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Inflammatory Masquerades of CSCR

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Abstract: ***Purpose: To reinforce clinical examination and demonstrate the clinical utility of OCT to differentiate Central serous chorioretinopathy (CSCR) and its inflammatory masquerades Synopsis – Two cases diagnosed outside as CSCR - 53-year-old male (unilateral) and 31-year-old female (bilateral) were evaluated in our retina department. On detailed evaluation and OCT imaging they were identified as inflammatory conditions masquerading as CSCR. Patients were started on Oral steroids with the resolution of the lesion as well as improvement in vision. In Some unusual situation's inflammatory conditions like Vogt-Koyanagi-Harada syndrome posterior uveitis posterior scleritis white dot syndromes can mimic CSCR. It is essential to differentiate the two conditions as the treatment for the two conditions is exactly the opposite. OCT features (ILM folds, sub retinal septa, RPE folds, RPE undulation index, hyperreflective foci in choroid) in conjunction with clinical examination gives diagnostic clues and helps in differentiating CSCR and inflammatory conditions.***

Keywords: VKH syndrome, Posterior scleritis, CSCR, OCT

1. Introduction

Central serous chorioretinopathy (CSCR) is characterized by localized serous detachments of the neurosensory retina with or without focal pigment epithelial detachments (PEDs) and altered retinal pigment epithelium (RPE). In Some unusual situations, inflammatory conditions like Vogt-Koyanagi-Harada syndrome, posterior uveitis, posterior scleritis, white dot syndromes can mimic CSCR especially when they have been partly treated prior to presentation to OPD.(1) In this case report, we present 2 cases of inflammation of ocular coats mimicking CSCR. We wish to highlight inflammatory masquerades of CSCR and relevant review of literature which can help us to differentiate the two clinical conditions.

2. Case 1

A 53-year-old male patient was referred to retina clinic as a case of CSCR. He gives history of painless diminution of vision in right eye (OD) for 1 week. Best corrected visual acuity (BCVA) in right eye and left eye (OS) was 6/9, and 6/6 respectively. Anterior segment examination in both eyes was within normal limits. Fundus examination of OD showed neurosensory detachment (NSD) of 1 disc diameter inferior to the superior arcade along with choroidal folds while OS fundus was within normal limits. OCT of OD showed multiple pockets of NSD along with folds of internal limiting membrane (ILM) and choroidal elevations. Ultrasound (USG) examination of OD showed fluid in sub-tenons space. Based on clinical features and imaging we revised the diagnosis as OD posterior scleritis and started the patient on oral Indomethacin 75 mg for 3 weeks. At 3 weeks follow-up patient presented with improvement of vision to 6/6. Fundus examination showed collapse of NSD with deposition of hard exudates at macula supporting our clinical diagnosis. OCT at follow-up visit showed normal inner retinal layers, few hyper reflective dots in outer plexiform layer suggestive of hard exudates along with decrease in size of NSD and normal choroid. (Figure 1)

