

A 49 Year Old Male with Atypical Central Serous Chorioretinopathy Presenting as Bilateral Exudative Retinal Detachment

Michael F Fernandez*

Retina Fellow Adviser, Dr. Jocelyn L. Sy, University of Santo Tomas Hospital Eye Institute, Philippine

***Corresponding Author:** Michael F Fernandez, Retina Fellow Adviser, Dr. Jocelyn L. Sy, University of Santo Tomas Hospital Eye Institute, Philippine.

Received: March 13, 2020; **Published:** May 29, 2020

Abstract

A 49 year-old male consulted because of blurring of vision of left eye on January 2015 and was assessed with swelling of posterior part of OS. He was advised to rest but didn't comply. Interval history showed involvement the right eye with tinnitus which led consult to retina specialist. Assessment was Posterior Uveitis with Exudative RD OS. Oral steroids 60 mg/day was started which provided no improvement. On May 2015, he was referred to another retina specialist and repeat FA showed leakage at midphase but was signed out: t/c VKH, inferior detachment OS. VA that time was 20/80 for right eye and 20/200 for left eye. He was given 65 mg/day and intravitreal steroid device but without improvement. On June 2015, he was referred to Uveitis specialist who confirmed VKH. VA was OD:20/200 and OS:CF. Methotrexate 7.5 mg/week and periorbital Depomedrol 40mg/ml were added to treatment. Subretinal scarring and granulomatous lesions were noted hence, referral to another retina specialist and uveitis specialist was done. Assessment was multifocal CSR with exudative retinal detachment OU. VA during this time was 20/400 for right eye and LP for left eye. Repeat FA showed chronic CSR, multiple neurosensory detachment with exudative retinal detachment OD and retinal detachment with proliferative vitreoretinopathy with subretinal fibrosis OS. OCT of macula showed neurosensory detachment and intraretinal edema without choroidal folds and subretinal septae. Steroids were discontinued and topical nsaid, acetazolamide 125 mg BID, azathioprine 50 mg TID were started. Improvement of vision from 20/400 to 20/80 and resolution of exudative retinal detachment were noted 23 days after discontinuing steroid. Left eye underwent pars plana vitrectomy with endolaser and silicon oil. Twenty-three days after operation, VA for OS was 20/400 with +5.00D.

Keywords: *Vogt Koyanagi Harada Disease; Central Serous Retinopathy; Exudative Retinal Detachment; Blurring of Vision; Pars Plana Vitrectomy*

Introduction

Vogt Koyanagi Harada Disease is an uncommon multisystem disease of presumed autoimmune etiology that is characterized by chronic, bilateral, diffuse, granulomatous panuveitis with accompanying integumentary, neurologic, and auditory involvement in which the etiology and pathogenesis of VKH disease are still unknown. Current clinical and experimental evidence suggest a cell-mediated autoimmune process driven by T lymphocytes directed against self-antigens associated with melanocytes of all organ systems in genetically susceptible individuals. The clinical features of VKH disease also vary depending on the stage of the disease namely prodromal, acute uveitic, convalescent and chronic recurrent stages. A patient may present with sequential blurring of vision in both eyes 1 - 2 days after the onset of CNS signs. It is marked by bilateral granulomatous anterior uveitis, vitritis, thickening of posterior choroid with elevation

Citation: Michael F Fernandez. "A 49 Year Old Male with Atypical Central Serous Chorioretinopathy Presenting as Bilateral Exudative Retinal Detachment". *EC Ophthalmology* 11.6 (2020): 16-24.

of peripapillary retinal choroidal layer, hyperemia and edema of optic nerve and multiple serous retinal detachments. The focal serous retinal detachments are usually shallow detachments with cloverleaf pattern at the posterior pole, but may coalesce and evolve into large bullous exudative detachments. The diagnostic criteria for VKH is shown below in table 1.

