

Posterior Uveal Effusion Syndrome Following Ectopia Lentis Surgery in a Case with Marfan's Syndrome

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To describe a case of acute uveal effusion with hypermetropic refraction that was limited to the posterior pole in a patient with Marfan's syndrome and bilateral ectopia lentis who underwent surgery for her left eye. A 22-year-old female with Marfan's syndrome and concomitant bilateral ectopia lentis was admitted for surgery on her left eye. On post-operation day one, we detected a shallow posterior uveal effusion (UE) and disc edema that did not include peripheral retina. Shallow posterior UE and disc edema were almost totally recovered after 3 weeks of the operation. To the best of our knowledge, this is the first case of Marfan's syndrome with concomitant ectopia lentis in medical literatures who had uveal effusion limited to the posterior pole after surgery. It is considered as Marfan's syndrome might be a predisposing factor for posterior UE.

Key words: Choroidal diseases, Lentis Ectopia, Hypermetropia, Marfan's syndrome, posterior uveal effusion syndrome.

Uveal effusion (UE) is characterized by abnormal fluid collection in the suprachoroidal area following surgery and usually occurs in hyperopic eyes with abnormally thick scleras¹. Although it is often reported after intraocular surgery, rarely, non-surgical reasons including inflammation, infections, trauma, neoplasm, drug reactions, pan retinal photocoagulation, corneal ulcer and venous congestion have also been reported.² Visual acuity is generally decreased due to the affected visual axis. It usually starts from the pars plana or the ora serrata and spreads through the eye to the equator and sometimes to the posterior pole. Uveal effusion that is limited to the posterior pole is rare^{3,4}. There is no clearly defined predisposing factor of UE. To the best of our knowledge, there are no cases with uveal effusion in Marfan's syndrome that are related or unrelated to surgery.

Marfan's syndrome is an inherited disease that affects connective tissues which provides strength,

support and elasticity to tendons, heart valves, blood vessels, ocular tissues, and other vital tissues of the body⁵. Depending on the extend of genetic expression, cardiovascular, locomotor and ocular system abnormalities may occur⁵. Ectopia lentis is the most frequent eye pathology in Marfan's syndrome⁶. In this report, we present a young female patient with Marfan's syndrome who had ectopia lentis and had posterior UE with acute hyperopic refraction following surgery of her left eye.

CASE REPORT

A 22-year-old female with Marfan's syndrome presented to our outpatient clinic with the complaint of decreased vision. The patient met the criteria for Marfan's syndrome with characteristic ocular, locomotor and cardiovascular findings. The patient with Marfan's syndrome and concomitant bilateral ectopia lentis was admitted for surgery on her left eye.

Left lensectomy, anterior vitrectomy and intraocular lens implantation using scleral fixation were planned. Retrobulbar anesthesia was applied (a mixture of bupivacaine 0.75% and lidocaine 2%). On post-operation day one, we performed funduscopy and detected shallow posterior UE and disc edema that did not include peripheral retina. There was a mild anterior chamber reaction. Anterior chamber depth was 2.95 mm. We did not detect any leakage with Seidel test. We discovered acute hypermetropic shift related to posterior shallow uveal effusion (Fig. 1). On post-operation day 1 refraction was $+14.00 +2.00 \times 80$. Best corrected visual acuity was 5/200 on the first day after the operation. Axial length decreased from 25.34 mm to 21.25. Topical antibiotic, cycloplegic agent (1% Cyclopentolate qid), topical steroid (prednisolone acetate, per hour) as well as oral steroid (prednisolone 32 mg/day) and acetazolamide (500 mg/day) were given. After the 3rd day post-operation, axial length was 22.50 mm, refraction was $+10 +2.25 \times 90$ and corrected visual acuity was 20/200. On post-operation day 10, visual acuity was 20/50, refraction was $+6.00 +200 \times 90$ and axial length was 24.01 mm. Regular post operative funduscopy showed that retinal folds and

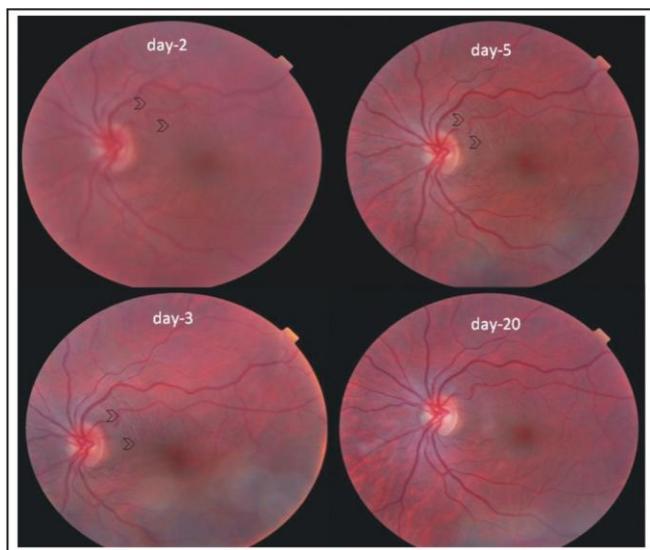


Fig. 1: Left fundus on post-operation day 2 and day 20. Disc edema and folds (arrow heads) around the optical nerve are disappearing after topical and systemic steroid treatment.

