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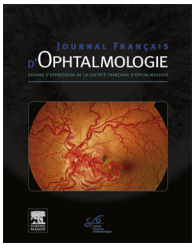

ORIGINAL ARTICLE

The retinal bacillary layer detachment: Clinical features and outcomes in posterior uveitis

Décollement bacillaire rétinien dans les uvéites postérieures : une étude descriptive

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KEYWORDS
Bacillary layer;
Choroiditis;
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Posterior uveitis;
Optical coherence tomography

Summary
Purpose. — To describe the clinical characteristics, presentation and response to treatment in posterior uveitis patients with bacillary layer detachment (BLD) seen on optical coherence tomography (OCT).
Materials and methods. — Retrospective review of patients with posterior uveitis and SD-OCT scans consistent with BLD. Data collected included demographics, uveitic etiology, treatment and duration of follow-up. Outcome measures included macular volume, central subfoveal thickness and visual acuity.

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Results. — Sixteen patients (20 eyes) were included. Twelve were female (75%). The mean age was 43.68 ± 14.7 years. The most frequent etiology of the uveitis was Vogt-Koyanagi-Harada (VKH) disease ($n = 10$), followed by sympathetic ophthalmia ($n = 2$). BLD was bilateral in four patients. Eight patients were treated with intravenous methylprednisolone boluses. Immunosuppressive therapies were required in 8 patients. The mean follow-up was 70 months (range: 2.0—216.0).
Conclusion. — BLD was observed in a series of posterior uveitis cases of various etiologies, showing functional and structural resolution with treatment in most cases.

MOTS CLÉS
Couche bacillaire ;
Choroïdite ;
Vogt-Koyanagi-Harada ;
Uvéite postérieure ;
Tomographie de cohérence optique

Résumé
Objectifs. — Décrire les caractéristiques cliniques, la présentation et la réponse au traitement des uvéites postérieures associés au décollement de la couche bacillaire (DCB) de la rétine observés par la tomographie de cohérence optique (OCT).
Méthodes. — Revue rétrospective de patients avec uvéites postérieures et leurs images compatibles avec un DBC sur OCT. Les données démographiques, l'étiologie des uvéites, le traitement et le temps de suivi ont été collectés. Le volume maculaire, l'épaisseur centrale subfovéale sur OCT et l'acuité visuelle, entre autres, ont été inclues dans les mesures à analyser.
Résultats. — Seize patients (20 yeux) ont été inclus. Douze étaient des femmes (75 %). La moyenne d'âge était de 43,68 ± 14,7 ans. L'étiologie la plus fréquente était la maladie de Vogt-Koyanagi-Harada ($n = 10$), suivie de l'ophtalmie sympathique ($n = 2$). Le DCB était bilatéral en quatre cas. Huit patients ont reçu des bolus intraveineux de méthylprednisolone. Le traitement immunodépresseur a été nécessaire pour 8 patients. La moyenne de suivi a été de 70 mois (rang : 2,0—216,0).
Conclusions. — Le DCB a été décrit dans une série d'uvéites postérieures de diverses étiologies avec une résolution fonctionnelle et structurelle dans la plupart des cas après le traitement.

Introduction

Posterior uveitis is an anatomic definition for immune-mediated diseases targeted towards the retina and/or choroid, according to the SUN (Standardization of Uveitis Nomenclature) classification [1]. Etiologically, posterior uveitis can be divided into infectious — such as toxoplasmic chorioretinitis - and non-infectious — such as Vogt-Koyanagi-Harada (VKH) disease.

Retinal bacillary layer has been described in 1941 by the neuroanatomist Polyak as the combination of both photoreceptor inner segments and outer segments [2]. The development of optical coherence tomography (OCT) has permitted a better understanding of the retinal layers by different reflectivity response patterns. In this regard, layers with high concentration of mitochondria are suggested to generate a hyperreflective signal in OCT scans [3]. With the development of enhanced definition OCT imaging, the inner segments of the photoreceptor layer have been divided in the myoid and ellipsoid zones [4].