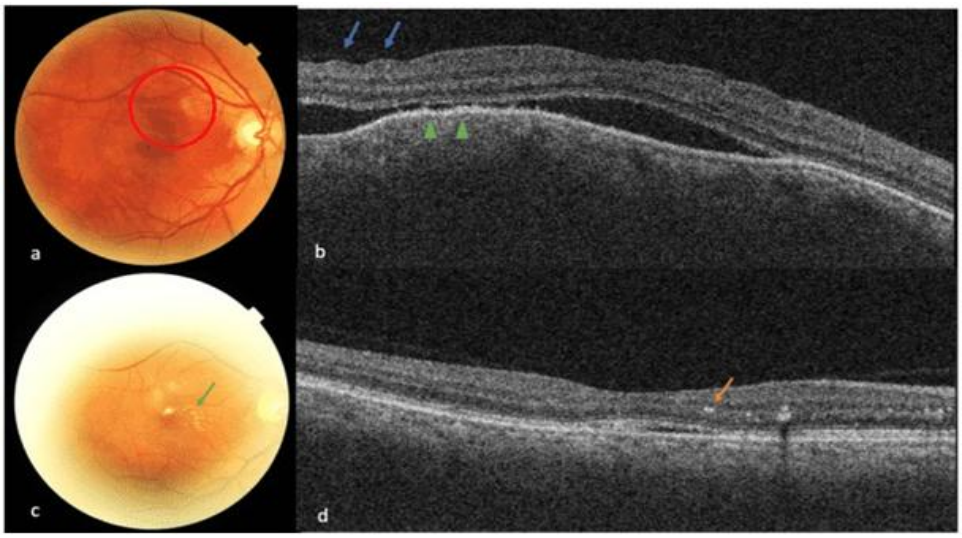

Figure 1: a. Fundus photos of right eye showing NSD with Choroidal folds (Red circle), b. OCT of RE passing through the lesion showing ILM fold (blue arrow), NSD and Choroidal folds (green arrow head), c. Fundus photo at follow-up showing resolution of NSD with hard exudates at macula (green arrow) d. OCT follow up showing resolution of NSD with hard exudates (orange arrow)

3. Case 2

A 31-year-old female patient who is a known diabetic and hypertensive was referred to retina OPD as a case of CSCR. She gives history of painless diminution of vision in RE for 2 days. There is no history of trauma or previous eye surgeries. Her BCVA was 6/18 in RE and 6/6 in LE. Anterior segment examination was normal in both eyes. Fundus examination of OD showed a 2 DD NSD inferior to superior arcade along with ILM folds, while OS fundus was within normal limits. OCT of OD showed ILM folds, Neurosensory detachment along with RPE undulations. Our Provisional diagnosis was RE choroiditis. We advised blood investigations to look for cause of choroiditis. After 1 week, she presented with diminution of vision in both eyes to 6/60. Her blood investigations were normal. Anterior segment examination in both eyes was normal. Fundus examination revealed blurred disc margin, dilated blood vessels, multiple neurosensory detachments at posterior pole in both eyes. OCT showed ILM folds, Multiple pockets of neurosensory detachment along with subretinal septa, subretinal cysts, RPE undulations and hyper reflective dots in choroid. USG showed increased choroidal thickening. We revised the diagnosis to OU posterior uveitis - Probable Vogt-koyanagi- Harada's syndrome and started her on oral prednisolone 1mg per kg body weight and referred her to rheumatologist to start on steroid sparing immunomodulatory therapy. At 1 week follow-up her vision improved to 6/9 in both eyes. Fundus examination revealed normal optic disc and blood vessels, and collapsed NSD at posterior pole supporting our clinical diagnosis. Fig 2&3 shows clinical pictures and OCT features of right and left eyerespectively.

