Choroidal effusion as a manifestation of central serous chorioretinopathy: A case report

Tuan Tran a, b, c, *, Mali Okada b, c, Jonathon Goh b, c, Thomas J. Gin b, c, C. Alex Harper b, c

a University of Sydney, Save Sight Institute, Sydney, NSW, Australia
b Royal Victorian Eye and Ear Hospital, East Melbourne, Victoria, Australia
c Centre for Eye Research Australia, East Melbourne, Victoria, Australia

ARTICLE INFO

Keywords:
Central serous chorioretinopathy
Choroidal effusion
Pachychoroid

ABSTRACT

Purpose: To report a case of bullous central serous chorioretinopathy presenting with large choroidal effusions. Observations: A patient presented with typical features of bullous central serous chorioretinopathy with large choroidal effusions. He had a previous history of bullous central serous chorioretinopathy in his other eye. The condition worsened after a short course of oral prednisolone, consistent with central serous chorioretinopathy. Surgical management with sclerectomies resulted in resolution of serous retinal detachment, choroidal effusions and subfoveal fluid.

Conclusions: We report choroidal effusions as a potential manifestation of central serous chorioretinopathy which may aid in our understanding in the pathogenic mechanisms of this condition. Furthermore, we demonstrate that surgical sclerectomies as a potential treatment option for serous retinal detachment and choroidal effusions in this condition.

A 64 year-old male patient of Salvadoran background presented with mildly reduced vision in his right eye three months after routine cataract surgery. His medical history consisted of well controlled insulin dependent diabetes mellitus, hypertension and hyperlipidaemia. His ophthalmic history was significant for left chronic bullous central serous chorioretinopathy (CSC) which started five years ago and eventually resulted in chronic poor vision. The previous episode of bullous CSC in his left eye was precipitated by the use of topical corticosteroid cream for dermatitis. After a year of observation for fluctuating disease, he underwent two sessions of focal thermal photocoagulation to the superotemporal choroidal polypoidal lesion and other extrafoveal areas of leak guided by fundus fluoresceine angiogram (FFA) (Fig. 1). Due to the ongoing persistence of bullous serous retinal detachment, chronic nature of disease and severity of vision loss, he was then commenced on oral eplerenone 50mg daily which was ceased after 12 months due to the lack of any clear clinical effect. Unfortunately, indocyanine angiography (ICGA) and photodynamic therapy (PDT) were unavailable at our institution. After two years of persistent disease the bullous serous retinal detachment and subfoveal fluid gradually resolved. As a result, he developed wide-spread RPE atrophy at the posterior pole and inferior retina along with diffuse outer retinal degenerative changes of the macula. The best corrected visual acuity (BCVA) in his left eye was 6/96 while his right eye remained unaffected at the time with BCVA of 6/5. He did not develop choroidal effusions at any time during the course of disease in his left eye. The condition remained stable and inactive for the following three years without further episodes of recurrence until his current presentation.

On his current presentation, he complained of mildly reduced vision in his right eye three months after routine cataract surgery. Post-operatively, he used topical prednisolone acetate 1%/phenylephrine hydrochloride 0.12% (Prednefrin Forte®) four times a day for four weeks. His one-month post-operative BCVA in the right eye was 6/9.

On examination, his BCVA of his right eye had reduced to 6/18 while his left remained at 6/96. Intraocular pressures were 14 mmHg in each eye. Preoperative axial length of the right eye was 22.51mm and left eye was 22.83mm (IOL Master 700, Carl Zeiss Meditec, Jena, Germany). Clinical examination of his left fundus remained unchanged. The right anterior chamber was deep with trace cells in both the anterior chamber and vitreous. The right posterior segment had large peripheral choroidal effusions with an inferior shifting serous retinal detachment without

Abbreviations: BCVA, Best corrected visual acuity; CSC, Central serous chorioretinopathy; FAF, Fundus Autofluorescence; FFA, Fundus fluoresceine angiogram; OCT, Optical coherence tomography; UES, Uveal effusion syndrome.

* Corresponding author. University of Sydney, Save Sight Institute, Sydney, NSW, Australia.
E-mail address: tuantran86@gmail.com (T. Tran).

https://doi.org/10.1016/j.jaocase.2022.101311
Received 22 October 2020; Received in revised form 28 June 2021; Accepted 18 January 2022
Available online 22 January 2022
2451-9936/© 2022 The Authors. Published by Elsevier Inc. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).
signs of chorioretinal inflammation (Fig. 2A). OCT demonstrated subfoveal fluid without an underlying pigment epithelial detachment (Fig. 2D). B-scan ultrasonography demonstrated hypoechoic choroidal detachments without abnormal scleral thickening, enhancing or masses.

