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Profound improvement in vision and electroretinogram after intensive steroid treatment in unexplained visual loss after silicone oil removal

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ABSTRACT

Purpose: Unexplained vision loss after silicone oil removal is a well-documented but incompletely understood entity for which there is no effective treatment described in the existing literature. We present a case where intensive oral and periocular steroid treatment resulted in significant subjective and objective clinical improvement.

Observations: After successful pars plana vitrectomy with silicone oil endotamponade to repair a macula sparing retinal detachment, the patient's best corrected visual acuity was 20/20 with silicone oil in the operative eye. However, seven weeks after uncomplicated combined silicone oil removal and cataract extraction with intraocular lens insertion, best corrected visual acuity was 20/250 with no new ophthalmic pathology to explain the vision loss. After a four week course of oral prednisone and three periocular triamcinolone injections over a period of nine weeks, visual acuity improved to 20/25 -2 in the operative eye. Serial multifocal electroretinography initially showed severely diminished amplitudes but improved markedly over the course of steroid treatment.

Conclusions and Importance: Although no effective treatments are described in the existing literature, improvement in visual acuity, visual field, and electroretinogram in this case suggests that intensive steroid treatment (periocular and systemic) may be efficacious in treating unexplained vision loss after silicone oil removal.

1. Introduction

Unexplained vision loss after silicone oil removal (UVLSOR) is characterized by a drop in visual acuity following uncomplicated silicone oil removal as compared to when silicone oil was in the eye, excluding cases in which other ophthalmic pathology develops to explain the decrease. The entity was first described by Newsom et al., in 2004 and then by Cazabon and many others in the years following.[1–11] (see Fig. 1–5)

In the first of the two largest observational studies to date, Moya et al. reported that 3.3% of patients were thought to have unexplained vision loss after silicone oil removal, with a mean loss of vision of 3.7 Snellen lines in this subset.[3] Five of the fourteen patients with UVLSOR in this study exhibited some degree of visual recovery, only two of which recovered visual acuity to what it had been with oil in situ. Separately, Roca et al. reported that 5.9% of patients developed UVLSOR in their study, with most patients losing at least two lines of best corrected visual acuity (BCVA).[4] In a smaller study conducted by Oliveira-Ferreira et al., including 46 patients as opposed to 324 in that of Roca and 421 in that of Moya, 10.9% of patients developed UVLSOR, with each patient losing at least two Snellen lines.[3–5] In a recent multicenter case series examining 11 patients thought to have UVLSOR, the mean BCVA in the operative eye decreased from 20/72 (0.6 ± 0.2 logMAR) with oil in the eye to 20/458 (1.4 ± 0.4 logMAR) 1 month after SO removal.[6]

Patients with UVLSOR generally show diminished macular photoreceptor amplitudes on electroretinography.[1–3,7] Despite this observation, the pathophysiology of UVLSOR is still not definitively understood. Newsom et al. first theorized that tissue infiltration by silicone oil may cause damage to the optic nerve.[1] Cazabon et al. speculated that at the time of SOR, a sudden disruption of potassium buffering at the interface of the retina and vitreous may cause an accumulation of potassium ions in the retina and resulting excitotoxicity.[1] Dogramaci et al. later said that increased foveal light exposure at the time of SOR may point to phototoxicity as a cause for vision loss in these cases.[8]

While UVLSOR has now been widely acknowledged to exist, the literature on the topic has been solely observational. To our knowledge, no published reports exist on treatment of UVLSOR. The lack of concrete understanding of its pathophysiology is a major obstacle to formulating

