Central Serous Chorioretinopathy mimicking Idiopathic Uveal Effusion Syndrome

Abbreviated title: CSCR and Uveal Effusion Syndrome

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Summary Statement: Four eyes (3 patients) presented with an acute inferior exudative retinal detachment associated with an annular choroidal detachment mimicking uveal effusion syndrome. They had a history of central serous chorioretinopathy in the same eye or in the fellow eye. The role of choroidal venous congestion and scleral thickening is discussed.

Abstract

Purpose: To describe central serous chorioretinopathy (CSCR) cases presenting as uveal effusion syndrome, providing new insights into “pachychoroid spectrum” diseases.

Methods: Clinical charts, color fundus photographs, fluorescein angiography, indocyanine green angiography, optical coherence tomography, ultrasound imaging, cerebral magnetic resonance imaging and biometry of four eyes of three patients were assessed. A literature review was conducted.

Results: The three patients had peripheral choroidal detachment and inferior bullous retinal detachment associated with CSCR features detected using multimodal imaging, including fluorescein and indocyanine green angiography. The choroid was thick in the three patients, and uveal effusion occurred after steroid treatment in all cases. Subretinal fluid drainage and deep sclerectomy with flaps of 4x4 mm in both inferior quadrants were performed in 3 eyes of
2 patients with good outcomes. One patient was treated with photodynamic therapy. All three patients developed a typical leopard-spot pigmentary pattern in the fundus.

**Conclusion:** A severe presentation of highly exudative CSCR may occur in rare cases with a peripheral choroidal detachment mimicking uveal effusion syndrome. These severe cases highlighted the role of choroidal thickening and hyperpermeability, choroidal vein dilation and possible scleral thickening in both entities.

**Keywords:** Central serous chorioretinopathy, Choroidal detachment, Choroidal veins, Exudative retinal detachment, Pachychoroid, Uveal effusion syndrome

**Introduction:**

Uveal Effusion (UE) is defined as a “serous detachment of the peripheral choroid and ciliary body, associated with bullous serous detachment of the retina” \(^1\). It can be due to an inflammatory or hydrostatic condition. When no cause is identified, the term “Idiopathic uveal effusion syndrome” (IUES) is used. Central serous chorioretinopathy (CSCR) may also present, in rare cases, with an inferior bullous retinal detachment associated with subretinal fluid leakage but no ciliochoroidal detachment. We analyzed 4 eyes of 3 patients with an unusual aspect of IUES associated with typical signs of CSCR in the same eye and in the fellow eye.

**Material and methods**
Three patients were managed between 2004 and 2015 in the Ophthalmology Department of Lariboisière Hospital and at the Center for Imaging and Laser (Paris, France). Color fundus photographs, fundus autofluorescence (FAF), fundus fluorescein angiography (FA), indocyanine green angiography (ICGA), optical coherence tomography (OCT), ultrasound (US) imaging, optic biometry and orbital magnetic resonance imaging (MRI) of the four eyes of three patients were retrospectively assessed and compared. A literature review was conducted using the MEDLINE and Web of Science databases to identify previous associations between UE and CSCR. No search restriction was used. Four articles were identified but only one was relevant. Additional articles were also identified using wider requests (Uveal effusion syndrome: 171 results, Central serous chorioretinopathy: 2,193 results) and the reference list of the screened articles.

Case Reports:

Case 1: A 45-year-old man was referred for a four-month history of variable decreased vision in the right eye (RE). He was treated with salbutamol and corticosteroid spray for asthma. The best-corrected visual acuity (BCVA) was 20/32 in the RE and 20/20 in the left eye (LE). Fundus examination showed a 360° ciliochoroidal detachment in the RE associated with an inferior retinal detachment involving the posterior pole (Figure 1A). There was no vitritis, no papilledema, no vasculitis and no retinal tear. FA of the RE showed retinal pigment epithelium (RPE) clumping at the posterior pole and beyond, several leaking points in the posterior pole (Figure 1B). Areas of choroidal hyperpermeability were seen during the mid-phase of ICGA where the leaking points were on FA, with a more intense leakage and staining close to the temporal inferior vessels (Figure 1C). The OCT B-scan of the RE showed a choroid thickened at more than 840 µm subfoveally with a bumpy anterior surface. The RPE
presented extensive granulations, and the retina was slightly detached at the posterior pole. The retina was focally thickened, by a massive posterior hyperreflective edema and presented superficial retinal folds (Figure 1 F). The ultrasound B-scan and MRI confirmed the presence of an annular ciliochoroidal detachment and an inferior retinal detachment. The axial length was 23.9 mm.