FFA demonstrated typical features of CSC with multiple areas of ink-blot leakage without signs of an early diffuse leopard spot pattern or retinochoroidal folds typically seen in UES (Fig. 2C). The areas of leakage did not correspond with subretinal fluid and appeared small and topographically unrelated to the large inferior serous retinal detachment or peripheral choroidal effusions. As such, we felt that focal laser photocoagulation would not provide significant benefit, similar to previous unsuccessful attempts on the left eye, thus was not performed.

Blood tests including complete blood count, blood biochemistries, erythrocyte sedimentation rate, C-reactive protein, angiotensin converting enzyme and serological tests for syphilis and tuberculosis were all within normal limits. MRI brain and orbits with contrast was unremarkable with no signs of orbital vascular congestion or posterior scleritis. The scleral thickness at the posterior pole measured by MRI was normal with both measuring 0.7mm, correlating with similar measurements on B-scan ultrasonography. After excluding secondary causes of uveal effusions and without similarly described presentations of choroidal effusions in pachychoroid disease in the literature, he was initially thought to have idiopathic uveal effusion syndrome (UES) precipitated after routine cataract surgery. The patient was commenced on a short trial of oral prednisolone 75mg/day for seven days, which was promptly tapered due to the lack of clinical effect.

After two months, the large peripheral effusions persisted and there was gradual worsening of both the inferior shifting serous retinal detachment and sub-foveal fluid (Fig. 3A). BCVA of his right eye had significantly reduced to 6/60 while the left remained at 6/96. Due to the progression of disease and severity of vision loss, he underwent surgical drainage of his choroidal effusions via three full-thickness deep sclerrotomies under partial thickness scleral flaps (4mm × 5mm) at the inferonasal, inferotemporal and superotemporal quadrants. It was thought that treating three quadrants would be enough to treat the effusions and the specific quadrants were deemed to best protect the

![Fig. 1. (A) Fundus Colour photos from his history of CSC five years ago with a left inferior bullous serous retinal detachment (arrow), elevated choroidal polypoidal lesion and the absence of choroidal effusions. The right macular has irregular pigmented changes in the macular. (B) FFA demonstrating multiple areas of late ink-blot leakage and inferior serous retinal detachment (arrow). (C) OCT enhanced depth imaging of his left eye showing sub-foveal choroidal thickness of 449μm and pachyvessels (arrowheads). The right eye with outer retinal changes at the macular consistent with CSC. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)](image-url)
macula in this case. The consistency of the sclera was noted to be normal intraoperatively. Histopathology of biopsied sclera did not show any abnormal extracellular mucin deposition or excess glycoaminoglycans (GAGs) typically found in UES (Fig. 4). Three months post-operatively the choroidal effusions, shifting serous retinal detachment and subfoveal fluid had resolved (Fig. 3C), and his BCVA had improved to 6/36. The superonasal choroidal effusion required more time to resolve as a superonasal sclerotomy was not performed (Fig. 3B).

This case demonstrates an atypical presentation of bullous CSC with large peripheral choroidal effusions after routine cataract surgery. At the time of submitting this manuscript, this was the first case to our knowledge of bullous CSC manifesting with peripheral choroidal effusions. Upon revising the manuscript, Boulanger et al.1 has recently published further cases that similarly worsened after receiving oral corticosteroids and were successfully treated with sclerectomies.

We believe the presentation of choroidal effusions were related to his underlying condition of bullous CSC for a number of reasons. Firstly, the left eye which had not had any history of choroidal effusions, was managed as chronic bullous CSC for several years. This was demonstrated on OCT by sub-foveal serous detachments, increased sub-foveal choroidal thickness and typical multifocal ink-blot leaks on FFA. Furthermore, the left eye had several other OCT features often seen in bullous CSC including; large peaked PED of uniform hyperreflectivity, hyperreflective dots within the choriocapillaris and hyperreflective choroidal vascular walls (Fig. 2B).2

The patient was initially thought to have UES in the right eye as cataract surgery is a common precipitating factor of UES.3 However, several features argue against this. The OCT of the right eye demonstrated intraretinal hyper-reflective dots, subretinal fibrin deposits and hyper-reflective dots at the level of choriocapillaris. Outer-retinal degenerative changes from long standing CSC can be seen in the left eye along with features of bullous CSC including hyper-reflective choroidal vascular walls and hyper-reflective dots within the choriocapillaris.
the choroid include: vortex vein outflow, transscleral macromolecular diffusion, transscleral hydrostatic water movement, and bulk flow around vessels and nerves of scleral emissaries. CSC is thought to occur from increased fluid leakage from presumed hyperpermeability at the level of the choriocapillaris. If this leakage is redirected posteriorly rather than anteriorly towards the sub-RPE or subretinal space and if leakage occurs to a degree that overwhelms these mechanisms of suprachoroidal fluid drainage, then theoretically choroidal effusions may occur.

