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Resident essay

Idiopathic uveal effusion syndrome

Joanne L. Bachman*

VA Puget Sound Health Care System, American Lake Division, USA

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Abstract

Idiopathic uveal effusion syndrome (IUES) is a rare condition characterized by a spontaneous detachment of the choroid, often with a secondary non-rhegmatogenous retinal detachment, in healthy middle-aged men. It is hypothesized that the primary cause of the condition is a congenital scleral anomaly causing abnormal uveoscleral protein transport. The following case report reviews a typical presentation of IUES. The patient was treated with lamellar sclerectomy and sclerostomy with resolution of the choroidal detachment over the next several months. Three months later, the patient developed unilateral disc swelling and a non-ischemic central retinal vein occlusion which eventually resolved without sequelae.

Keywords: Idiopathic uveal effusion syndrome; Choroidal detachment; Choroidal effusion; Sclerectomy; Sclerostomy

1. Introduction

Idiopathic Uveal Effusion Syndrome (IUES) is an uncommon ocular condition characterized by a spontaneous serous detachment of the choroid and ciliary body [1–10]. A bullous, serous non-rhegmatogenous detachment of the retina may also occur. This condition usually affects healthy middle-aged men. IUES typically presents unilaterally, but approximately 85% of patients will develop involvement of the fellow eye after a period of months to years [2]. Although the pathogenesis of this condition is uncertain, investigators have proposed that abnormal uveoscleral protein transport plays a primary role in the development of the clinical findings. It has been hypothesized that a congenital scleral abnormality exists which acts as a barrier to the uveoscleral outflow of protein [1–4]. Consequently, the impaired venous drainage of the choroid results in an outpouring of a protein rich exudate into the supraciliochoroidal space [1,2,9]. With retinal pigment epithelial cell decompensation, fluid and protein leak into the subretinal space thereby producing a serous retinal detachment. The natural course of this condition is prolonged, with remissions and exacerbations, often leading to a decline in visual acuity [1–4]. Unlike uveal effusion secondary to hypotony, topical cycloplegics and corticosteroids do not have a role in the management of patients with IUES. Since intraocular pressure (IOP) is normal and the abnormality is at the scleral level, surgical intervention utilizing a sclerectomy and sclerostomy procedure has been advocated as a possible treatment. The following case report reviews a typical case of idiopathic uveal effusion syndrome along with the pathogenesis and management of this condition.

2. Case report

A 48-year-old white male presented to the Eye Clinic with a two week history of white 'flashing lights off center, like an interference pattern' in the left eye. Although a history of direct trauma to the eye could not be elicited, he reported being hit on the left side

* 2820 Queens Way Apt #F3, Milton, WA 98354, USA.

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of his face with a soccer ball approximately 15 days prior to the examination. His medical history was remarkable for vascular headache syndrome. The last headache episode was 2 months prior and resolved after 30 days, upon initiating treatment with oral prednisone. Ocular history was unremarkable. The patient denied any other known ocular abnormalities, inflammations, injuries, or surgeries. At the time of examination, the patient was not taking any medications and reported no known medication allergies.

Ocular examination revealed best corrected visual acuities of 20/20 in the right eye and 20/30 in the left eye with minimal refractive error. Extraocular muscle movements were intact without restrictions. Pupils were briskly reactive to direct light without an afferent pupillary defect. Biomicroscopy revealed no abnormalities of the lids, lashes, conjunctiva, or cornea bilaterally. A slightly shallowed peripheral anterior chamber was noted in each eye without inflammatory or pigment cells. Intraocular pressure by Goldmann tonometry was 18 mmHg, OU. Gonioscopy showed open angles and no blood in Schlemm's canal. Vitreous examination revealed trace cells in the left eye. Dilated fundus examination was unremarkable in the right eye. The left optic nerve was noted to have mild disc edema (Fig. 1). Retinal examination of the left eye demonstrated a 360° choroidal detachment extending to the midperiphery without retinal breaks (Fig. 2). A peripheral serous retinal detachment extended inferiorly to the midperiphery. The serous fluid was noted to shift with a change in head position. The macula was not involved. B-scan ultrasonography revealed a dramatically thickened sclera bilaterally. A diagnosis of idiopathic uveal effusion syndrome was made. After consultation with several retinal specialists, a quadrantic lamellar sclerectomy and sclerostomy was performed 3 weeks later. Gradual resolution of the serous and choroidal detachments was observed over the next several months.

