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Vitrectomy and Silicone Oil Tamponade in the Management of Retinal Detachment in Marfan Syndrome Patient

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ABSTRACT

The objective of this case report is to present outcome of vitrectomy with silicone oil tamponade in management of subtotal retinal detachment with multiple breaks in Marfan Syndrome. A case report, a 21 years old man, came to outpatient clinic with blurred vision in his both eye since childhood. Visual examination in his both eyes was hand movement, shallow anterior chamber, ocular tension of the right eye (RE) was 17.3 mmHg, left eye (LE) was 14.6 mmHg and his lens position in the RE was superotemporal direction subluxated and nasal superior direction subluxated in the LE, but both lenses part remains in the visual axis. Marfan Syndrom diagnosed based on ocular disorder, arachnodatily and arm span greater than height. Three Mirror Goldman (TMG) contact lens examination showed in the RE retinal break at 12 o' clock, multiple holes at 4 and 5 o'clock position, subtotal retinal detachment, PVR grade II; in the LE multiple hole at 11 o'clock position with PVR grade I and subtotal retinal detachment inferiorly with macular involvement. USG examination showed elongation of axial length up to 28 mm in the RE and 30 mm in the LE. Management consist of circling band, vitrectomy with silicone oil tamponade with endolaser had been performed on the left eye. One week after vitrectomy, visual acuity became two meters counting finger, retina totally attached. His RE will be operated after his LE fully recovery. Vitrectomy with silicone oil tamponade is suggested in the management of subtotal retinal detachment with multiple breaks.

***Key words**: vitrectomy, silicone oil, retinal detachment*

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INTRODUCTION

Marfan Syndrome is a connective tissue disorder with autosomal dominant inheritance that has a prevalence of 1 case per 5000 to 10.000 individuals and is caused mainly by FBN 1 mutation. The cardinal features involve the ocular, cardiovascular and skeletal system. The skin, lung and dura also may be involved.[1]

In 1998, the Gent criteria specified characteristics of the phenotype and genotype that can be assessed through history, bedside examination, imaging and molecular genetic testing. "Major criteria" carried more diagnostic weight and included features not commonly found in the general population (such as ectopia lentis, aortic root aneurysm and dural ectasia). "Minor criteria" included that were not only common in the general population but can occure together in people either by chance or a part of condition confused with Marfan Syndrome, such as some types of Ehlers-Danlos Syndrome and MASS phenotype.[2]

Ectopia lentis, bilateral and symmetrical, is present in 80% of cases. Subluxation is most frequently superotemporal, but may be in any meridian. Because the zonule is frequently intact, accommodation is retained, although rarely the lens may dislocate into the anterior chamber or vitreus. The lens may also be microspherophakic. Angle anomaly is present in 75% of eyes. It is characterized by dense iris processes and thickened trabecular sheets, and may be responsible for glaucoma, Retinal detachment associated with lattice degeneration and high axial myopia is the most serious complication, Other features include hypoplasia of spinchter dilator pupillae (rendering mydriasis difficult), peripheral iris transillumination, strabismus, flat cornea and blue sclera.[3] Major criteria is ectopia lentis (lens dislocation), minor criteria are myopia, flat cornea, iris or ciliary muscle hypoplasia.[4]

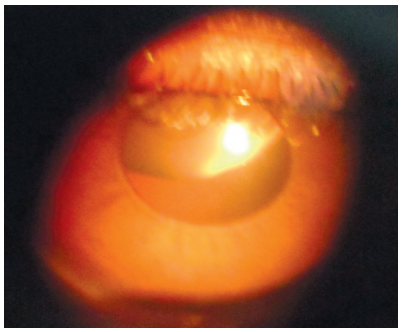
Objective: to report a case of rhegmatogenous long standing retinal detachment with PVR and pathologic myopia which must be performed vitrectomy and silicon oil tamponade, without conducting lens extraction, because of no cataract and visual part of lens remaining in the visual axis.

CASE REPORT

A 21 years old man, came to outpatient clinic with blurred vision in his both eye since childhood. Visual examination in his both eyes were hand movement, shallow anterior chamber, ocular tension were 17.3 mmhg and his lens position in the right eye (RE) was superotemporal direction sublucsated and nasal superior direction sublucsated in the left eye (LE), but both lens part remains in the visual axis.

Three Mirror Goldman (TMG) contact lens examination showed in the RE retinal break at 12 o clock, multiple hole at 4 and 5 o'clock position, subtotal retinal detachment, PVR grade II; in the LE showed multiple hole at 11 o'clock position with PVR grade I and subtotal retinal detachment inferior, with macular involvement.