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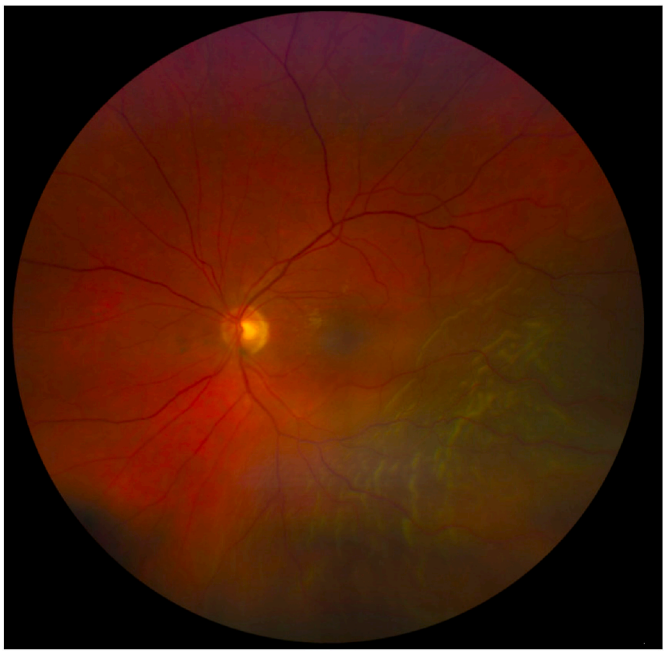

Fig. 1. Color fundus photos showing inferotemporal macula-sparing retinal detachment at initial presentation.

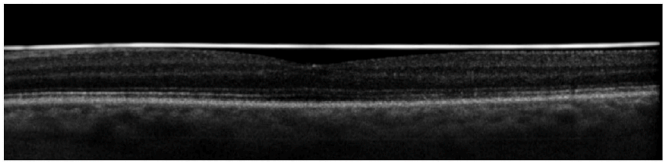

Fig. 2. Postoperative macular OCT showing normal retinal anatomy with silicone oil meniscus.

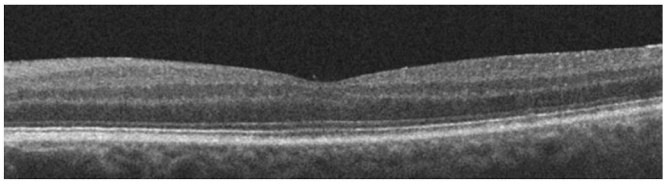

Fig. 3. Postoperative macular OCT after silicone oil removal showing normal retinal structures.

an effective treatment proposal. Here, we report a case of UVLSOR that showed marked improvement in visual acuity, visual field testing, and electrophysiologic testing over a course of intense steroid treatment.

2. Case report

The patient, a 63 year old male, initially presented with five days of a dark shadow in the peripheral vision of his left eye. Indirect ophthalmoscopy with scleral indentation revealed an inferotemporal macula-sparing rhegmatogenous retinal detachment associated with a horseshoe retinal tear at 4:30. The patient's pinhole visual acuity (VA) was 20/50 in the left eye and 20/20 in the right eye.

He was taken the same day for retinal detachment repair with combined pars plana vitrectomy (PPV) and scleral buckling. In preoperative discussion, the patient opted for silicone oil as opposed to gas endotamponade, as he had air travel that he could not cancel in the immediate future. The surgery proceeded without complication, and at