Three months later, the patient presented with a complaint of decreased peripheral vision in the left eye. The best corrected visual acuity was 20/20 in the

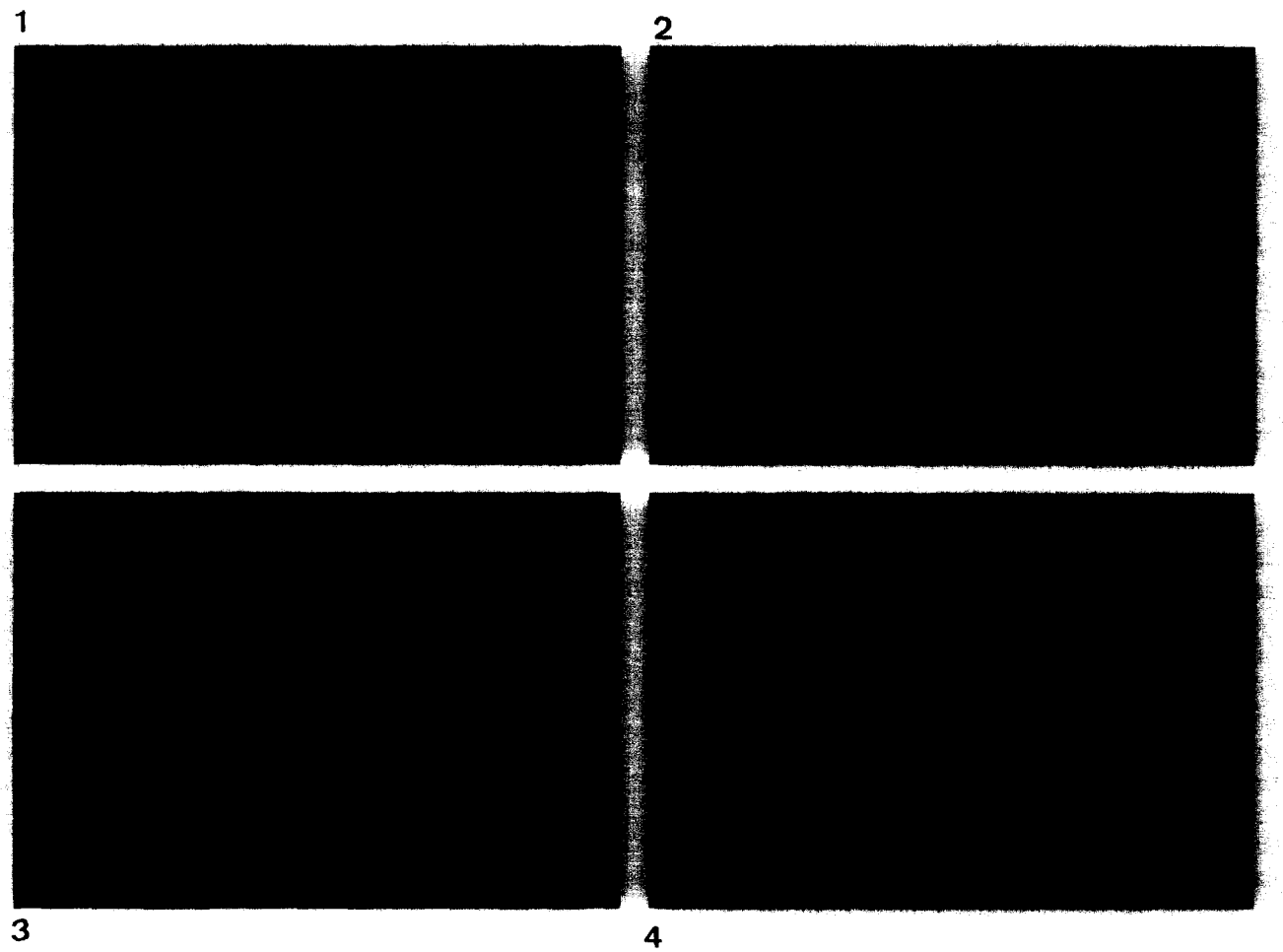

Fig. 1. Initial appearance of left optic nerve with mild disc edema in a patient with IUES.
Fig. 2 Left peripheral fundus exam reveals a ciliochoroidal annular detachment without retinal breaks.
Fig. 3 Left eye 3 months after sclerectomy surgery with moderate optic disc edema and peripapillary flame shaped hemorrhages.
Fig. 4 Leopard spots noted inferiorly in the area of the resolved choroidal detachment.