Table 6-6 Revised Diagnostic Criteria for Vogt-Koyanagi-Harada Syndrome
<p>Complete Vogt-Koyanagi-Harada syndrome</p> <p>I. No history of penetrating ocular trauma or surgery</p> <p>II. No clinical or laboratory evidence of other ocular or systemic disease</p> <p>III. Bilateral ocular disease (either A or B below must be met):</p> <p>A. Early manifestations</p> <ol style="list-style-type: none"> 1. Diffuse choroiditis as manifested by either: <ol style="list-style-type: none"> a. Focal areas of subretinal fluid, or b. Bullous serous subretinal detachments 2. With equivocal fundus findings, then both: <ol style="list-style-type: none"> a. Fluorescein angiography showing focal delayed choroidal perfusion, pinpoint leakage, large placoid areas of hyperfluorescence, pooling of dye within subretinal fluid, and optic nerve staining b. Ultrasonography showing diffuse choroidal thickening without evidence of posterior scleritis <p>B. Late manifestations</p> <ol style="list-style-type: none"> 1. History suggestive of findings from IIIA, and either both 2 and 3 below, or multiple signs from 3 2. Ocular depigmentation <ol style="list-style-type: none"> a. Sunset glow fundus, or b. Sugiura sign 3. Other ocular signs <ol style="list-style-type: none"> a. Nummular chorioretinal depigmentation scars, or b. RPE clumping and/or migration, or c. Recurrent or chronic anterior uveitis <p>IV. Neurologic/auditory findings (may have resolved by time of examination):</p> <ol style="list-style-type: none"> A. Meningismus B. Tinnitus C. Cerebrospinal fluid pleocytosis <p>V. Integumentary findings (not preceding central nervous system or ocular disease)</p> <ol style="list-style-type: none"> A. Alopecia B. Poliosis C. Vitiligo <p>Incomplete Vogt-Koyanagi-Harada syndrome Criteria I to III and either IV or V from above</p> <p>Probable Vogt-Koyanagi-Harada syndrome Criteria I to III from above must be present Isolated ocular disease</p>

Adapted from Read RW, Holland GN, Rao NA, et al. Revised diagnostic criteria for Vogt-Koyanagi-Harada disease: report of an international committee on nomenclature. *Am J Ophthalmol.* 2001;131(5):647-652.

Table 1: Diagnostic criteria for VKH.

The acute stage of VKH disease is exquisitely responsive to early and aggressive treatment with topical, periocular, and systemic corticosteroids and cycloplegic and mydriatic agents. Initial dosages typically are 1.0 - 1.5 mg/kg/ day of oral prednisone or 200 mg of intravenous methylprednisolone for 3 days followed by high-dose oral corticosteroids, although the route of administration has no demonstrable effect on changes in visual acuity or the development of visually significant complications.

Central serous retinopathy (CSR) also known as central serous chorioretinopathy or choroidopathy (CSC), is an idiopathic condition characterized by the development of a typically well-circumscribed, serous detachment of the sensory retina. Different risk factors include type A personality, psychological stress, pregnancy and hypercortisolism. Exogenous administrations of steroids and endogenous hypercortisolism are also included as predisposing factors. It occurs primarily in otherwise healthy men between 25 and 55 years of age. The detachment results from altered barriers and deficient pumping functions at the level of the retinal pigment epithelium (RPE), even though the primary pathology may involve the choriocapillaris. Symptomatic patients describe the sudden onset of blurred and dim vision, micropsia (objects appear smaller than they are), metamorphopsia (objects appear distorted), paracentral scotoma, or decreased color vision accompanied by a migraine-like headache. In general, visual acuity ranges from 20/20 to 20/200, but in most patients it is better than 20/30 sometimes. Most (80% - 90%) eyes with CSC undergo spontaneous resorption of subretinal fluid within 3 - 4 months;

Citation: Michael F Fernandez. "A 49 Year Old Male with Atypical Central Serous Chorioretinopathy Presenting as Bilateral Exudative Retinal Detachment". *EC Ophthalmology* 11.6 (2020): 16-24.

recovery of visual acuity usually follows but can take up to 1 year. Laser photocoagulation at the site of fluorescein leakage can induce rapid remission; resorption of subretinal fluid may occur within several weeks of therapy. Another option for the treatment of CSC, especially when the leakage site is too close to the center of the fovea, is the use of verteporfin photodynamic therapy (PDT). On rare occasions, atypical CSR can present with bullous exudative RD, usually associated with multiple large retinal pigment epithelial detachments that are often hidden beneath a cloudy subretinal serofibrinous exudate.

This case report highlights that Central Serous Retinopathy and Vogt Koyanagi Harada disease can both present with gradual painless unilateral blurring of vision accompanied by exudative retinal detachment. VKH disease and acute central serous retinopathy are two common disorders with serous retinal detachment caused by dysfunction of choroid. However, VKH is an inflammatory disease while CSR occurs due to vascular compromise in choroid. The treatment of VKH and CSR are opposite. High dose systemic corticosteroid therapy is required in VKH but forbidden in CSCR. Failure in differentiating CSCR from VKH diseases may result in inappropriate use of corticosteroids, leading to exacerbation of disorder.