In the LE, FAF showed hyperfluorescent gravitational tracts (Figure 1 G). Pigmentary changes in the posterior pole had an annular disposition, and two leaking points were seen on FA (Figure 1H). Multiple areas of choroidal hyperpermeability were seen on ICGA (Figure 1H). The choroidal thickness was 874 µm. Secondary causes of UE were ruled out such as an orbital mass or a scleritis. The medical and biological workups were normal, orbital MRI was performed to rule out inflammation or tumor infiltration. The biological workup included blood cell counts, and a simple analysis of inflammatory proteins. The patient did not develop any systemic disease during the 5-year follow-up. Inhaled steroids were discontinued, but the choroidal detachment and the retinal detachment did not improve. Subretinal fluid drainage and deep lamellar sclerectomy of 4x4 mm in both inferior quadrants was then performed. One month after surgery, the choroidal and retinal detachments had totally regressed. Four years after the onset, the final BCVA was stabilized to 20/63. The retina remained attached and RPE disturbances became widespread. ICGA better showed the presence of large choroidal veins, especially at the initial location of the leakage and fibrin (Figure 2).

Case 2: A 69-year-old man was diagnosed with chronic CSCR in both eyes since 2010. The patient had chronic obstructive pulmonary disease (COPD) requiring inhaled and nasal steroids. His RE was treated at this time with focal laser and photodynamic therapy (PDT). The LE also presented with alterations of the RPE at the posterior pole, including an atrophic gravitational track (Figure 3A). The OCT B-scan showed a flat irregular PED in the macula (Figure 3B). The situation remained stable, but
in 2014, an acute exacerbation of COPD required eight days of oral steroids. One week later, in the LE, an inferior bullous retinal detachment occurred, involving the macula, associated with an annular ciliochoroidal detachment (Figure 3C). There was no vitritis, no papilledema, no vasculitis and no retinal tear. The BCVA dropped to 20/100. FA showed several leaking points above the optic disc (Figure 3D) and ICGA showed large dilated choroidal veins surrounding the optic disc with an intense peripapillary late choroidal staining (Figures 3G and H). The OCT B-scan showed a choroidal thickening at 540 µm subfoveally (Figure 3E) and near the optic disc. Ultrasonography (US) and MRI confirmed the annular choroidal detachment and the retinal detachment in the LE (Figure 3F). There was no measurable scleral thickening, no orbital mass and no inflammation. The axial length was 22.6 mm in the RE and 22.7 mm in the LE. In the RE, the subfoveal choroid was also thickened at 440 µm.

A systemic workup ruled out intraocular lymphoma, orbital inflammation and tumor infiltration, Vogt-Koyanagi-Harada disease, and Waldenström's macroglobulinemia. Steroids could not be discontinued due to COPD severity, and one month later, the retinal detachment had increased in the LE, subretinal fibrosis had appeared above the optic disc as well as an extensive subretinal leopard-spot pigmentation. Full-dose PDT was then applied to the hyperfluorescent spots supero-nasal to the optic disc. The retinal and choroidal detachments progressively regressed and were resorbed at 6 months. No recurrence was observed over the next five years. The final BCVA was 20/63 in the RE and 20/80 in the LE.

Case 3: A 54-year-old man was referred for blurred vision in the RE. His medical and ocular history were unremarkable. The BCVA was 20/80 in the RE and 20/20 in the LE. Fundus examination of the RE showed an inferior bullous retinal detachment with shallow macular detachment (Supplemental Digital Content, http://links.lww.com/ICB/A137) and an annular choroidal detachment confirmed by ultrasonography. FA showed choroidal folds, many subretinal leaking points at the posterior pole and around the optic disc, and hyperfluorescent
pinpoints. ICGA showed choroidal vein dilation at the posterior pole, and a prolonged
hyperfluorescence of the temporal inferior vortex veins (Supplemental Digital Content). In the
LE, FA showed multifocal hyperfluorescent RPE scars with one leaking point supero-
temporal to the macula, and a small extra-macular PED. On ICGA, the RPE scars had a
typical necklace pattern above the optic disc. Mid-phase ICGA showed a hyperfluorescent
choroidal plaque at the leaking point seen on FA (Figure 4A and B). The features of the LE
were highly suggestive of CSCR but the diagnosis was not made at this time in the RE. The
axial length was 21.1 mm in the RE and 21.3 mm in the LE. A comprehensive assessment to
detect inflammatory diseases (including lumbar puncture, HLA typing and ear, nose, and
throat examination) was unremarkable.