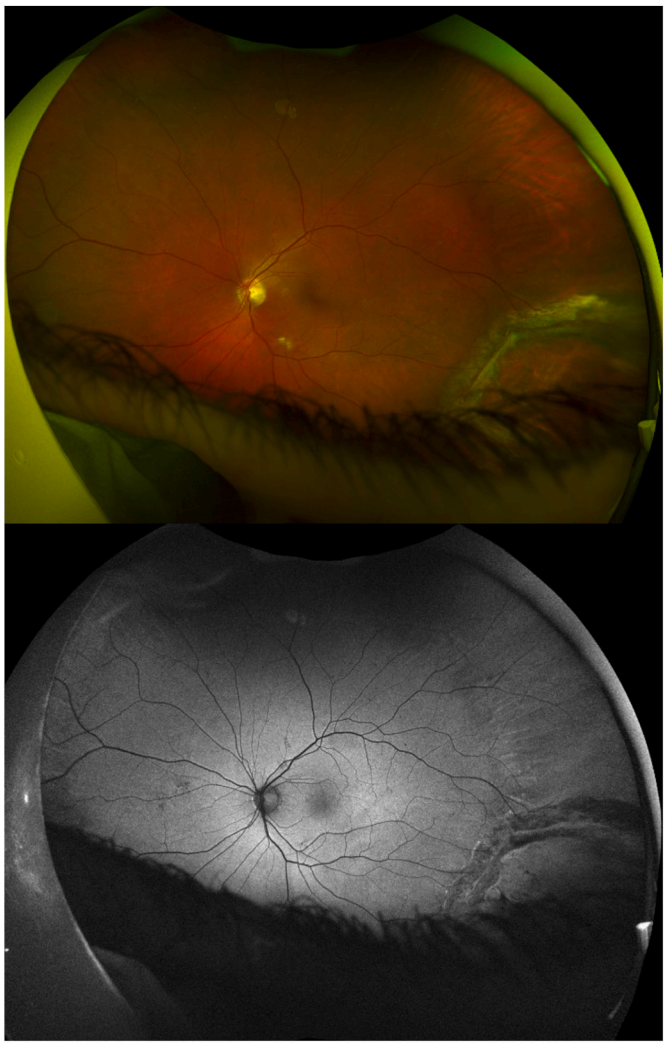

Fig. 4. Standard and autofluorescence fundus photos showing reattached retina and no new visible pathology six weeks after silicone oil removal.

his postoperative visits on day one and week one, the retina remained attached under silicone oil with normal intraocular pressure.

Twenty days after surgery, BCVA in the operative eye was 20/20 with a temporary refraction.

The remainder of the postoperative period proceeded uneventfully, other than the expected development of progressive nuclear sclerosis in the operative eye. Nearly five months after the initial retinal detachment repair, a combined vitrectomy, silicone oil removal and cataract extraction with intraocular lens placement was carried out without complication.

Postoperative day and week one visits were unremarkable, with normal intraocular pressure readings and expected postoperative appearance. The patient was able to count fingers postoperative day one, although VA testing was likely limited by partial air fill. His acuity improved to 20/300 without improvement by pinhole at week one (air resolved). The retina remained attached, and the intraocular lens was well-centered.

Two weeks after surgery, the patient's VA was 20/300 without improvement by pinhole. Examination revealed no inflammatory cells, posterior capsule opacification (PCO), or other new ophthalmic pathology. Three weeks later, pinhole VA remained 20/300 (without improvement by pinhole), significantly worse than anticipated given the operative eye refracted to 20/20 before the surgery. The patient also reported a central scotoma in the operative eye that appeared after

silicone oil removal. Optical coherence tomography (OCT) of the macula revealed normal retinal structures, and OCT of the retinal nerve fiber layer (RNFL) showed neither edema nor atrophy. Refraction the following day did not improve acuity.

Six weeks after oil removal, standard and autofluorescence fundus photos were collected, displaying a successfully reattached retina and providing no explanations for the lower than anticipated acuity.

Seven weeks after surgery, pinhole VA in the operative eye was 20/250, and visual field testing demonstrated a central scotoma.

At this time, exam of the operative eye revealed no inflammatory cells, PCO, or other new ophthalmic pathology. The retina remained attached and flat on scleral buckle. From the time of initial presentation, the patient's intraocular pressure (IOP) remained within the normal limits, and the cup to disc ratio remained stable at 0.2. A check by physician revealed no afferent pupillary defect.

A multifocal electroretinogram (mfERG) was done at this time, revealing markedly diminished responses compared to the fellow eye.

Given the significantly decreased visual acuity in the operative eye after SOR with no clear cause, the diagnosis of unexplained vision loss after silicone oil removal was considered most likely. Without treatment guidance from existing literature, a trial of steroid - both periocular and oral - was proposed to the patient. The rationale for treatment was that even though the putative initial injury causing vision loss could not be reversed, limiting any subsequent inflammation could only be of benefit.

The patient was amenable to proceed, and seven weeks after surgery, a periocular triamcinolone injection (40mg/mL) was administered to the affected eye and a course of oral prednisone was initiated. The patient, weighing 85kg (188lbs), was to take 40mg of oral prednisone per day for two weeks, then 20mg per day for one week, then 10mg per day for one week, then discontinue.

Five weeks later, after the completion of the course of oral prednisone, the patient's pinhole VA improved to 20/50 in the operative eye. Given this improvement and no indication of elevated IOP in response to the steroid, and additional periocular triamcinolone injection was given.

Four weeks later, pinhole VA in the affected eye had improved to 20/40. The patient also reported improvement in the central scotoma, corroborated by visual field testing shown below. The mfERG was also repeated at this time, which showed improved, but still diminished, amplitudes in the affected eye (image B of Fig. 6) (see Fig. 7 and 8).