Case Report

A previously well 49 year-old male consulted last January 2015 because of 1 month history of gradual blurring of vision of left eye. He was assessed with swelling of posterior part of the involved eye. He was advised to rest, however, he didn't comply. On March 2015, fluorescein angiography was requested which showed the following results: multiple areas of inkblot expansile dye leakage seen over the superior nasal area and para macular area of the left eye during midphase (Figure 1 and 2).

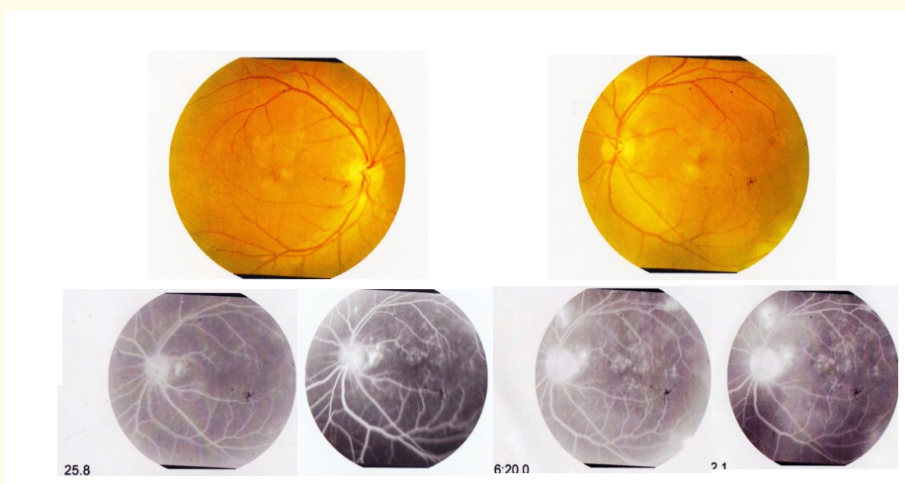


Figure 1: FA on March 2015 showing multiple areas of inkblot expansile dye leakage at midphase.

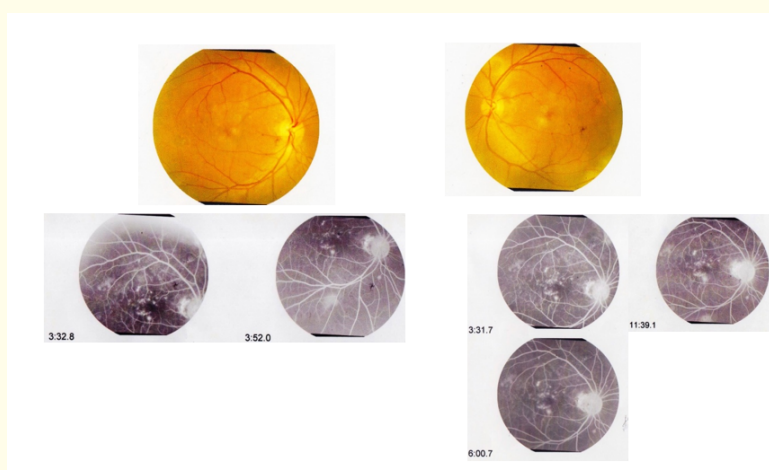


Figure 2: FA on the right eye showing dot leakage over the inferior perifoveal area and paramacular area.

Interval history showed persistence of symptom but now accompanied by tinnitus which led to consult to a retina specialist in Cebu where he was assessed with Posterior Uveitis with Exudative Retinal Detachment OS. Oral steroids 60mg/day was started which provided no improvement. Documented visual acuity that time was 20/80 for right eye and 20/200 for left eye. He noted persistence of blurring of vision now affecting the right eye. The use of intravitreal injection of slow releasing steroid drug was suggested however the patient refused.

On May 2015, he consulted another retina specialist in Manila who suggested a repeat Fluorescein Angiogram that showed increased areas of dot hyperfluorescence with expanding dye leakage during Mid AV phase but was signed out as: Posterior Uveitis, to consider Vogt-Koyanagi-Harada disease with inferior detachment of Left eye (Figure 3 and 4). Another cycle of steroid treatment was started with prednisone 65mg/day. Intravitreal steroid device was also given but didn't provide improvement. Comparison of FA done in March and May showed more areas of leakage seen at midphase (Figure 5).