Disc edema began to disappear the first week after the operation. After 3 weeks of the operation, corrected visual acuity was 16/20, axial length was 25.16 mm

and refraction was $+1.00 +200 \times 85$. Shallow posterior UE and disc edema were almost totally recovered after 3 weeks of the operation (Fig. 2, 3).

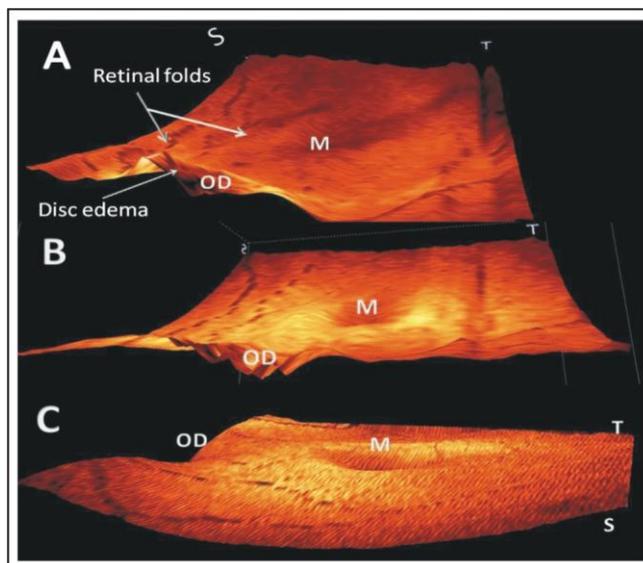


Fig. 2: A. 3-D OCT image of the left eye, on post-operation day 2.
B. Disc edema and folds around the optic nerve had resolved on post-operation day 20.
C. 3-D OCT image of a normal fellow eye of the same patient. M = macula, OD: optic disc, S: superior, T: temporal.



Fig. 3: Subluxated lens before surgery. Anterior segment on post-operation day 2 and day 20.

DISCUSSION

Uveal effusion is choroidal capillary leakage and accumulation of this fluid in the space between uvea and sclera. There is no clear, well - defined genetic predisposition of UE and risk factors are also unclear. The most important risk factor is low intraocular pressure. Also, inflammation related to surgery is also

associated with serous choroidal detachment. It can be thought that decreasing intraocular pressure and preventing inflammation may decrease the risk of UE. Inflammation increases the capillary permeability of choroid and a decrease in intraocular pressure may enhance the fluid collection in the interstitial space. Uveal effusion is related to increased capillary permeability of the choroid, decreased intraocular pressure as a result of an increased outflow of aqueous humor, disrupted scleral permeability as seen in nanophthalmic eyes and decreased production of aqueous humor as a result of iridocyclitis.

It was thought that UE is more common at the anterior side of the equator because uveoscleral connections at this part are weak⁷. The form that is limited to the posterior pole is rare^{3,4}. In our case, predisposing factors for UE included trauma of surgery, hypotony, minimal inflammation and combinations of these factors. On the other hand, Gass hypothesized that aging, hormonal changes in the collagen and ground substance of the congenitally abnormal sclera are responsible for reducing the scleral permeability to protein⁸. In time the eye becomes incapable of handling even small amounts of extravascular protein occurring from minor injuries to the uveal vasculature. In this respect it is not clear whether Marfan's syndrome, a congenital disorder of collagen that exist in vascular tissue and in the sclera, has any direct effect on the pathogenesis of posterior UE or not.

Local inflammation may provoke effusion at the posterior pole by increasing permeability in the choroidal vessels. In our case, it can be suggested that Subtenon's anesthesia may stimulate the sclera around the injection area increasing intraorbital pressure temporarily and this may induce a local inflammatory reaction that reaches to the choroid⁹. Similarly, during the retrobulbar anesthesia, retrobulbar venous flow might be damaged which could affect the choroidal circulation. Moreover, it has been shown that injected medications may pass through the choroidal circulation even if there is no retrobulbar venous damage¹⁰. Although, our patient did not report any drug sensitivity, she could have developed a hypersensitivity reaction to the injected substances (mixture of bupivacaine 0.75% and lidocaine 2%).

There are several mechanisms that can explain the unexpected hyperopic shift after surgery. Incorrect IOL power or intraocular fluid leakage causing shortening in the axial length may explain hyperopic shift. In our case, however, abnormal fluid collection

in the suprachoroidal area caused an internal elevation of the choroid and formation of folds in the retina thereby shortening the axial length. During follow-up, retinal folds disappeared and axial length reached the pre-operative levels as accumulated fluid receded.

We conclude that acute hyperopic shift following surgery for ectopia lentis in the Marfan's syndrome should cause one to suspect a posterior UE syndrome. It was observed that monitoring axial length, refractive changes, fundus imaging and proper treatment were adequate for the patient. On top of that, it is considered as Marfan's syndrome, a congenital disorder of collagen might be a predisposing factor for UE.

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