right eye and 20/25 in the left eye. The pupils were briskly reactive without an afferent pupillary defect. Intraocular pressure by Goldmann tonometry was 16 mmHg, OU. Dilated fundus examination of the right eye was again unremarkable. Examination of the left eye revealed moderate disc edema and hyperemia with peripapillary flame shaped hemorrhages (Fig. 3). The peripheral retina was flat without retinal breaks. Leopard spots at the level of the RPE were noted inferiorly (Fig. 4) in the area of the resolved choroidal detachment. The patient was followed every 1–2 weeks for the next several months with progressive hemorrhages in four quadrants and increasing venous engorgement. The patient was diagnosed with a non-ischemic central retinal vein occlusion (CRVO). The patient was advised to quit smoking and was placed on one aspirin tablet per day. The retinal hemorrhages and disc edema resolved over the next 3 months, with vision acuity stabilizing at 20/40.

3. Discussion

A ciliochoroidal or uveal effusion is an abnormal accumulation of serous fluid in the outer layer of the ciliary body and choroid [3,5,7,8]. Throughout the literature the terms effusion and detachment are used interchangeably [3]. A uveal effusion can be caused by a variety of ocular or systemic conditions, most of which will fall into one of the following four categories: (1) hydrodynamic; (2) inflammatory; (3) neoplastic; (4) idiopathic or secondary to abnormal sclera [3,5,7] (Table 1).

In the normal choroidal capillaries, the transmural hydrostatic pressure gradient is in equilibrium with the colloid osmotic pressure gradient [3,8]. Since there are no lymphatics in the eye, protein escaping from the choroidal capillaries into the extravascular space moves across the sclera by bulk flow, driven by intraocular pressure (IOP). Hypotony, which may occur after cataract extraction or glaucoma filtration procedures, has been reported as the most common cause of ciliochoroidal detachment [4]. With hypotony, the bulk flow of fluids and proteins through the sclera decreases causing an increase in the transmural hydrostatic pressure gradient. This change facilitates the leakage of fluid into the supraciliochoroidal space [3,4,10].

Table 1
Classification of uveal effusion [3]

•Hydrodynamic factors
Ocular hypotony
Wound leak
Glaucoma filtering surgery
Cyclodialysis cleft
Penetrating ocular trauma
Rhegmatogenous retinal detachment
•Inflammatory factors
After trauma or surgery
After photocoagulation or cryotherapy
Uveitis
Posterior scleritis
Vogt Koyanagi Harada Syndrome
•Neoplastic conditions
Metastatic carcinoma
Malignant melanoma
Lymphoid, leukemic, or melanocytic choroidal infiltration
•Secondary to abnormal sclera
Idiopathic uveal effusion syndrome

Ocular inflammation can produce an increase in capillary protein permeability with a subsequent leakage of protein into the extravascular space promoting fluid retention and uveal effusion [3,5]. This can occur from ocular trauma, uveitis, scleritis, intraocular surgery, pan-retinal photocoagulation or cyclocryotherapy. It is less common to see uveal effusion with neoplastic conditions, however, cases have been reported with choroidal metastatic tumors, malignant melanomas, leukemia, lymphoma, and reactive lymphocytic hyperplasia [3].

Idiopathic Uveal Effusion Syndrome (IUES) was first described by Schephens and Brockhurst in 1963 [4,5]. They reported on 17 patients, predominately middle-aged men, who spontaneously developed a ciliochoroidal detachment along with a bullous non-rhegmatogenous retinal detachment. Clinical features noted were abnormally thick sclera in six of 13 eyes and elevated cerebral spinal fluid protein in 11 of 13 patients tested [4]. In 1983, Gass hypothesized the primary cause of IUES is a congenital scleral abnormality which acts as a barrier to uvealscleral outflow of protein [1,2]. With increased resistance to transcleral protein outflow, fluid is retained within the suprachoroidal space. In the early stages, clinical findings of uveal effusion manifest as an anterior choroidal detachment with a smooth grayish-brown surface often extending in a circumferential fashion [5]. Long-standing effusion is associated with decompensation of the retinal pigment epithelium (RPE). With RPE decompensation there is leakage of fluid into the subretinal space. Accumulation of fluid and protein in the subretinal space results in a non-rhegmatogenous retinal detachment. There is a marked tendency for the serous fluid to shift with a change in head position [2,5,9]. 'Leopard spots' are large hyperpigmented spots at the level of the RPE which typically occur after a choroidal detachment has resolved.

