Case Report

Two Patients with Atypical Choroidal Detachment

Joseph W. Fong Heather V. Broyles Nour Y. Atassi Ahmed B. Sallam
Sami H. Uwaydat

Department of Ophthalmology, Jones Eye Institute, University of Arkansas for Medical Sciences (UAMS), Little Rock, AR, USA

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Abstract

Serous choroidal detachment that is caused by rhegmatogenous retinal detachment (RRD) may present a significant diagnostic challenge as delayed recognition and repair of the underlying RRD can severely impact the final anatomical and visual outcome. We report 2 consecutive patients with atypical choroidal detachments who were later found to have underlying RRDs. A 71-year-old female presented with a 1-week history of painful vision loss and floaters in the left eye. Examination revealed choroidal detachments in the nasal and temporal periphery and an overlying retinal detachment with shifting subretinal fluid. However, no retinal breaks were identified. An extensive laboratory workup and imaging of the orbits were unrevealing. She was treated with 80 mg oral prednisone daily for 2 weeks with subsequent resolution of the choroidals but persistence of the retinal detachment. Similarly, a 52-year-old male presented with a 3-week history of flashes and floaters followed by painful vision loss in the left eye 1 day prior to presentation. He had hand motion vision OS and the intraocular pressure was undetectable by hand-held tonometry OS. Dense brunescant cataract prevented adequate viewing of the posterior pole. B-scan ultrasonography revealed a funnel retinal detachment, with homogenous choroidal echogenicities suggestive of hemorrhagic choroidal detachment. Extensive laboratory workup was unrevealing. The patient was started on 60 mg oral prednisone and re-evaluated every 2 days, but ultrasonography revealed persistence of the choroidal detachment after 1 week. The diagnosis of RRD with an associated choroidal detachment should be considered, even in the absence of an identifiable causative retinal break.

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Correspondence to:
Sami H. Uwaydat, shuwaydat@uams.edu

Introduction

An unexplained serous choroidal detachment may present a significant diagnostic challenge. The differential diagnosis of choroidal detachment with overlying retinal detachment includes inflammatory conditions (posterior scleritis), neoplastic etiologies (choroidal melanoma and choroidal metastatic lesions), and idiopathic etiologies such as uveal effusion syndrome [1–3]. However, when resolution of the choroidal detachment occurs with persistence of the retinal detachment following steroid therapy, the diagnosis of rhegmatogenous retinal detachment (RRD) with an associated choroidal detachment (RRDCD) should be considered, even in the absence of an identifiable causative retinal break. We present 2 consecutive patients with RRDCD and their outcomes.

Case Presentation 1

A 71-year-old female presented with a 1-week history of painful vision loss and floaters in the left eye. She had no history of myopia, previous ocular trauma, or surgery. On initial examination, her visual acuity was 20/30-2 OD and 20/80 OS with intraocular pressures of 21 mm Hg OD and 17 mm Hg OS. Examination of the unaffected right eye was within normal limits, with a mild nuclear sclerotic cataract, and no evidence of intraocular inflammation. Examination of the left eye revealed rare anterior chamber cells, 1+ vitreous cells, choroidal detachments in the nasal and temporal periphery, and an overlying retinal detachment with shifting subretinal fluid as shown in Figure 1a. No retinal breaks were identified. Her medical history was nonrevealing for underlying medical conditions, specifically hypertension, autoimmune diseases, and systemic malignancy. Complete blood count, basic metabolic panel, HIV, T-spot, syphilis screen, and antinuclear cytoplasmic antibody panel were all normal. MRI of the brain and orbits revealed mild enhancement of the posterior sclera but no intraocular mass. Fluorescein angiography showed late leakage in the temporal periphery, fundus autofluorescence image showed the normal autofluorescent pattern of retinal pigment epithelium (Fig. 1b), and optical coherence tomography of the macula showed shallow subretinal fluid and a posterior vitreous detachment (Fig. 1c). The patient was started on 80 mg oral prednisone for a presumed diagnosis of serous retinal detachment secondary to posterior scleritis. Two weeks later, the choroidal detachments resolved but the retinal detachment persisted, exhibiting a more corrugated appearance as shown in Figure 1d, yet no retinal breaks were identified. Intraocular pressures remained normal.

After 4 weeks of observation and treatment with oral steroids, the retinal detachment extended to involve the superior quadrants with development of fixed retinal folds in the inferior quadrants. No retinal breaks could be identified on serial fundus examinations. At this point, the patient underwent pars plana vitrectomy combined with phacoemulsification with intraocular lens implantation and placement of an encircling band. Intraoperatively, a small peripheral break was identified at the 11 o'clock meridian. An inferior retinectomy was performed to mobilize and flatten the retina, and the vitreous cavity was filled with silicone oil. At the 6-week visit, the retina remained reattached and the patient's vision had improved to 20/400.