In the case series of choroidal effusions in patients with bullous CSC by Boulanger et al., there are common features with our case which include; the presence of multi-focal leaks on FFA, increased choroidal thickness, chronic changes of CSC and a previous history of CSC. In attributing potential mechanisms of choroidal effusions in CSC, increased choroidal permeability by ICGA has been demonstrated in idiopathic UES. Furthermore, cases of UES with pachychoroid features have recently been described. Perhaps these cases represented atypical manifestations of bullous CSC similar to our case or whether choroidal hyperpermeability is a possible feature of disease overlap.

Spaide and Ryan postulated on the possibility of CSC causing choroidal effusions when examining a large series of patients with CSC. By using optimal binning of their data they demonstrated that as subfoveal choroidal thickness exceeds approximately 400 μm, fluid may saturate the choroidal stroma and accumulate in the posterior choroid or suprachoroidal space. They proposed that as the choroidal stroma expands to a certain limit, any incremental fluid accumulation would accumulate elsewhere, whether it be anteriorly towards the subretinal space or posteriorly within the suprachoroidal space. Anatomic differences of the choroidoscleral junction between the posterior pole and the periphery may explain the usual absence of choroidal effusions in CSC, as choriocapillaris leakage generally occurs in discrete areas at the posterior pole. Peripheral choroidoscleral attachments are weak and bounded by vortex veins allowing for its lobular appearance. In contrast, they are firmly connected anchored at the posterior pole. Therefore, it seems plausible that in extreme cases of bullous CSC, fluid

Fig. 3. Each demonstrating wide-field pseudo-colour image (top), autofluorescence (middle) and OCT (bottom) (A) pre-operatively, (B) one month post-operatively and (C) three months post-operatively. 3A. Subretinal fibrin can be seen inferonasally within bullous retinal detachment which is often found in chronic bullous CSC. 3B. Significant improvement of choroidal effusion at locations of sclerectomies with the superonasal effusion remaining at one month post-operatively. 3C. Resolution of choroidal effusions and subfoveal fluid with outer retinal changes on OCT three months post-operatively. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

Fig. 4. Histologic examination of the patient’s sclera under light microscopy with Alcian blue staining (original magnification x40) demonstrating normal scleral tissue architecture. There is minimal alcianophilic staining to suggest abnormal excess accumulation of extracellular mucin or glycoaminoglycans. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)
leakage from the choriocapillaris may accumulate in the posterior choroid at both posterior and peripheral locations. This may overwhelm the normal physiological mechanisms of suprachoroidal fluid drainage and result in choroidal effusions as we believe to have occurred in our patient.

Our case describes large choroidal effusions in the presence of active bulbar CSC which improved after surgical drainage. We caution the use of oral corticosteroids without a clear inflammatory cause of uveal effusions and with a previous history or features of chronic CSC. This may worsen an underlying atypical bulbar CSC variant such as in our case. Multi-focal leaks on FFA may be a distinguishing feature of CSC from other causes. Surgical sclerotomies may effectively treat the choroidal effusions and serous retinal detachments in bulbar CSC. Further studies are required to understand the mechanisms of fluid leakage and drainage within the choroid under physiological conditions and in CSC to explain this atypical manifestation.

Patient consent
Written consent to publish case details was obtained by the patient.

Disclosures
The authors declare no financial support was provided to conduct this research.

Funding
No funding or grant support

Authorship
All authors attest that they meet the current ICMJE criteria for Authorship.

Ethics
Written consent was obtained in writing from the patient. Formal institutional ethics approval was not required.

Declaration of competing interest
None.

Acknowledgements
The authors thank A/Prof Penny McKelvie from the Department of Pathology, St Vincent’s Hospital, Melbourne Australia, for reporting the histopathological findings and providing these images.

