may also be required in resistant cases. Intravenous antibiotics are used in orbital cellulitis particularly ceftriaxone and fluocoxacilin.

Systemic immunosuppression (posterior scleritis, vasculitides, autoimmune disease like VKH, sympathetic ophthalmitis idiopathic frosted branch angiitis) like high dose steroids (1-2mg/kg/day tapered over 4-6 months, this may be preceded by intravenous methylprednisolone pulse therapy (500-1000mg/day) along with topical steroids. Steroid resistant cases may require immunosuppressives such as methotrexate, azathioprine, biological blockers such as infliximab. Intravitreal and periocular corticosteroids are also used for treating inflammatory diseases. Timely consultation with other medical specialists, when indicated to initiate treatment, is critical in connective tissue disorders and other systemic disorders.

Blood sugar or blood pressure management.
In some diseases (e.g., ICSC), simple observation is sufficient.

Laser photocoagulation in the treatment of vascular anomalies (Coats’ disease) and tumors (capillary hemangiomias). In ICSC that does not resolve spontaneously, focal laser can be applied to sites of RPE leakage that are sufficiently distant from the fovea. Laser can also be applied to well-circumscribed choroidal hemangiomias, although external beam radiotherapy is indicated for more diffuse lesions.

PDT with verteporfin has been used recently in ICSC, most especially the chronic form with demonstrated improvement. The use of intravitreal anti-VEGF therapy can be valuable in the treatment of exudative AMD, Coats’ disease, and choroidal melanomas and hemangiomasketologies of serous retinal detachment.

Benign lesions require careful observation, choroidal melanomas are treated with brachytherapy when tumor is less than 20 mm in basal diameter, other treatment modalities include external beam therapy, stereotactic radiotherapy, transpupillary thermos therapy uses infrared beams trans-scleral chorioretectomy for thick tumors that cannot be treated with radiotherapy, and enucleation. Retinoblastoma is treated in same manner but chemotherapy is the mainstay of treatment in most cases which includes carboplatin, etoposide and vincristine(CEV).
Finally, in certain cases of serous retinal detachment, surgical intervention like vitrectomy is warranted. The creation of scleral windows in uveal effusion syndrome serves as one example of a primary surgical intervention.

**COURSE AND OUTCOME**

As a rule, resolution of subretinal fluid occurs with observation or appropriate treatment of the underlying local or systemic pathology. However, the longer a serous detachment is present, the less likely it is that full resolution and, more importantly, restoration of visual function can be achieved. Involvement of the macula also portends a poorer outcome. Visual outcomes with serous detachment tend to be better than with chronic rhegmatogenous and tractional detachments, and this can help to reassure patients as appropriate treatment is initiated.

**References**