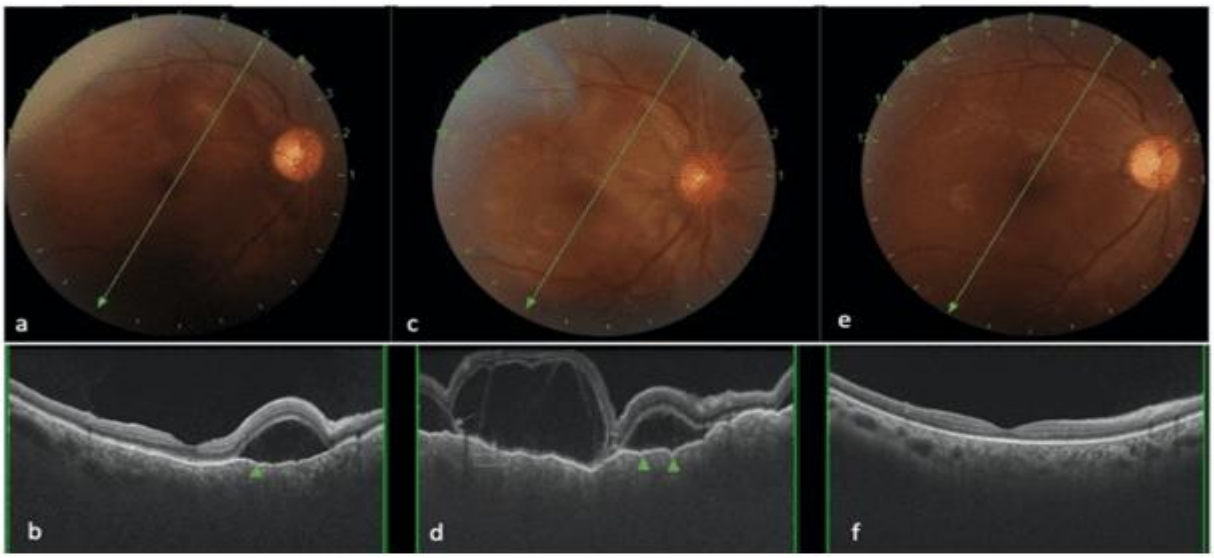

Figure 2: a. b Fundus photos and OCT of right eye at initial visit showing NSD and RPE folds (green arrow head), c, d. Fundus photo and OCT at 1 week sowing multiple pockets of NSD with RPE folds (green arrow head), subretinal septa (blue arrow) and hyper-reflective dots (blue rectangle). i. e. Fundus photo and OCT following steroid treatment showing resolution of NSD

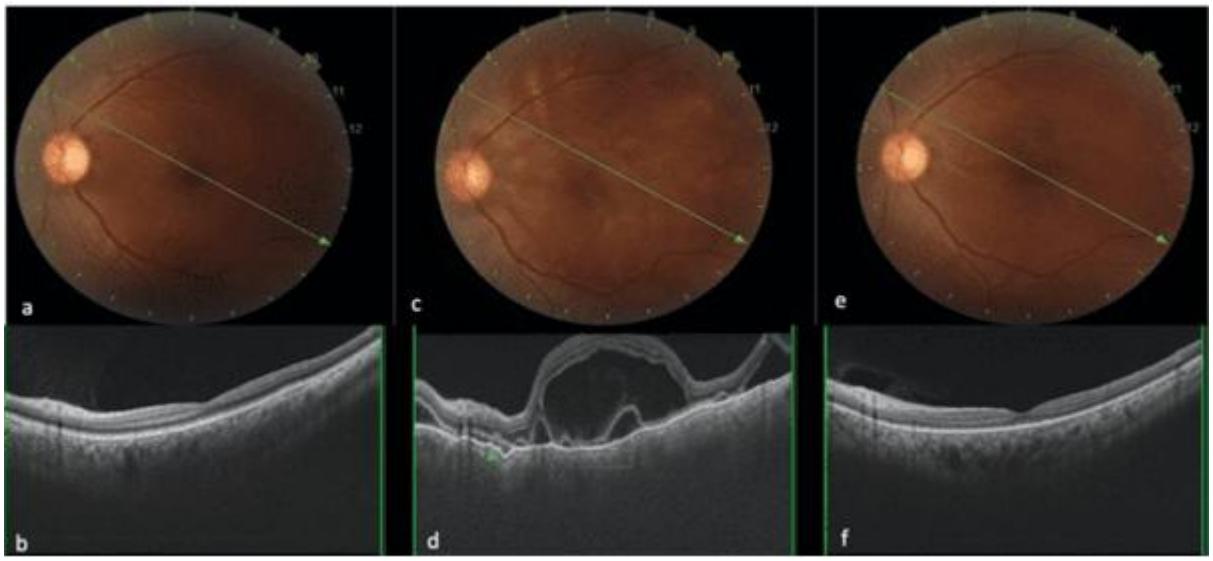

Figure 3: a. b Fundus photos and OCT of left eye at initial visit showing normal fudus and normal OCT c, d. Fundus photo and OCT at 1 week sowing multiple pockets of NSD with RPE folds (green arrow head), and hyper-reflective dots (blue rectangle). i. e. Fundus photo and OCT following steroid treatment showing resolution of NSD