The nature of the scleral abnormality in the IUES is not completely understood. Johnson and Gass give three different theories linking the scleral abnormality with IUES. In the first theory, the sclera is thicker

than normal. With intraoperative findings, the sclera was estimated subjectively to be thicker in 70% of cases (14 of 20) [1]. This thickening might be explained by scleral edema resulting from long-standing proteinaceous fluid in the suprachoroidal space. In the second theory, an abnormal scleral composition may contribute to the pathogenesis. Ward and associates reported on one patient with IUES to have abnormal collagen size and arrangement, and an increased acid mucopolysaccharide content [11]. In the final theory, fewer or smaller than normal scleral emissary channels may play an important role in uveoscleral protein flow in IUES. Intraoperative findings showed the vortex veins were reduced in number (less than four), caliber, or both in 68% (13 of 17) of the eyes [1]. Gass hypothesized that a vortex venous obstruction is secondary to scleral swelling from a high protein concentration [1,2].

Other clinical findings associated with IUES include mild vitreous inflammation, elevated protein without lymphocytosis in the cerebral spinal fluid, dilated episcleral veins and blood in Schlemm's canal [1–8] (Table 2). Vitreous cells could result from either mild uveal tract inflammation, passive movement of cells out of congested ciliary processes, or a macrophage response to increased protein level [4]. The high subretinal and choroidal protein concentration may be responsible for diffusion of protein into the perioptic subarachnoid space which elevates the cerebral spinal fluid protein level [2]. Signs of increased uveal pressure are dilation of episcleral veins and blood in Schlemm's canal on gonioscopy [1]. One IUES series reported 75% (15 of 20) of patients with dilated episcleral veins and 70% (seven of ten) with blood in Schlemm's canal [1]. Fluorescein angiography (FA) may show a delay in perfusion of the choroid. It can also demonstrate localized areas of leakage at the level of the RPE [4].

Most patients with IUES are middle aged at the time of diagnosis. Although it has been hypothesized that a congenital abnormality of the sclera is linked with the development of IUES, it has not been well elucidated why IUES develops in middle age. Some authors postulate protein handling becomes compromised by what would be an innocuous event for a normal eye. For example, a transient change in intravascular hydrostatic pressure or capillary permeability from slight blood pressure elevation, viral infection, mild trauma, or subclinical uveitis might shift the balance into overt ciliochoroidal effusion which would then be self perpetuating [1]. In the case reported, it is possible that the recent head trauma may have caused an iritis with associated hyposecretion of aqueous humor and reduction in intraocular pressure precipitating the development of ciliochoroidal effusion.

Table 2
Clinical features of idiopathic uveal effusion syndrome

Healthy middle-aged men
Choroidal and ciliary body detachment
Non-rhegmatogenous retinal detachment
Normal intraocular pressure
Dilated episcleral veins
Blood in Schlemm's canal
Thickened sclera with ultrasonography
Mild to moderate vitreous cells
Optic disc edema
'Leopard spots' after resolution of choroidal detachment
Elevated protein in cerebral spinal fluid