Given the continued improvement in vision and mfERG, and with IOP still within normal limits, a third periocular triamcinolone injection was given. Three weeks later, pinhole VA was improved to 20/25 -2. No additional treatment was given at this time.

Six months after SOR, the patient's BCVA was 20/30 with a −0.75 sphere spectacle refraction. The patient reported continued symptomatic improvement in the central scotoma in the affected eye, which was once again consistent with the results of visual field testing. A final mfERG was conducted at this time, revealing continually improving amplitudes in comparison to the second mfERG (image C of Fig. 6).

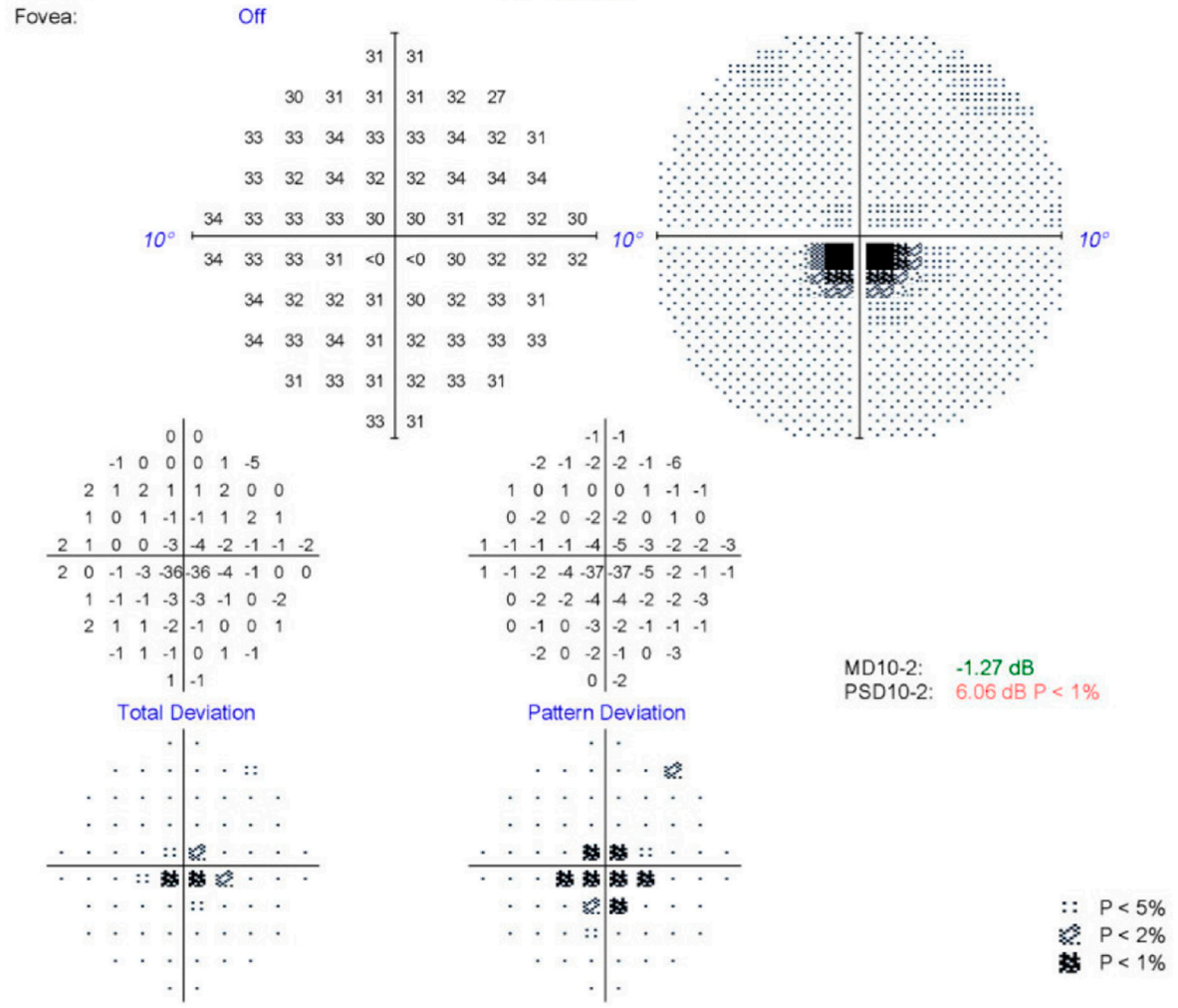

Fig. 5. Humphrey 10-2 visual field showing central scotoma seven weeks after silicone oil removal.