Recently, bacillary layer detachment (BLD) has been defined as the disruption within inner segment (myoids and ellipsoids), after histologic findings of macular cone photoreceptor degeneration [5]. Scarce reports of BLD in toxoplasmic chorioretinitis [6,7], VKH [8,9], acute posterior multifocal placoid pigment epitheliopathy (APMPPE) [10], acute idiopathic maculopathy (AIM) [10], age macular degeneration (AMD) [5] and blunt eye trauma [11] have been described. Although the pathogenesis of BLD is still uncertain, an intense choroidal swelling is suggested to play a major role.

Herein, we describe clinical features and prognosis of patients with posterior uveitis of different etiologies presenting with BLD as detected by spectral domain optical coherence tomography (SD-OCT).

Materials and methods

Retrospective chart review of patients with BLD seen on SD-OCT, (HD Cirrus, Carl Zeiss, Germany) in the context of posterior uveitis at four Spanish tertiary referral centres from January 2015 to January 2021. Data collected included: demographics, age at diagnosis, sex, evolution time of the uveitis to BLD treatment and time of follow-up. Anterior chamber cell (ACC) count was assessed with slit lamp examination and graded according to Standardization of Uveitis Nomenclature (SUN) Working Group scale.

Table 1 Baseline characteristics.

Variable	*n* = 16
Age; mean (SD)	44,6 (15,01)
Sex; *n* (%)	
Male	4 (25%)
Female	12 (75%)
Uveitis etiology; *n* (%)	
Vogt-Koyanagi-Harada	10 (62%)
Sympathetic ophthalmia	2 (12.5%)
Multifocal choroiditis	1 (6%)
Relentless placoid maculopathy	1 (6%)
Serpiginous chorioretinitis	1 (6%)
Toxoplasmic retinitis	1 (6%)
Laterality	
Unilateral	12 (75%)
Bilateral	4(25%)
Systemic corticosteorids; *n* (%)	
Oral prednisone (PDN)	15 (93.7%)
Metilprednisolone megadose (MTP)	8 (50%)
BCVA, logMar; mean (SD)	
Baseline (*n* = 14)	0.87 (0.59)
1 year follow-up (*n* = 14)	0.09 (0.21)

Vitreous haze (VH) was evaluated using dilated indirect ophthalmoscopy and compared with the National Eye Institute (NEI) scale. Laterality of the uveitis, anatomic subtype, etiological diagnosis and phenotypic descriptors were collected. Main outcome measures on SD-OCT at presentation and follow-up included: presence of serous retinal detachment (SRD), main BLD measurement (μm), central subfield thickness (CST, μm) and macular volume (MV, mm^3). The BCVA was converted to log- MAR units for quantitative analysis.

The outer hyperreflective bands were identified according to the IN-OCT consensus guidelines [12]. The myoid zone was identified as the hyporreflective zone between the external limiting membrane and ellipsoid zone. The BLD was defined as a separation within the myoid zone, separating the ellipsoid zone from the inner cell bodies. BLD thickness was measured manually by two specialists (I.H, A.M) using the caliper tool available on OCT-SD as a vertical line from the edges of the intraretinal cyst: the hyperreflective line on the retinal pigment epithelium (RPE; below) and the outer border of the external limiting membrane (ELM; above).

Outcome measures (macular volume, main BLD and central subfoveal thickness by SD-OCT, and visual acuity) are described with the mean, standard deviation (SD) and range.

Wilcoxon test was used for statistic analysis. A *P*-value < 0.05 was considered significant.

Results

Sixteen patients (20 eyes) were included in the study, 12 females and 4 males. The mean age at the time of BLD diagnosis was 43.68 ± 14.72 years (range: 10 to 63).

The mean follow-up was 70 months (range: 2.0—216.0 years). The most frequent etiology of the uveitis was VKH in 14 eyes from 10 patients (62%), followed by sympathetic ophthalmia (SO) in 2 eyes from 2 patients (12,5%). There was one patient each diagnosed of multifocal choroiditis, persistent placoid maculopathy (PPM), toxoplasmic chorioretinitis and tuberculosis-associated serpiginous choroidopathy presenting with BLD in at least one eye. BDL was unilateral in 12 patients (75%), while 4 patients (25%) showed bilateral simultaneous BLD, all in the context of acute VKH.