Case Presentation 2

A 52-year-old male presented with a 3-week history of flashes and floaters followed by painful vision loss in the left eye 1 day prior to presentation. He had no history of myopia, previous ocular trauma, or surgery but reported a family history of retinal detachment in 2

Fig. 1. **a** Optos color fundus photography showing choroidal detachments in the nasal and temporal periphery with an overlying retinal detachment. **b** Fundus autofluorescence image showing the normal autofluorescent pattern of retinal pigment epithelium. **c** OCT of the macula showed shallow sub-retinal fluid. **d** Optos color fundus photography showing resolution of choroidal detachments with persistent retinal detachment 2 weeks after starting oral steroids. OCT, optical coherence tomography.

immediate family members. On initial examination, his visual acuity was 20/200 OD and hand motion OS with intraocular pressures of 24 mm Hg OD and undetectable by hand-held tonometry OS. Examination of the unaffected right eye was within normal limits, with a 3+ brunescent nuclear sclerotic cataract, no evidence of intraocular inflammation, and an attached retina. Examination of the left eye revealed rare anterior chamber cells and 4+ brunescent nuclear sclerotic cataract which prevented adequate viewing of the posterior pole. B-scan ultrasonography revealed a funnel retinal detachment, with homogenous choroidal echogenicities suggestive of hemorrhagic choroidal detachment as shown in Figure 2a. His medical history was significant for type 2 diabetes and hypertension but negative for autoimmune diseases and systemic malignancy. Complete blood count, basic metabolic panel, HIV, T-spot, syphilis screen, and antinuclear cytoplasmic antibody panel were all normal. Computed tomography of the brain and maxillofacial structures did not reveal any intraocular mass but demonstrated prominent choroidal detachment as shown in Figure 2b. Chest X-ray was within normal limits. The patient was started on 60 mg oral prednisone and re-evaluated every 2 days. Serial ultrasonography revealed persistence of the choroidal detachment. One week after initial presentation, the patient underwent pars plana vitrectomy combined with phacoemulsification, drainage of serous and hemorrhagic choroidals, placement of an encircling band, and injection of a C3F8 gas bubble. Intraoperatively, inspection of the retina revealed a total retinal detachment with retinal breaks in the inferior and superior periphery. At the 1-month visit, the retina remained attached (Fig. 2c) and the patient's vision had improved to counting fingers near face.

Fig. 2. **a** B-scan ultrasonography revealed a funnel retinal detachment, with homogenous choroidal echogenicities suggestive of hemorrhagic choroidal detachment. **b** Computed tomography of the brain and maxillofacial structures did not reveal any intraocular mass but demonstrated prominent choroidals. **c** Optos color fundus photography at the 1-month visit showing the retina remained attached.

Discussion and Conclusions

The reported rate of RRDCD is 2.0–4.5% of all primary RRDs in Western countries, and it is reported as high as 18.1% of primary RRDs in China [4–6]. Risk factors for RRDCD include high myopia, history of ocular trauma, older age, and pseudophakia or aphakia [5]. RRDCD has a poor prognosis, primarily due to poor visualization and difficult identification of causative breaks that may cause delay in treatment, difficult application of retinopexy, and rates of proliferative vitreoretinopathy reported to be as high as 35.4–52.4% after initial surgery [7, 8].

The etiology of RRDCD is unknown. It is believed that in a RRD, liquefied synergetic vitreous containing inflammatory cytokines accesses the subretinal space, inducing choroidal vessel hyperpermeability. This hyperpermeability leads to exudation of fluid into the supraciliary and suprachoroidal spaces and subsequently results in ciliary body edema. Ciliary body edema reduces aqueous production which results in hypotony and the development of a choroidal detachment [8]. Our first patient did not have hypotony in the affected eye, but her IOP was 3–9 mm Hg lower in the affected eye than in the unaffected eye on serial exams. This is consistent with Seelenfreund's report that in 48 RRDCD patients who had IOP recording, all but 1 had a lower IOP in the affected eye compared to the fellow eye [5]. Our second patient, on the other hand, demonstrated frank hypotony.

Early diagnosis of RRDCD is crucial as delay in surgical intervention to repair the underlying RRD can negatively impact the final anatomical and visual outcome. RRDCD should be suspected when resolution of the choroidal detachment is noted with persistence of the retinal detachment. When this is observed, the diagnosis of RRDCD should be entertained despite the absence of an identifiable causative retinal break in the clinic setting, and prompt surgical intervention should be strongly considered to locate an occult retinal break. Despite clear visualization of the fundus, definite retinal breaks may not be found in 2.2–4% of phakic retinal detachments [1–3]. Other differential diagnoses of choroidal detachment with overlying retinal detachment include inflammatory conditions (posterior scleritis), neoplastic etiologies (choroidal melanoma and choroidal metastatic lesions), and idiopathic etiologies such as uveal effusion syndrome. The complaint of pain associated with vision loss, especially in case 1, initially led us to strongly suspect an inflammatory etiology, but in retrospect, the pain may be due to the presence of blood in the suprachoroidal space.

In case 1, although treatment with oral steroids or observation would have been appropriate for the management of a serous choroidal detachment alone, the progression of the patient's retinal detachment over the course of 1 month, despite the resolution of the choroidal detachment, warranted surgical intervention to identify and manage the causative retinal break. The retinal break that was identified intraoperatively explains the inferior retinal detachment seen on presentation, based on Lincoff's rules. We had an increased index of suspicion for the possibility of RRDCD with our second patient and elected to pursue aggressive surgical management much earlier in his clinical course especially considering the inability to identify a causative retinal break due to the dense cataract. Early recognition and surgical management of RRDCD decreases the likelihood of vision-threatening sequelae such as proliferative vitreoretinopathy and re-detachment. In summary, in patients who present with choroidal detachment, the possibility of a RRDCD should always be entertained, even when no retinal breaks can be identified or media opacities preclude adequate visualization of the peripheral retina.

Statement of Ethics

Written informed consent for this case report was obtained from the patient.

Conflict of Interest Statement

The authors declare that there are no conflicts of interest to disclose.

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Author Contributions

J.W.F.: writing and reviewing. H.V.B.: writing and reviewing. N.Y.A.: writing and reviewing. A.A.S.: reviewing. S.H.U.: reviewing.

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