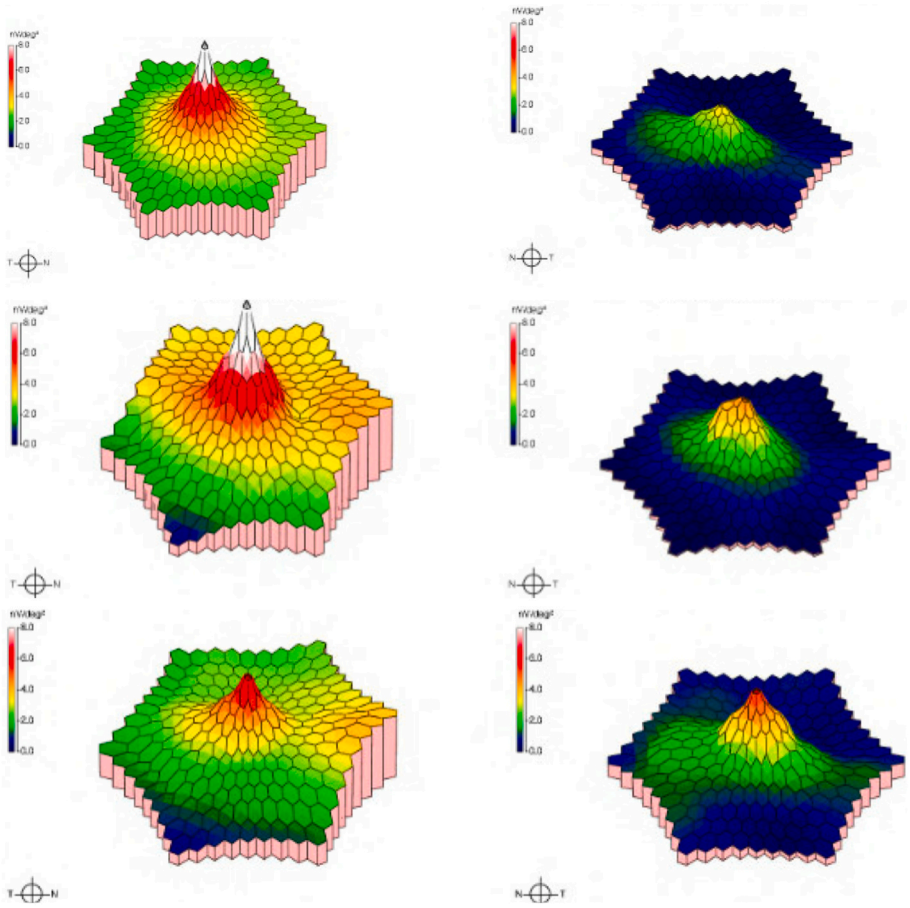

Fig. 6. Multifocal ERG results in the right eye were consistently normal on all three readings. On the initial Multifocal ERG of the left eye (image A), seven weeks after SOR, amplitudes were significantly diminished. Amplitudes in the left eye were improved from the initial result to the second result (image B), conducted after the course of oral prednisone and two periocular triamcinolone injections. Amplitudes are once again improved from the second to the third reading (image C), conducted after the administration of the final periocular triamcinolone injection.

3. Discussion

We describe here a case of UVLSOR that improved after intensive local and systemic steroid treatment. By now, UVLSOR is a well-reported, if incompletely understood, phenomenon. Various theories exist on the mechanism of injury, but findings across case series have several features in common: a severe decline in vision following SOR, usually accompanied by central scotoma, and a complete absence of explanatory findings on clinical exam and imaging studies.[2,4]