References
1. Boulanger E, Bonnin S, Delahaye-Mazza C, et al. Central serous chorioretinopathy mimicking idiopathic uveal effusion syndrome. Retin Cases Brief Rep. 2021. https://doi.org/10.1097/ICB.0000000000001170, 2021/06/04.
2. Balaratnasingam C, Freund KB, Tan AM, et al. Bulbous variant of central serous chorioretinopathy: expansion of phenotypic features using multimethod imaging. Ophthalmology. 2016;123:1541–1552. https://doi.org/10.1016/j.ophtha.2016.03.017, 2016/04/17.
3. Elagouz M, Stanescu-Segall D, Jackson TL. Uveal effusion syndrome. Surv Ophthalmol. 2010;55:134–145. https://doi.org/10.1016/j.survophthal.2009.05.003, 2010/02/18.
4. Gass JD. Bulbous retinal detachment. An unusual manifestation of idiopathic central serous choriodopathy. Am J Ophthalmol. 1973;75:810–821. https://doi.org/10.1016/0002-9394(73)90887-8, 1973/05/01.
5. Gass JD, Jallow S. Idiopathic serous detachment of the choroid, ciliary body, and retina (uveal effusion syndrome). Ophthalmology. 1982;89:1018–1032. https://doi.org/10.1016/s0161-6420(82)34685-6, 1982/09/01.
6. Uyama M, Takahashi K, Koizumi Y, et al. Uveal effusion syndrome: clinical features, surgical treatment, histologic examination of the sclera, and pathophysiology. Ophthalmology. 2000;107:441–449. https://doi.org/10.1016/s0161-6420(99) 00414-4, 2000/03/11.
7. Shields CL, Roelofs D, Di Nicola M, et al. Uveal effusion syndrome in 104 eyes: response to corticosteroids - the 2017 Axel C. Hansen lecture. Indian J Ophthalmol. 2017;65:1093–1104. https://doi.org/10.4103/ij.o.752.17, 2017/11/15.
8. Lam A, Sambursky RP, Maguire M. Measurement of scleral thickness in uveal effusion syndrome. Am J Ophthalmol. 2005;140:329–331. https://doi.org/10.1016/j.jao.2005.02.014, 2005/08/10.
9. Gass JD. Pathogenesis of disciform detachment of the neuroepithelium. Am J Ophthalmol. 1967;63(suppl 1-139), 1967/03/01.
10. Scheider A, Nasemann JE, Lund OE. Fluorescein and indocyanine green angiography of central serous chorioretinopathy. Arch Ophthalmol. 1994;112:1057–1062. https://doi.org/10.1001/archoph.1994.01090200063023, 1994/08/01.
11. Scheider A, Nasemann JE, Lund OE. Fluorescein and indocyanine green angiographies of central serous choroidopathy by scanning laser ophthalmoscopy. Am J Ophthalmol. 1993;115:50–56. https://doi.org/10.1016/0002-9394(93)73524-8, 1993/01/01.
12. Kumar A, Kesler S, Singh RP. The indocyanine green findings in idiopathic uveal effusion syndrome. Indian J Ophthalmol. 2002;50:217–219, 2002/10/03.
13. Jin S, Francisconi CLM, Wong DT. Pachychoroid findings in a case of uveal effusion syndrome. Can J Ophthalmol. 2020;55:e74–e76. https://doi.org/10.1016/j. cjo.2019.08.002, 2019/11/13.
14. Tumurbayashi Y, Morishita S, Kohimoto R, et al. Type III uveal effusion syndrome suspected to be related to pachychoroid spectrum disease: a case report. Medicine (Baltimore). 2020;99.e21441. https://doi.org/10.1097/MD.0000000000012441, 2020/08/07.
15. Spaide RF, Ryan Jr EH. Loculation of fluid in the posterior choroid in eyes with central serous chorioretinopathy. Am J Ophthalmol. 2015;160:1211–1216. https://doi.org/10.1016/j.ajo.2015.08.018, 2015/08/25.
16. Spaide RF, Orlock DA, Herrmann-Delemazure B, et al. Wide-angle indocyanine green angiography. Retina. 1998;18:44–49. https://doi.org/10.1097/00006982-199801000-00008, 1998/03/21.
17. Pang CE, Shah VP, Sarraf D, et al. Ultra-widefield imaging with autofluorescence and indocyanine green angiography in central serous chorioretinopathy. e362 Am J Ophthalmol. 2014;158:362–371. https://doi.org/10.1016/j.ajo.2014.04.021, 2014/ 05/06.
18. Mesic RA. Detachment of ciliary body-anatomical and physical considerations. Invest Ophthalmol. 1965;4:935–941, 1965/10/01.