4. Discussion

CSCR is a self-limiting clinical condition characterized by painless, sub-acute loss of vision in middle aged individuals and carries a good visual prognosis. However certain choroidal vascular conditions such as polypoidal choroidal vasculopathy, choroidal neovascular membrane; inflammatory conditions such as posterior scleritis, choroiditis; retinal vascular diseases such as retinal angiomatous proliferans and juxtafoveal telangiectasia; and few miscellaneous conditions such as optic disc pit and dome shaped macula can mimic as CSCR.

1) In our cases we highlight few inflammatory conditions mimicking as CSCR. It is essential to differentiate the two conditions as the treatment for the two conditions is exactly opposite.

In case one, patient presented with painless diminution of vision, without congestion or tenderness of anterior segment, and isolated neurosensory detachment without any disc edema mimicking as CSCR. However, on careful fundus examination we can see choroidal folds which is unusual in CSCR. In our case OCT showed NSD along with ILM folds and choroidal folds which is a red flag sign, which made us to perform an USG which revealed sub tenon fluid and confirmed our diagnosis. Posterior scleritis is painful inflammation of the sclera posterior to the ora serrata and is commonly misdiagnosed in 74% cases. Though pain is an important clinical feature, it can be absent in 21.7% of cases. (2) It can manifest as isolated serous retinal detachment in 28% of cases.(3) B-scan in posterior scleritis shows increase in posterior coats thickness, and T-sign in severe cases which is a pathognomonic of scleritis.(4)

In case two, patient initially presented with unilateral diminution of vision in RE for 2 days, with normal anterior segment, and a neurosensory detachment which can be misdiagnosed as CSCR. But OCT showed neurosensory detachment along with RPE folds indicating an inflammatory pathology. In an article by Lin et al (5) on comparison of OCT features of acute VKH and CSCR ILM folds, RPE folds have been identified as markers to exclude a case of CSCR. We advised blood investigations to rule out systemic disorders. At one week she presented with typical VKH features suggesting left eye to be in prodromal phase. Vogt-koyanagi-haradas disease is bilaterally asymmetric granulomatous uveitis, and is often associated with exudative retinal detachment, and with extraocular manifestations such as meningismus, tinnitus, alopecia, and vitiligo. Acute stage of the disease is confused with CSCR in about 22% cases. (6) VKH can be either complete, incomplete and probable according to revised diagnostic criteria for VKH.(7) All the three categories have absolute requirement of bilateral eye disease. However, a unilateral or delayed involvement of the other eye can occur in rare cases. The differentiating features of CSCR, posterior scleritis and VKH are summarized in Table 1.

To conclude, in a set-up where multimodal imaging is not available, OCT in conjunction with clinical examination gives diagnostic clues and helps in differentiating CSCR and inflammatory conditions.

Table 1: Clinical and OCT features to differentiate between CSCR, Posterior scleritis and VKH

	CSCR	**Posterior scleritis**	**VKH**
Age at presentation	Middle age usually in 40's	Middle age	2^{nd} – 5^{th} decade
Sex	Men	Women	Women
Anterior segment	Normal	Conjunctival congestion and tenderness (52.2%) (2)	Aqueous cells, flare
Symptoms	No pain	Pain (78.3%) (2)	Blurring of vision
Systemic associations	-	Present (17%) (8)	Poliosis, Vitiligo, alopecia Meningismus
Disc hyperemia	absent	present (43.4%) (8)	present
OCT features	• PED • Subretinal hyper-reflective dots • Double layer sign • Elongated photoreceptor outer segments • Increase in sub-foveal choroidal thickness	• ILM folds • RPE undulations • Choroidal elevations • Increase in choroid thickness • Hard exudates	• Subretinal septa (84.6%)(5) • Folds of RPE (67.7) (5) • ILM folds (52.3%)(5) • Increase in RPE undulation index

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