This decline in vision does not typically improve spontaneously, as our search of the literature yielded only two publications reporting spontaneous recovery of vision in UVLSOR.[3,6] Moya et al. reported that out of 14 patients who lost at least two lines of acuity due to UVLSOR, 2 recovered vision to the same level as with oil in situ. 3 more re-gained some vision following UVLSOR, although vision remained significantly diminished. Two patients' VA improved from 3/60 to 6/36, and another's improved from CF to 6/18[3]. More recently, in a case series of 11 eyes with UVLSOR, Pakravan et al. reported a mean BCVA improvement from 20/458 (1.4 ± 0.4 logMAR) 1 month after SOR to 20/219 (1.0 ± 0.5 logMAR) at the time of last follow up.[6] Given the format in which this data is presented, it is unclear what proportion of patients exhibit spontaneous visual recovery or to what extent individual patients have recovered vision, but the mean acuity remains substantially diminished from 20/72 (0.6 ± 0.2 logMAR) with oil in. Aside from these two publications, no other reports of spontaneous recovery were noted in our literature review.[1–11]

To our knowledge, there are no reports on treatment for this condition in the existing literature, and because the mechanism of UVLSOR is not understood, no obvious treatment exists to address the root cause. In deciding what kind of treatment may be reasonable, a brief review of the proposed mechanisms of vision loss in UVLSOR is warranted. One possible mechanism is damage to the retina and optic nerve from silicone oil infiltration, which has been demonstrated on imaging studies and enucleated eye specimens,[12] though it would not adequately explain why the vision loss only manifests *after* the oil is removed. Another proposed mechanism is excitotoxicity due to potassium ion hyperconcentration at the vitreoretinal interface during oil removal, though one study found no increased K+ concentration in the retro-oil space.[9]

Another proposed mechanism is damage from light toxicity which is theorized by Dogramaci to be more severe at the time of SOR.[8] While photochemical injury can be subtle on ophthalmoscopy, it generally is manifested eventually by some abnormality of outer segment structures or pigment epitheliopathy on OCT. UVLSOR, by contrast, does not demonstrate any kind of abnormality on OCT. The existing theories of a causative mechanism may be plausible, but as yet, none seems overwhelmingly likely.

Risk factors for developing UVLSOR are similarly unrevealing as to a potential mechanism. Michel described young age and macula-on retinal detachment with giant retinal tear as potential risk factors,[7] while Roca identified high intraocular pressure and long duration of oil in the eye as risk factors.[4] Data presented by Pakravan seemingly supports macula on retinal detachment as a potential risk factor, as 7 of 11 eyes (64%) with UVLSOR met this criteria.[6] It is interesting that there is a higher