Mean BCVA in logMar of the fourteen eyes with BLD and with one year of follow-up was: 0.87 ± 0.59 (range: 0.2—2.3) at presentation, 0.12 ± 0.19 (range: 0—0.7) at 1 month, 0.06 ± 0.21 (range: 0—0.8) at 6 months and 0.09 ± 0.21 (range: 0—0.8) at one year of follow-up. Baseline characteristics of BLD are shown in Table 1.

Location of BLD with respect of the foveal area was: subfoveal in 13 eyes (65%), temporal in 2 eyes (10%), nasal in 3 eyes (15%), superior in 2 eyes (10%), and inferior in one eye (5%). Of the 20 eyes with BLD, 7 eyes showed a single BLD (35%) and 13 eyes (65%) showed multiple BLD. Fifteen out of 20 eyes (75%) showed adjacent serous retinal detachment.

Mean measurement of the main BLD at presentation was 517.46 ± 321.99 μm (15 eyes).

Of the fourteen BLD affected eyes with one-year follow-up, mean CST at presentation was 541.5 ± 253.8 (range: 187—915), 225.2 ± 21.8 (range: 183—257) at 1 month, 262.78 ± 70.2 (range: 210—490) at 6 months, and 252.64 ± 27.85 (range: 215—306) at 1 year. Of the fourteen BLD affected eyes with one-year follow-up, mean MV at presentation was 14.4 ± 3.71 (range: 8.4—20,5), 9.7 ± 1.3 (range: 6.3—11.4) at 1 month, 9.4 ± 1.7 (range: 4.8—10.6) at 6 months, 9.4 ± 1.7 (range: 4.8—10.6) at 1 year.

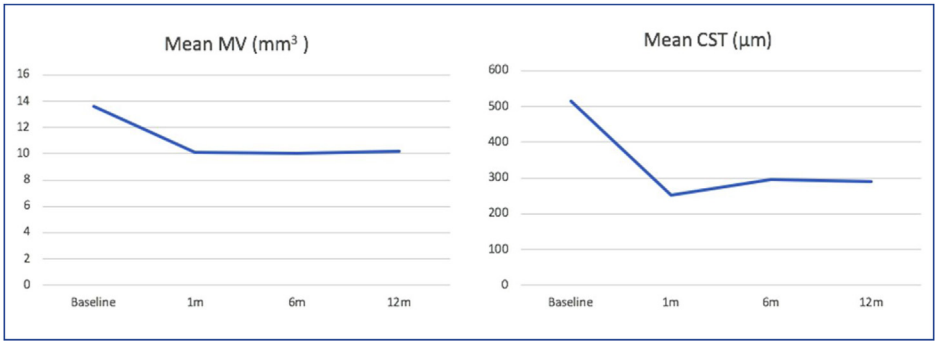

Figure 1. Mean MV and CST at baseline, 1 month, 6 months and one year of follow-up. Only the fourteen patients that achieved 1 year of follow-up were included.