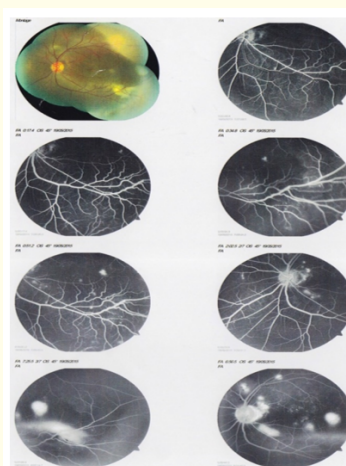


Figure 3: May 2015 FA showing leakage at Mid Phase, not in early phase.

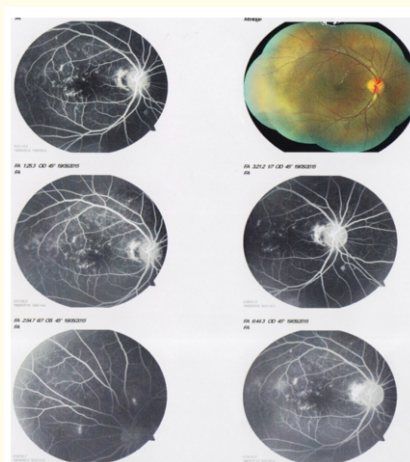


Figure 4: May 2015 FA showing similar dot hyperfluorescence.

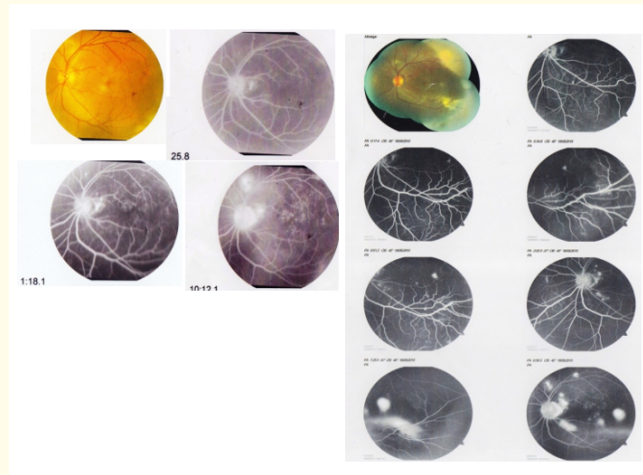


Figure 5: FA comparison of March(left) and May(right) studies showed more areas of dot leakage seen during midphase.

On June 2015, he was referred to a Uveitis specialist who confirmed Vogt Koyanagi Harada disease as the diagnosis. Visual acuity that time was 20/200 for the right eye and counting fingers for the left. Methotrexate 7.5 mg/week and periorbital methylprednisolone acetate injection of 40 mg/ml were added to treatment. On subsequent follow up, fundus examination revealed subretinal scarring and granulomatous lesions, which led to referral to other retina and uveitis specialists. Visual acuity during this time was 20/400 for right eye and light perception for left eye. After reviewing the history and doing ophthalmologic examination, he was then assessed with Multifocal Central Serous Retinopathy with Exudative Retinal Detachment OU (see fundus photo below in figure 6).