3.1. Treatment

Treatment of IUES can be very arduous. This condition is rarely improved with conventional retinal detachment repair procedures [2,4]. Systemic corticosteroids have been prescribed to reduce the choroidal detachment. However, in one study with 15 patients who were treated with high-dose systemic or periocular corticosteroids, only two resulted in transient improvement. Treatment with cytoxic agents have also had little success [1]. Laser photocoagulation to areas of RPE leakage identified on fluorescein angiography usually has no lasting beneficial effects [2,3]. It is difficult to know whether or not improving treated cases represent spontaneous resolution [1,2,4]. In one study where natural history information is available, 45% (nine of 20 cases) reattached spontaneously in 4 months to 6 years. The other 11 cases remained detached at the end of the observation period (the range of follow-up in this study was 3 months to 8 years). Although considerable disagreement exists regarding the timing of surgical intervention, some clinicians advocate surgical intervention only when the macula becomes threatened by subretinal fluid. In contrast, many retinal surgeons promote early quadrantic sclerectomies and sclerostomies. In the case reported, the patient was provided with the treatment options and elected surgery. Johnson and Gass found that quadrantic lamellar sclerectomies and sclerostomies had favorable results [1]. In their study, 23 eyes of 20 patients were studied after undergoing a rectangular 5×7 mm, 1/2–2/3 thickness sclerectomy in each quadrant, centered just anterior to the equator and placed outside the meridian of each vortex vein. A linear sclerostomy about 2 mm was made in the center of each sclerectomy bed and enlarged with a 1–2 mm scleral punch. Since suprachoroidal fluid drainage was noted at the time of surgery most ciliochoroidal detachments improved immediately post-operatively. The mean post-operative follow-up was 41 months (range 6–86 months). Resolution of subretinal and/or supraciliochoroidal

fluid occurred within 6 months in 19 eyes (83%) after 1 procedure and in 22 eyes (96%) after one–two additional procedures [1]. The mean interval of reattachment of 19 eyes with non-rhegmatogenous retinal detachments was 2.4 months with an average pre-operative detachment duration of 18.5 months. Reoccurrences developed in 23% of the eyes, all of which resolved spontaneously or with repeat sclerectomy surgery. Visual acuity improved by 2 or more lines in 13 eyes (56%), stayed stable in 8 (35%) and decreased in 2 (9%) [1]. With the absence of an unoperated control group with similar disease severity, one cannot exclude the possibility that reattachments observed were spontaneous and unrelated to surgical intervention. However, the procedure is believed to be responsible for the improvement due to the short postoperative time to reattachment.

4. Conclusion

Idiopathic Uveal Effusion Syndrome is an uncommon ocular condition characterized by a spontaneous serous detachment of the choroid and ciliary body with a secondary non-rhegmatogenous retinal detachment in otherwise healthy middle-aged men. Other clinical findings that may be present include: dilation of episcleral veins, blood in Schlemm's canal, mild vitreous cells and elevated protein in the cerebral spinal fluid. It is hypothesized that a congenitally thickened sclera is the primary underlying cause which acts as a barrier to uvealscleral outflow of protein. These patients have a protracted clinical course with remissions and exacerbations, however, surgical quadrantic sclerectomies and sclerostomies have been reported to be a relatively safe and effective treatment for patients with this condition.

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References

[1] Johnson MW, Gass DM. Surgical management of the idiopathic uveal effusion syndrome. Ophthalmology 1990;97:778–785.
[2] Gass DM. Uveal effusion syndrome: a new hypothesis concerning pathogenesis and technique of surgical treatment. Trans Am Ophthalmology Soc 1983;81:247–260.
[3] Johnson MW. Ciliochoroidal effusions. In: Margo CE, Hamed LM and Mames RN, eds. Diagnostic Problems in Clinical Ophthalmology. Philadelphia: W.B. Saunders, 1994:373–381.
[4] Gass JD, Sulayman J. Idiopathic serous detachment of the choroid, ciliary body, and retina (uveal effusion syndrome). Ophthalmology 1982;89:1018–1031.
[5] Brockhurst RJ. Ciliochoroidal (uveal) effusion. In: Ryan SJ, ed. Retina. St. Louis: CV Mosby, 1989;2:729–734.
[6] Casswell AG, Gregor ZJ, Bird AC. The surgical management of uveal effusion syndrome. Eye 1987;1:115–119.
[7] Finlay AE, Fogle JA, Green RW. Ciliochoroidal effusion. Clin Ophthalmol, 1984;4(52):1–19.
[8] Pollalis S, Tragakis M. Idiopathic choroidal detachment. Mod Probl Ophthal 1979;20:23–29.
[9] Wilson RS, Hanna C, Morris MD. Idiopathic chorioretinal effusion: an analysis of extracellular fluids. Annu Ophthalmol 1977;9:647–653.
[10] Brubaker RF, Pederson JE. Ciliochoroidal detachment. Surv Ophthalmol 1983;27:281–289.
[11] Ward RC, Gragoudas ES, Pan DM, Albert DM. Abnormal scleral findings in uveal effusion syndrome. Ophthalmology 1988;106:139–146.