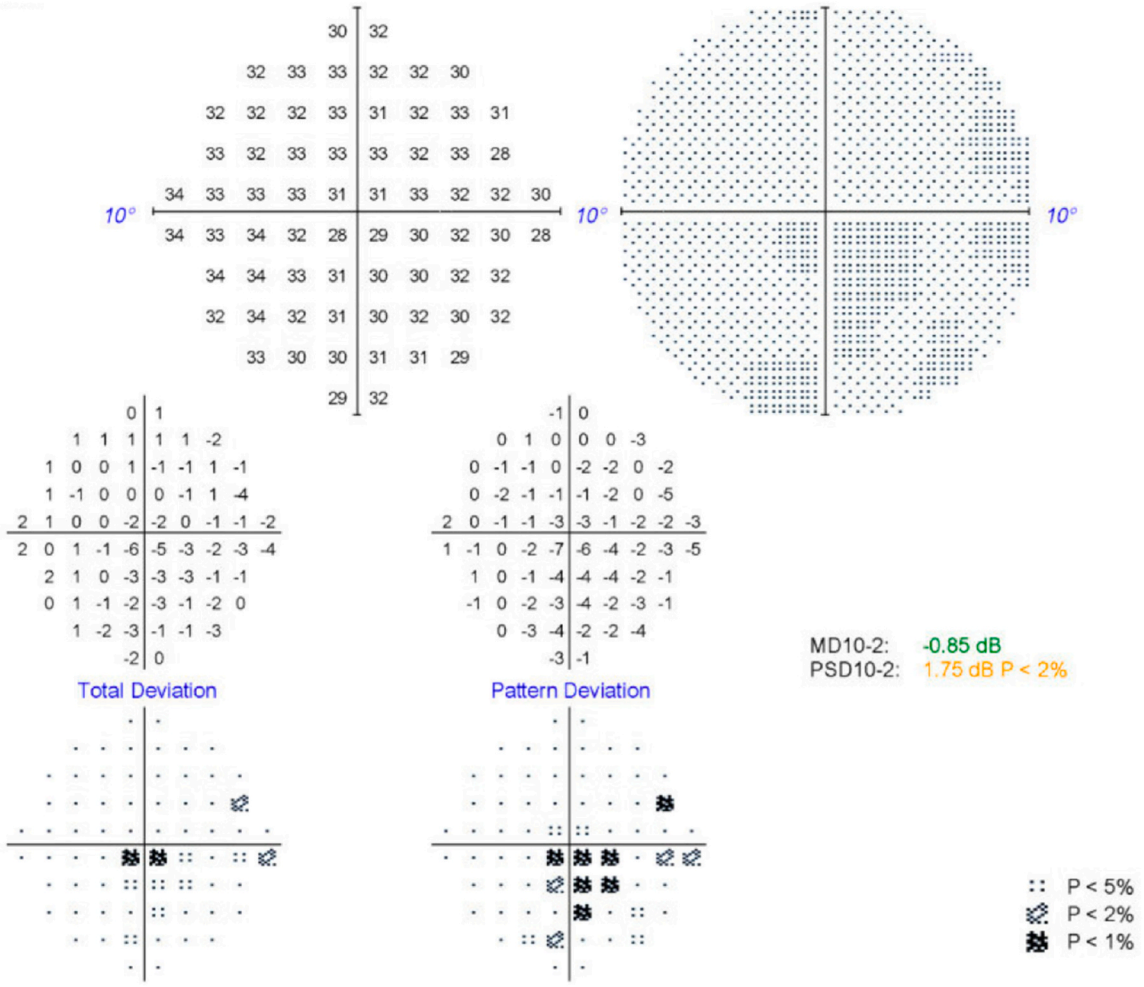

Fig. 7. Humphrey 10-2 visual field showing improvement of the central scotoma.

incidence of UVLSOR in cases of macula-on retinal detachment even though it appears the macula is the primary site of injury in UVLSOR. One possible explanation for this finding is that UVLSOR may be present but more silent in maculae that were detached; postoperative vision loss in those cases may be attributed to macular damage from the original detachment, as opposed to UVLSOR.

Without a clear mechanism of injury, we are left to speculate as to what kinds of intervention may be helpful. We understand from multiple studies, including this report, that ERG abnormalities in UVLSOR localize to the photoreceptors and middle retinal structures.[1,2,7,9] In other cases of injury and inflammation involving outer retinal structures (white dot syndromes, for example), steroids have been shown to provide some benefit.[13,14] While white dot syndromes are not exactly analogous, the utility of steroid treatment in those cases, coupled with the lack of clear options in our present case, made a reasonable case to initiate a course of steroid treatment. In this patient's previous post-operative courses, there was no IOP spike while on topical steroid, so the risk of steroid-induced ocular hypertension was thought to be acceptable. The patient was also pseudophakic, therefore eliminating another potential complication of steroid treatment.

We were pleased to see consistent improvement in subjective and objective testing after the initiation of steroid treatment. Despite the clear improvement, we cannot definitively conclude that the treatment was responsible. It is entirely possible that the improvement occurred not because of steroid treatment, but because it would have improved with the passage of time. Indeed, reported cases of UVLSOR show spontaneous improvement without treatment. However, we believe the notable scarcity of these reports, as described above, lends credence to the possibility that steroid treatment may have played a positive role in visual recovery. Nonetheless, we must acknowledge the limits of our study, namely that the treatment was only applied to one individual and there is no control data.

Despite the obvious limitations of our conclusions, the possibility of even some beneficial treatment for this condition is encouraging. Further study is needed to develop more insight into the efficacy of steroids in this application, and we believe that it is reasonable to consider steroid treatment in patients fitting the clinical profile of UVLSOR, especially given the generally poor visual prognosis and lack of other treatments.

4. Conclusions

There are no prior reports of effective treatment for unexplained vision loss after removal of silicone oil removal, but our case suggests that steroid treatment may be beneficial in similar cases.

Patient consent

The patient consented verbally to publication of the case.