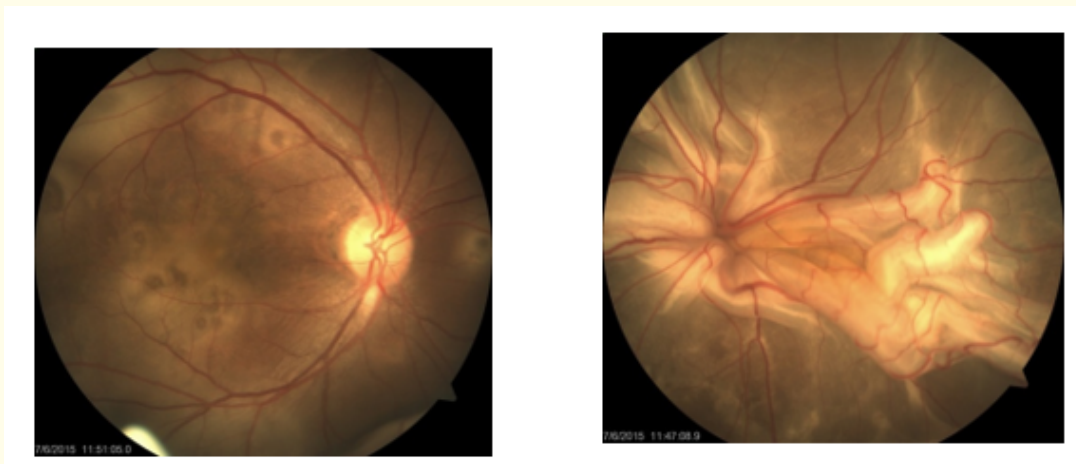


Figure 6: Fundus Photo upon consult to Uveitis and Retina specialists in Manila, assessed to have atypical central serous retinopathy.

Repeat fluorescein angiogram was requested and showed chronic Central Serous Retinopathy, multiple neurosensory detachments with exudative retinal detachment OD and retinal detachment with proliferative vitreo-retinopathy with subretinal fibrosis OS (See figure 7). OCT of macula showed neurosensory detachment and intraretinal edema without choroidal folds and subretinal septae (See figure 8).

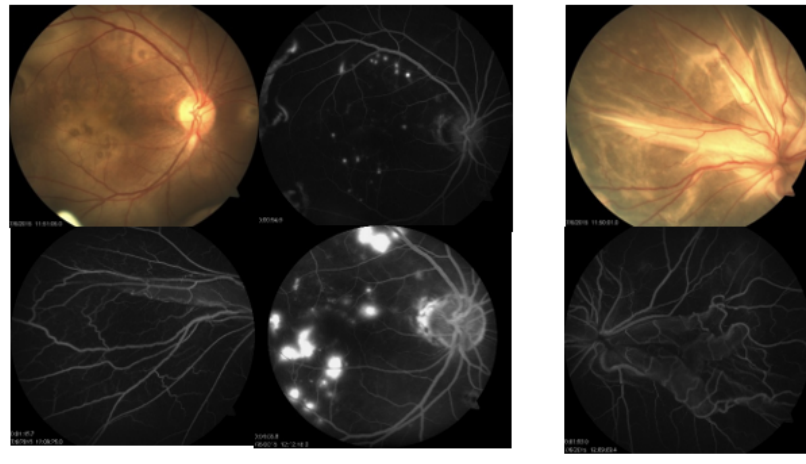


Figure 7: Figure 7a: FA of right eye showing chronic Central Serous Retinopathy, multiple neurosensory detachments with exudative retinal detachment. Figure 7b: FA of left eye showing retinal detachment with PVR with subretinal fibrosis.

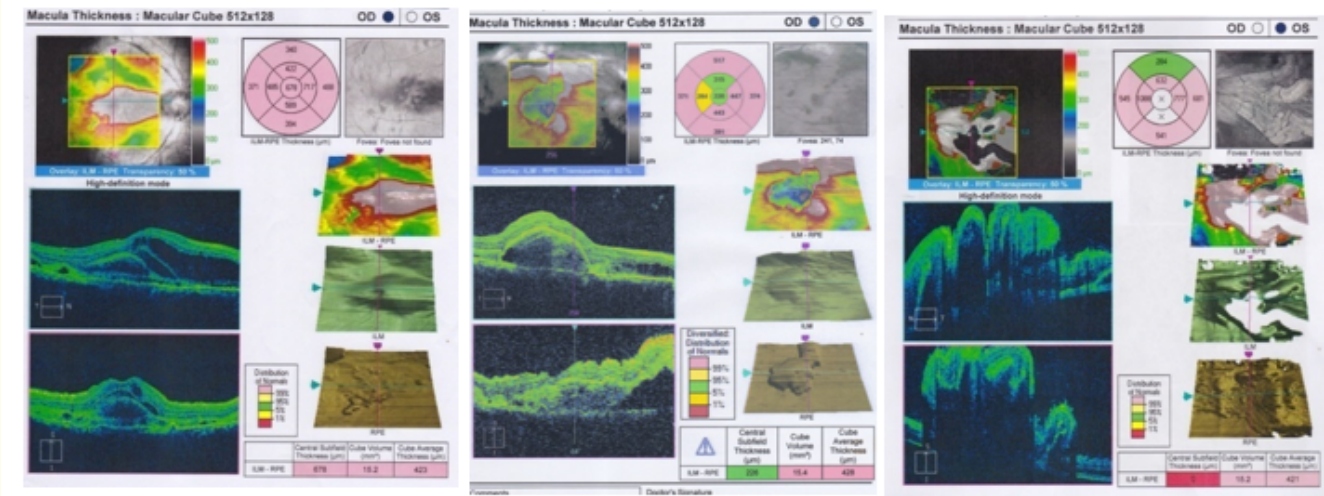


Figure 8: OCT macula: neurosensory detachment and intra retinal edema, note no choroidal folds seen, no sub retinal septa.