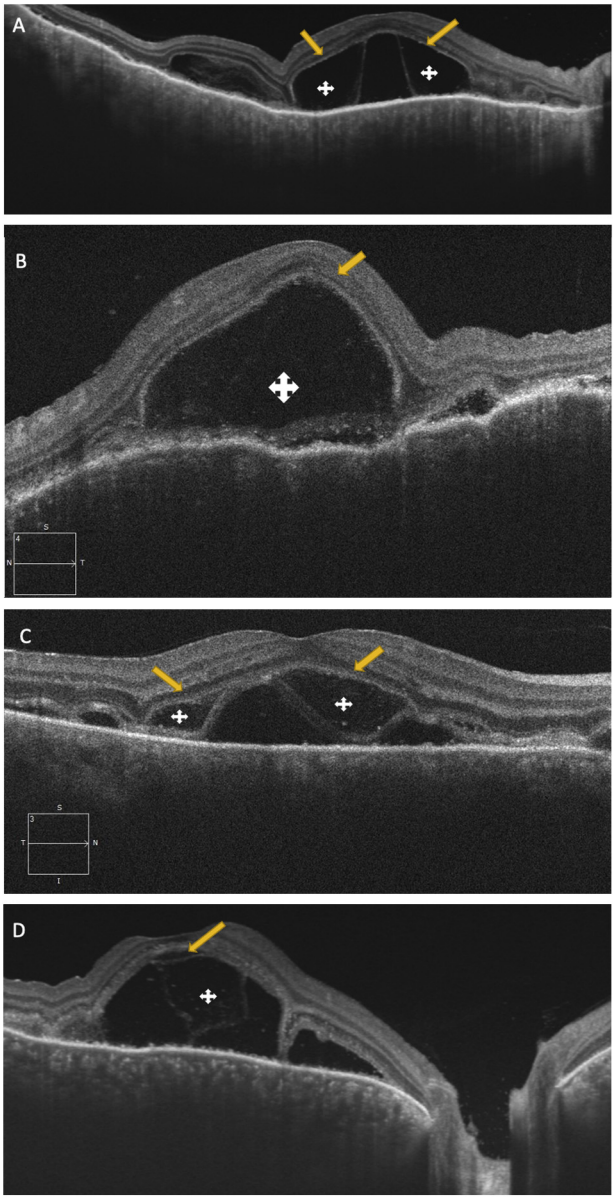

Figure 2. Bacillary layer detachment in four patients with acute Vogt-Koyanagi-Harada at baseline. OCT showed a cystic space (white cross) with hyperreflective line at the level of ellipsoid band due to the splitting of the photoreceptors (yellow arrow). Thickness of the choroid was also observed in all eyes.

At one-year follow-up ($n = 14$), BCVA improved significantly compared to baseline ($P = 0.001$) (Fig. 1), whereas a significant decrease in CST ($P = 0.009$) and MV ($P = 0.005$) were detected on SD-OCT (Figs. 2 and 3).

Fifteen patients (93.7%) were known to be treated with systemic corticosteroids, including intravenous methyl prednisolone megadoses (500 to 1000 mg/day) during three consecutive days in eight of them. Immunosuppressive therapy was required in 8 out of 14 patients (50%) during the first year of follow-up to control ocular inflammatory activity.

At one-year of follow-up, ten eyes (71%) showed a complete visual recovery (LogMar 0.00) while four eyes (28%) disclosed visual impairment in different degrees of severity (logMar range: 0.1—0.8) due to persistent outer retinal damage.