Due to the very unusual aspect of the retinal and choroidal detachments in the RE, Harada's
disease could not be ruled out and the patient received high doses of steroids. The retinal
detachment increased and involved the macula. IUE syndrome was suspected and subretinal
fluid drainage and deep lamellar sclerectomy of 4x4 mm in both inferior quadrants were
performed. Six months after the onset, an inferior bullous retinal detachment and an annular
choroidal detachment also occurred in the LE (Figure 4 C, D, E) that showed RPE changes
attributable to CSCR six months earlier. Lamellar sclerectomy resulted in retinal flattening.
Thirteen years later, the leopard-spot pigmentation had evolved to RPE atrophic scars. BCVA
was limited to 20/400 in the RE but reached 20/25 in the LE. The patient did not develop any
systemic disease.

Discussion:

We described 4 eyes of 3 patients with an inferior exudative retinal detachment and an
annular ciliochoroidal detachment. In all cases, one or both eyes presented signs of chronic
CSCR or a history of CSCR. The eyes involved showed signs of subretinal leaking points on
FA, choroidal hyperpermeability on ICGA, severe choroidal thickening on OCT, and the retinal detachment was triggered or worsened by corticosteroid treatment.

Uveal effusion is a rare disease characterized by the association of an annular ciliochoroidal detachment and an inferior retinal detachment, typically shifting with the patient's position. Later, the term of IUES has been used to differentiate this condition from nanophthalmic UE. Only a few multimodal images of IUES are available in the literature. Our cases had an axial length ranging between 21.1 and 23.9 mm, ruling out the diagnosis of nanophthalmos. They shared with IUES the presence of an annular ciliochoroidal detachment and an inferior non-rhegmatogenous retinal detachment, but they showed on FA defined subretinal leaking points located above the limits of the inferior bullous retinal detachment while no fluorescein leakage has been reported in IUES. In addition, one eye had a known history of chronic CSCR, and in all cases the fellow eye presented with signs of either past or active CSCR. Also, as in IUES, our cases showed a "leopard-spot" appearance of the fundus characterized by a mottling hyperpigmentation at the RPE while "leopard spots" have also been reported in severe CSCR. In a recent series of 21 CSCR eyes with inferior bullous retinal detachment, no ciliochoroidal detachment was reported.

Our cases had the particularity of presenting the aspect of a bullous variant of CSCR but associated with an annular ciliochoroidal detachment that is specific of IUES. To our knowledge, only one case showing some similarities with our cases has been reported, but no OCT image and no image of the periphery were shown, and the authors have concluded to IUES rather than to CSCR.

IUES is thought to be due to a scleral thickening or an abnormal scleral structure that could lead to choroidal thickening through different mechanisms. First, the scleral thickening and stiffness could narrow the transscleral passage of the vortex veins, impairing the venous
outflow and leading to choroidal congestion. A second hypothesis is that the abnormal scleral thickness and structure could impede the normal flow of transscleral fluid through the sclera resulting in increasing fluid collection through an oncotic mechanism. CSCR publications have suggested that the choroidal vein dilation initially seen on ICGA could be due to an engorged vortex vein ampulla as seen in UES.

Partial thickness sclerectomy was performed in 3 eyes, because we still suspected a diagnosis of CSCR for which it has been reported to be effective in a case associated with inferior bullous retinal detachment. Lastly, it has been shown that CSCR occurs in emmetropic or moderately hypermetropic eyes, but not in myopic eyes. This is also the case for IUES, and our cases had an axial length ranging between 21.1 and 23.9 mm.

While the cause of pachychoroid in CSCR has not yet been elucidated, an exudation through choriocapillaris units in areas of dilated choroidal veins has often been suggested to explain the choroidal thickening and the loculation of the suprachoroidal fluid at the posterior pole in some CSCR cases. The fact that PDT targeted on choroidal plaques of hyperpermeability results in a diffuse reduction in choroidal thickness reinforces this hypothesis.

The management of this rare but severe retinal disease should include systematic medical and biological work-up, precise orbital imaging (MRI or US) and a close follow-up. A proper diagnosis of CSCR in these cases allows avoiding unnecessary or deleterious corticosteroid treatment. However, partial posterior sclerectomy could be an effective treatment as in IUES.

We were not able to definitively explain these cases of CSCR presenting as IUES, but they could correspond to transitional forms between both diseases that could have in common a short axial length, a scleral thickening and choroidal venous congestion.
REFERENCES


sLEGENDS FOR FIGURES

Figure 1. Forty-five-year-old man with inferior retinal detachment and annular choroidal detachment in the right eye. Visual acuity: 20/32.