Uveitis work-up was done and showed negative results for CBC, Toxoplasma titers, RF, ANA, PPD, chest x ray except for T spot that showed positive results, hence referral to a pulmonologist was made. Due to progression of exudative retinal detachment extending to nasal and superior area of the retina, steroids were discontinued. Topical NSAIDs, acetazolamide 125 mg BID, azathioprine 50 mg TID were started (See figure 9).

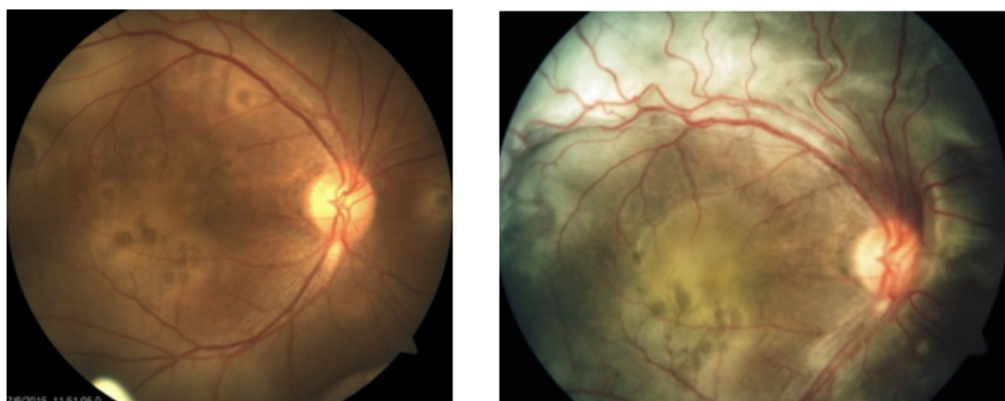


Figure 9: Progression of exudative retinal detachment.

Improvement of vision from 20/400 to 20/80 and resolution of exudative retinal detachment were noted 23 days after discontinuing steroid (Figure 10).

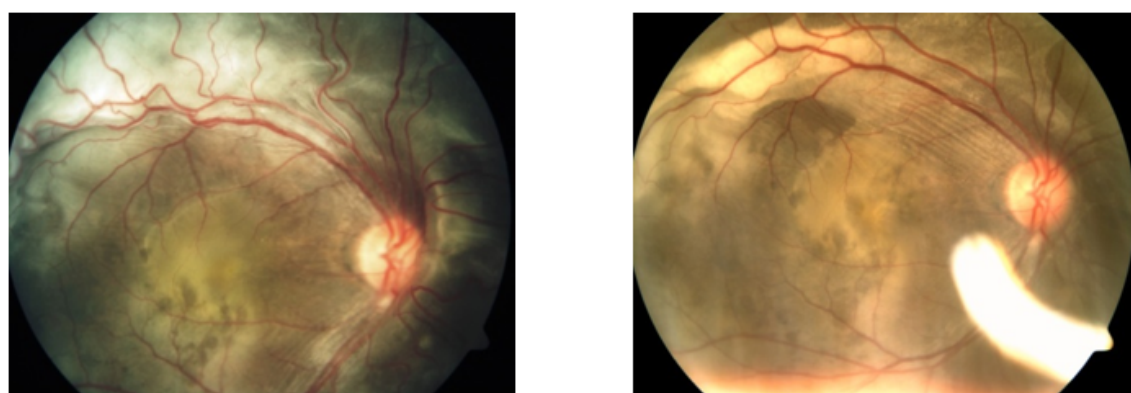


Figure 10: Improvement of exudative retinal detachment 23 days after discontinuing steroids.

The patient's left eye underwent pars plana vitrectomy with endolaser and silicon oil injection which showed improvement of visual acuity 23 days after operation which was 20/400 with +5.00D (Figure 11).