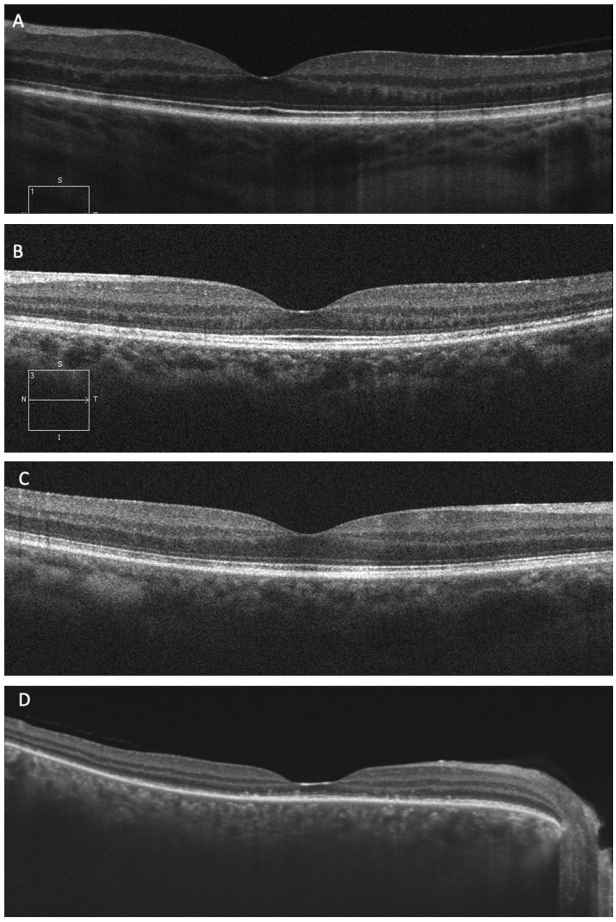

Figure 3. Resolution of bacillary layer detachment after systemic corticosteroid treatment was observed in OCT in the four cases of Vogt-Koyanagi-Harada shown in Fig. 2. Complete anatomic recovery of the retina was showed in all cases. Moreover, a decreased in the choroid thickness compared to baseline was observed in all cases.

Discussion

We describe the clinical features and outcomes of patients with posterior uveitis of different etiologies presenting with BLD as detected by spectral domain optical coherence tomography (SD-OCT). This new entity has been described in sparse reports and called by different names including ''atypical outer retinal fluid'' [13] or ''subretinal fluid with subretinal fibrin accumulation'' [14]. Recently, Mehta et al. described BLD as a new phenotype of macular thickening where fluid accumulates within a cystoid structure due to photoreceptors splitting, precisely at the level of the myoids. Our results support this hypothesis as showed by SD-OCT scans Fig. 1.

Myoids are considered to be a weaker structure in the photoreceptor layer, in contrast to the external limiting membrane (above) or the ellipsoid layer (below) [4]. In this regard, histology of macular cones in patients with age-related macular degeneration revealed their ability to loose inner segments under particular conditions [3].

The interpretation of the hyperreflective line on the floor of the cystoid space has been widely discussed. Traditionally, it was attributed to a proteinaceous material as a result of the RPE swelling [15]. However, Mehta et al. recently suggested this structure to be the deposition of mitochondria-rich ellipsoids that is presumed to be hyperreflective on OCT [7]. Besides, typical vertical columns within BLD walls have been hypothesised to be the degenerated Müller cells [16], instead of fibrinous deposits as thought in the past [15]. Correlation between retinal cell histology and structural image on SD-OCT has dramatically improved in the last years. However, for some authors, there is still a controversy in the third hyperreflective band [4]. Until the correlation of SD-OCT scans with retinal histology would not be cleared, the exact location of the retinal splitting in BLD according to OCT findings would not be confirmed.

The mechanism why fluid accumulates in this particular macular phenotype instead of in the shape of macular oedema or serous retinal detachment is still unknown. Cicinelli et al. hypothesize that a massive acute choroidal exudation — such as in VKH — would induce the fluid to penetrate into the neuroretina causing the photoreceptor splitting at the most fragile point: the myoid layer [16]. Herein, we report BLD in seven different ocular conditions that share the hallmark of hyperacute choroidal swelling at presentation, being VHK the most frequently described. In most of our cases, BLD was associated with subretinal fluid or serous retinal detachment, both typical signs of choroidal inflammation as suggested in the physiopathological mechanism of BLD.

Therefore, treatment with high dose systemic steroids is essential to halt chorioretinal swelling and to restore visual acuity. Treatment with systemic corticosteroids have shown a prompt resolution of BLD in most of our cases, with good visual prognosis at one year. However, two patients of different etiologies (PPM and SO) shown a shady visual prognosis at one year despite treatment due to reluctant macular oedema and possible subretinal fibrosis.

With the emergence of new OCT technologies, we support an increase in the prevalence of BLD. Nevertheless, this entity may still be underestimated due to the transient nature and rapid resolution after intensive corticosteroid treatment.

The present study was a heterogeneous case series and patients were analysed retrospectively. Further studies should be carried out to determine whether BLD could merge as a prognostic biomarker in posterior uveitis.

Conclusion

BLD has emerged as a new type of macular thickening of different etiologies in the context of posterior uveitis, which may help the specialist to better understand the underlined condition and to institute an appropriate and prompt treatment.

Disclosure of interest

The authors declare that they have no competing interest.

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