A: Ultra-wide-field color photo showing the inferior retinal detachment reaching the inferotemporal retinal vessels, associated with an annular choroidal detachment. Note the presence of an abnormal pigmentation in the posterior pole.

B: Mid-phase fluorescein angiography (FA) showing multiple subretinal hyperfluorescent leaking points (arrows), and a diffuse pigmentary disturbance.

C: Mid-phase Indocyanine Green Angiography (ICGA) showing hyperfluorescent choroidal plaques corresponding to the leaking points seen on FA, and a more intense leakage and staining just above the limit of the retinal detachment (arrow).

D: Ultrasonography B-scan showing the inferior retinal detachment, and the significant choroidal thickening.
E: Magnetic Resonance Imaging showing the retinal detachment and the ciliochoroidal detachment.

F: OCT B-scan passing through the inferior leaking zone (horizontal arrow on C) showing a choroidal thickening and bumping and a shallow retinal detachment. A focal posterior retinal edema (arrow) was noted at the ICG staining, the hyperreflectivity of which could be due to fibrin infiltration. Note also the presence of superficial retinal folds.

G–I: Left eye of the patient at the same time. Fundus autofluorescence shows several areas of hyperautofluorescence with a gravitational track (G). Fluorescein angiography (H) shows necklace retinal pigment epithelium lesions and leaking points (arrows). Mid-phase ICGA shows several hyperfluorescent choroidal plaques (I).

Figure 2. Same patient as in figure 1, 4 years after the onset.

A: Ultra-wide-field fundus autofluorescence showing hyper- and hypo-autofluorescent mottling retinal pigment epithelial (RPE) alterations in the posterior pole and the inferior periphery.

B: Early phase of indocyanine green angiography (ICGA) showing an abnormal dilated choroidal vein inferior to the macula where an intense ICG staining was located at the onset of the disease. The horizontal green line indicates the level of the B-scan shown in (C).

C. Horizontal B-scan: The choroid is still thick. A large choroidal vein is visible immediately beneath the RPE (large arrow) at the initial intense ICG staining. At this level, the retina remained focally thickened and raised by subretinal fibrosis.

Fig 3: Sixty-five-year-old man with a history of chronic central serous chorioretinopathy (CSCR) in both eyes, presenting with an inferior retinal detachment and an annular choroidal detachment in the left eye (LE).
A: Late phase of fluorescein angiography (FA) in the LE, one year before the occurrence of the exudative retinal detachment: note the presence of hyperfluorescent gravitational tracks nasal and inferior to the optic disc, a subretinal leaking point below the supero-temporal retinal vessels, and a small retinal pigment epithelial detachment (PED) inferior to the fovea, all these signs being suggestive of chronic CSCR.

B: Horizontal B-scan obtained at the same time, showing an alteration of the photoreceptor outer segments in the macula as well as a flat irregular PED.

C: Color photo one year later showing an inferior, mobile, folded retinal detachment (arrows) associated with an annular choroidal detachment (large arrow).

D: Late-stage FA performed the same day, with multiple subretinal leaking points, located mainly around the optic disc.

E: Vertical B-scan passing through the fovea, showing a massively thickened choroid with a bumpy surface. RPE granulations are present, and the retina is detached and presents superficial folds.

F: Magnetic resonance imaging showing the retinal and choroidal detachments.

G: Early-phase wide-field indocyanine green angiography (ICGA) showing extremely dilated choroidal veins around the optic disc.

H: During the late phase of ICGA, an intense peripapillary choroidal staining was seen.

Figure 4: Left eye of a 54-year-old man with inferior retinal detachment and annular choroidal detachment in both eyes 6 months apart (see supplemental figure for the right eye).

A and B: Left fundus upon presentation, 6 months before the onset of the retinal detachment. Fluorescein angiography (FA) (A) shows multiple alterations of the retinal pigment epithelium (RPE), including a subretinal leaking point (arrowhead) and a small pigment
epithelium detachment (arrow). Late-phase indocyanine green angiography (ICGA) showing several plaques of choroidal hyperfluorescence, including around the optic disc. Also note the circular arrangement of atrophic lesions of the retinal pigment epithelium above the optic disc.

C, D and E: Inferior retinal detachment and peripheral choroidal detachment in the same eye, 6 months later. Color photo montage (C) showing the inferior retinal detachment (small arrows) and the peripheral, annular choroidal detachment (large arrow); mid-phase FA shows the exacerbation of the subretinal leakage around the optic disc (D); late-phase FA montage shows the leopard-spot pigmentation and the choroidal detachment (arrow).

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