Picture 1. Right eye anterior segmen: superotemporal lens subluxation



Picture 2. Left eye anterior segmen: superonasal lens subluxation

Picture 3. Right eye USG: retinal detachment

Picture 4. Left eye USG: retinal detachment

In this patient have underwent management vitrectomy and silicon oil tamponade on the left eye. A week after vitrectomy, visual acuity became two meters counting finger, retina totally attached.

DISCUSSION

This patient underwent management vitrectomy and silicon oil tamponade with indication long standing retinal detachment, subtotal retinal detachment with multiple break, patologic myopia (long axial length) and fasilitate manipulation, intraoperative retinal evaluation and left eye as priority because from Three Mirror Goldman examination found less break and PVR grade I so expected better result than right eye.

Marfan syndrome is classified in to ocular disorder, musculoskeletal, abnormal growth of bone. In ophthalmology classified in to abnormality of axial length elongation, zonullae elongation causing lens displacement because of unparallel zonullae elongation in every quadrant.

Ocular disorder in this patient are ectopia lentis: right eye superotemporal lens subluxation, left eye superonasal lens

subluxation, high axial myopia, and retinal detachment.

The type of retinal detachment is Rhegmatogenous that occurs secondarily to a full-thickness defect in the sensory retina, which permits fluid derived from synchitic (liquefied) vitreous to gain access to the subretinal space.

The typical of retinal detachment in this patient are: long standing retinal detachment, subtotal with multiple break, pathologic myopia. Management in this patient is vitrectomy and silicon oil tamponade.

Although the vast majority of simple rhegmatogenous retinal detachment can be treated successfully by scleral buckling, vitrectomy has greatly improve the prognosis for more complex detachment in which retinal break cannot be visualized as a result of haemorage, vitreus debris, posterior capsular opacity, IOL edge effect, vitrectomy is crucial to provide an adequate retinal view of all asscociated breaks. Scleral buckling carries a high risk of failure and PVR in such circumstance if any retinal breaks are missed, in which retinal breaks cannot be closed by scleral buckling such as giant tears, posterior break.[3]

Causes of failure are missed breaks. It should be emphasized that about 50% of all retinal detachment are ascociated with more than one break. In most cases the breaks are located within 90 of each other. At surgery, the surgeon should therefore not be satisfied if only one breaks has found until a thorough search has been made for the presence of other breaks and the configuration of the retinal detachment corresponds to the position of the primary break. Buckle failure may be the result of the following: inadequate size-replace, incorrectly positioned reposition, inadequate height-drain SRF or consider intravitreal gas injection.[3] Proliferative vitreoretinopathy is the most common of late failure. The traction forces associated with PVR can occasionally open old breaks and create new ones. Presentation is typically between the fourth and sixth postoperative weeks. After an initial period of visual improvement following succesfull retina attachment the patient report a sudden and progressive loss of vision which may develop within a few hours. Reopening of retinal break in the absence of PVR as a result of inadequate cryotherapy of scleral buckling. It may occur when buckle height decrease either with time or following surgical removal.[3]

Silicon oils have a low specific gravity and are thus buoyant. They allow for more controlled intraoperative retinal manipulation and may also be used for prolonged postoperative intraocular tamponade. The most commonly used liquid silicones have relatively low viscosity (1000–5000 cs). The 1000 cs silicone is easy to inject and remove whilst 5000 cs silicone is less prone to the production of tiny droplets (emulsification).[3]

Miopia in this patient is patologic because of progressing. Retina degenerating process, vitreous degeneration inducing visual acuity decrease and rhegmatogenous retinal detachment.

Simple surgical approach is imposible in this case, because of complexity, the choice of vitrectomy with silicon oil tamponade is indicated.

The procedures must be done immediately to improve visual acuity, but with consequency in the future, left eye is the priority because of better prognose than right eye.

According to literature and clinical consideration that the presence of ectopia lentis remain in the position because of no cataract finding, lens position remaining in the visual axis. It has advantages to dynamically keep vitreous to the function of retinal attachment mainly in macula area.

One week after vitrectomy, visual acuity reach two meters counting finger with better visual field, make the patient satisfied, because long visual loss in the both eyes, suggested to the patient to control routinely to anticipate complication such as cataract, glaucoma, emulsification process due to long silicon use in anterior and posterior segmen.

Endolaser performed at 11 o'clock area to prevent enlargement of infiltration subretinal fluid. Silicon oil injected with viscosity 1000 cs, sufficient for internal indentation.

CONCLUSION

Vitrectomy and silicon oil tamponade had been successfully done as management of retinal detachment with multiple break, pathologic myopia and PVR in Marfan Syndrome. Silicon oil must be monitor for long time for stabil visual acuity. The location of lens still preserved to help the function of silicon to on placed the retinal adherence. Right eye consider to be operated with worst prognose.

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