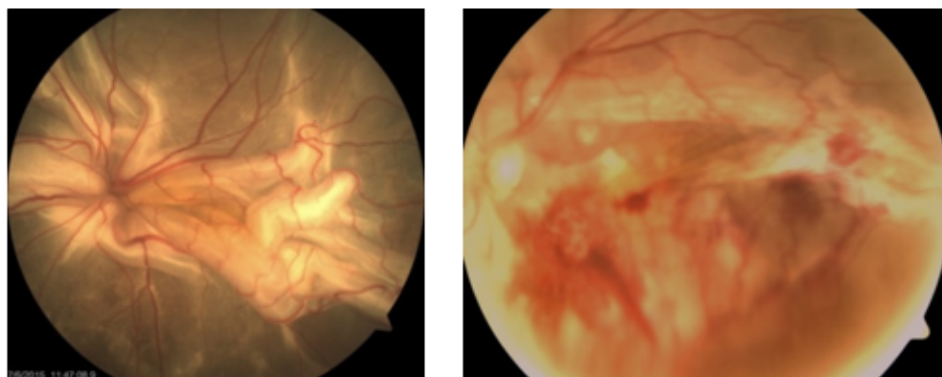


Figure 11: 23 days Post Operation of Pars plana vitrectomy with endolaser and silicon oil injection.

Discussion

Presentation of Central Serous Retinopathy may be atypical that can lead to misdiagnosis of various forms of uveitis such as Vogt Koyanagi Harada disease, sympathetic ophthalmia, posterior scleritis, multifocal choroiditis, serpiginous choroiditis, idiopathic posterior uveitis, or other uveitic entity that all usually require corticosteroid treatment. Unusual findings can occur including acute bullous retinal detachment, subretinal fibrin, subretinal fibrosis, hard exudates, and even retinal neovascularization. Atypical and chronic or recurrent forms of CSCR may be overlooked or misdiagnosed as chorioretinal inflammatory conditions, leading to inappropriate use of corticosteroids because they are not only ineffective, but they usually exacerbate the condition, leading to bilateral, severe, and chronic CSR with multifocal RPE detachments or diffuse retinal pigment epitheliopathy. This usually results in worsening of CSR and irreversible chorioretinal damage and visual impairment.

A careful clinical examination and history taking are mandatory to differentiate CSR from other differential diagnosis like VKH disease. Based on the revised diagnostic criteria for VKH disease, appropriate laboratory findings are needed to rule out other ocular or systemic diseases (See table 1 in appendix). In addition, the occurrence of tinnitus is not pathognomonic to have an assessment of VKH and the use of steroids should have not worsened the illness if it is a type of uveitis. VKH syndrome is considered to cover a wide spectrum of diseases affecting the eyes, meninges, skin, and audio vestibular system that manifest in symptoms of discuses, tinnitus, headache, vomiting, and flu-like symptoms for several weeks before the onset of ocular symptoms, vitiligo, poliosis and alopecia during the convalescent stage. Clinical findings should be correlated with FA findings (like delayed choroidal perfusion, pinpoint leakage, large placoid areas of hyperfluorescence) and ultrasound findings (diffuse choroidal thickening).

In patients with CSR exhibiting unusual and atypical findings, the use of multimodal imaging, including OCT, FA and ultrasonography, can provide us clues for the definitive diagnosis.

On FA, an expansile dot pattern of hyperfluorescence is the most common presentation in which the dot represents a small, focal, hyperfluorescent leak from the choroid through the RPE that appears in the early phase and increases in size and intensity in late phase. In some patients, multiple pinpoint leakage may be present. In rare cases just like in this patient, an extensive, often gravity-dependent, serous detachment of the retina may develop from one or more leakage points outside the central area or may be associated with a diffuse pattern of fluorescein leakage, often without any prominent leakage point. In Vogt-Koyanagi-Harada syndrome, the yellow-white lesions can appear similar to CSR and multiple pinpoint leakages on FA; however, the granulomatous uveitis present in the former helps differentiate the diseases which was not present in this patient.

On OCT, although both diseases may have some similarities in terms of having subretinal fluid, they also have distinct features that allow physicians to differentiate them. Pigment epithelial detachments were more common in CSR. Retinal pigment epithelial bulge defined as a small protrusion along the RPE layer was seen only in CSR. On the other hand, the presence of folds of RPE, fluctuations in internal limiting membrane and subretinal septae were only present in VKH (Figure 12). Lastly, the absence of vitreoretinal traction or choroidal thickening or infiltration associated with retinal detachment on ultrasonography in CSR.