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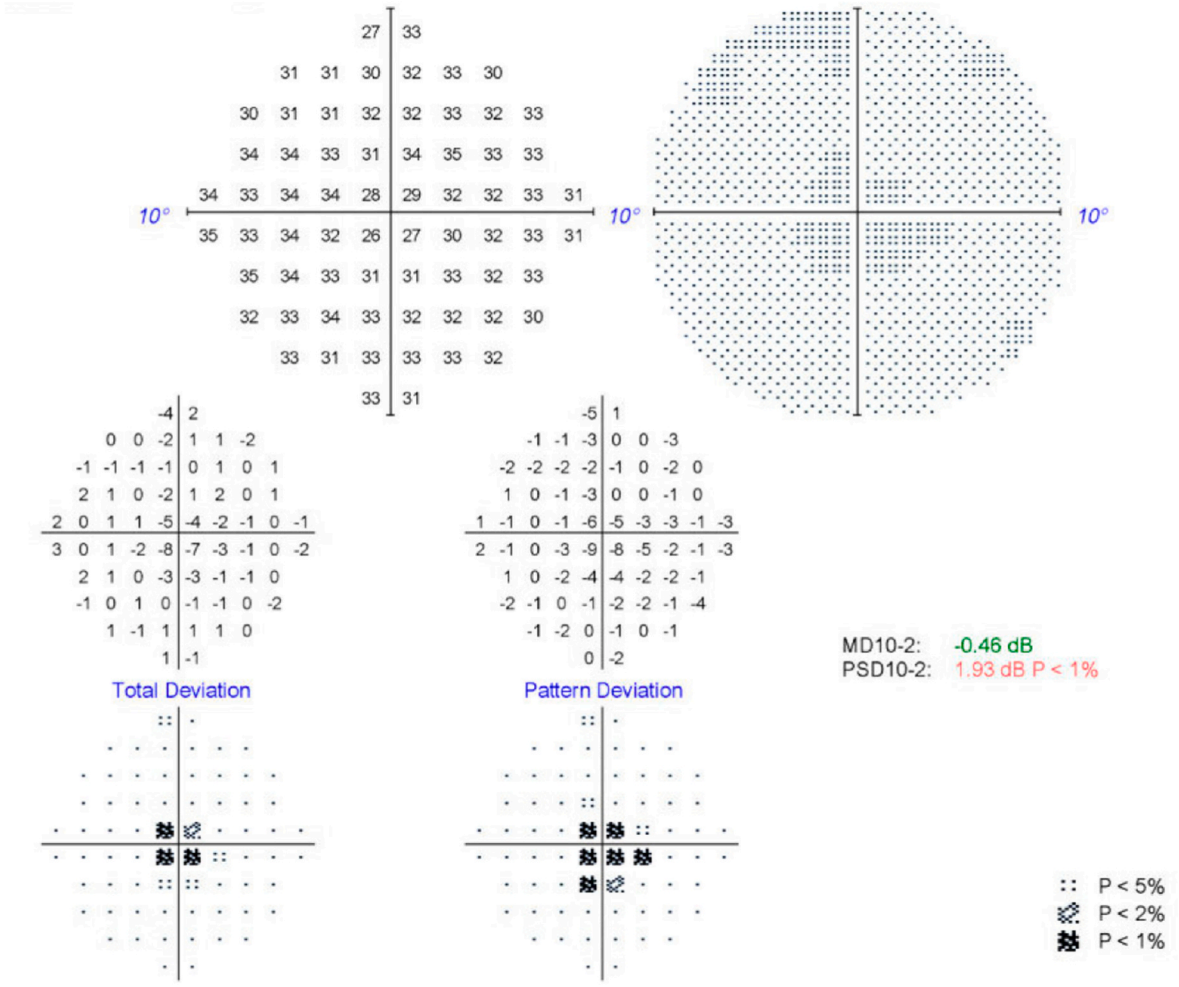

Fig. 8. Humphrey 10-2 visual field showing continued improvement of the central scotoma.

Authorship

All named authors meet the International Committee of Medical Journal Editors (ICMJE) criteria for authorship for this article, take responsibility for the integrity of the work as a whole, and have given their approval for this version to be published.

CRediT authorship contribution statement

Dane A. Jester: Writing – original draft. **Jesse M. Smith:** Conceptualization, Data curation, Writing – review & editing.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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