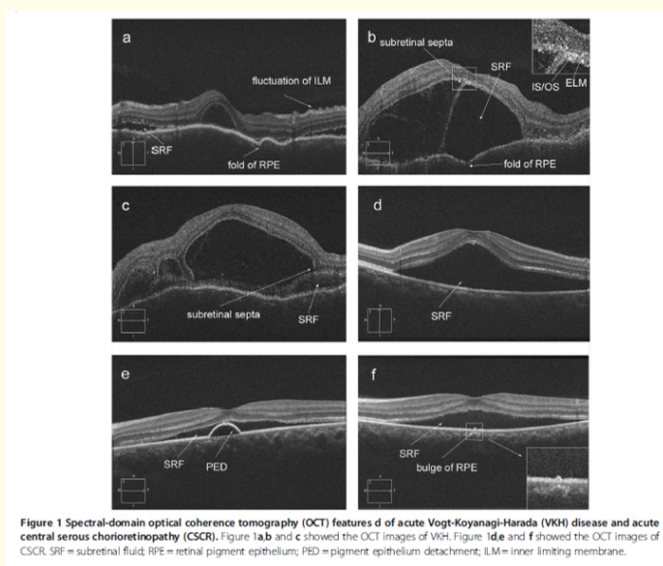


Figure 12: OCT findings in VKH and CSR.

For the natural course and management of CSR, most patients with CSR undergo spontaneous resorption of subretinal fluid within 3 - 4 months but recovery of visual acuity may take up to 1 year. However, stress can be a precipitating factor in our case since the patient didn't comply when he was advised to rest after being diagnosed with CSR that led to worsening of the disease. This also led to the misdiagnosis and mismanagement as a case of uveitis. In patients with corticosteroid-induced or worsened CSR, discontinuation of corticosteroids should be the first management step. Many cases actually improved following discontinuation of corticosteroid therapy with resolution of subretinal detachment, decreasing or disappearing of visual symptoms and improvement of visual acuity. Laser photocoagulation at the site of leakage can induce rapid resorption of subretinal fluid within 5 weeks of therapy but with no evidences of better visual acuity than in untreated eyes. Photodynamic therapy can be another option when the lesion is near the fovea. Unfortunately, CSR in this patient presented as an exudative retinal detachment where the subretinal fluid must be drained in order to carry out the appropriate therapy. Technique used in this case was an internal drainage using pars plana vitrectomy with endolaser photocoagulation and silicon oil injection that provided improvement in visual acuity of the patient to 20/400 with correction [1-6].

Conclusion

Aside from good clinical history and examination, there are multimodal imaging findings that may help us differentiate atypical central serous retinopathy from VKH to prevent inappropriate use of corticosteroids. FA patterns for CSR are the following: 1) expansile dot, 2) smokestack and 3) diffuse pattern while patterns for VKH can be multifocal leakages due to multiple sensory detachments but with signs of having granulomatous uveitis together with findings in the diagnostic criteria for the disease. For OCT, retinal pigment epithelial bulge occurs in CSR while folds in RPE, fluctuations in ILM and subretinal septa are more common in VKH.

Bibliography

1. "Intraocular inflammation and Uveitis". Section 9 AAO Basic Science and Clinical Course (2014-2015): 183.
2. "Retina and Vitreous". Section 12 AAO Basic Science and Clinical Course (2014-2015): 171.
3. Rim Kahloun., *et al.* "Multimodal Imaging for the Diagnosis of an Atypical Case of Central Serous Chorioretinopathy". *Middle East African Journal of Ophthalmology* 21.4 (2014): 354-357.
4. M Gemenetzi., *et al.* "Central serous chorioretinopathy: an update on pathogenesis and treatment". *Eye* 24.12 (2010): 1743-1756.
5. Dusheng Lin., *et al.* "Comparison of the optical coherence tomographic characters between acute Vogt-Koyanagi-Harada disease and acute central serous chorioretinopathy". *BMC Ophthalmology* 14 (2014): 87.
6. Eun Kang., *et al.* "Surgical Management of Bilateral Exudative Retinal Detachment Associated with Central Serous Chorioretinopathy". *Korean Journal of Ophthalmology* 20.2 (2006): 131-138.

Volume 11 Issue 6 June 2020

© All rights reserved by Michael F Fernandez.