Unusual Features of Developed Capsule Contraction Syndrome in Long Term Follow up Period

Geç Gelişen Kapsül Kontraksiyon Sendromunun Nadir Görünümü

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ABSTRACT

We describe two cases have unusual features of capsule contraction syndrome who were successfully treated with surgical procedure and Nd-YAG laser capsulotomy. The first case was 65 years old man who developed CCS without any risk factors, after 4 years later followed uncomplicated phacoemulsification surgery. He treated with extraction of intraocular lense (IOL) combined with complete capsulectomy, anterior vitrectomy and anterior chamber IOL implantation. The second case was 8 years old boy had operated for cortical cataract and ectopia lentis. Anterior capsular contraction was detected in patient one year after the surgery, combined with peripheral fibrotic thickenning, reduction of total equatorial diameter of capsular bag, banding of haptics over the optic and banding over each other both ends of capsular tension ring without posterior capsular opacification. The first case, visual acuity (VA) was improved from 20/200 to 20/25 at the end of firsth month without any complication. The second case who treated with Nd-YAG laser anterior capsulotomy, VA was improved from 20/200 to 20/20 after the first day. Surgical procedures and Nd-YAG laser are seen to be effective treatmet modalities for CCS and anterior capsular opacification.

Key Words: Capsule contraction, capsular opacification.

ÖZ

Kapsül kontraksiyon sendromunun alışılmadık özelliklerine sahip, cerrahi prosedür ve Nd-YAG lazer kapsülotomi ile başarılı bir şekilde tedavi edilmiş iki olgunun sunumu. İlk olgu, komplikasyonsuz katarakt cerrahisini takiben 4 yıl sonra kapsül kontraksiyon sendromu gelişen ve herhangi bir risk faktörü olmayan 65 yaşında erkek hasta. Olguya göz içi lens ekstraksiyonuyla beraber total kapsülektomi, ön vitrektomi ve ön kamaya göz içi lens implantasyonu yapıldı. İkinci olgu, kortikal katarakt ve lens ektopisi nedeni ile opere edilen 8 yaşında erkek çocukdu. Olguda cerrahi tedaviden bir yıl sonra ön kapsül kontraksiyonu ile beraber ön kapsül kenarında fibrotik kalınlaşma, kapsüler kese ekvatoryal çap mesafesinde azalma, göz içi lens haptiklerinin ve kapsül germe halkasının her iki uç kısmının arka kapsül kesafeti gelişmeden lens optiği üzerine bağlandığı gözlendi. İlk olguda birinci ay sonunda herhangi bir komplikasyon olmadan görme keskinliği 20/200'den 20/25'e yükseldi. Nd-YAG lazer ön kapsülotomi ile tedavi edilen ikinci olguda görme keskinliği bir gün sonra 20/200'den 20/20'ye yükseldi. Cerrahi yöntemler ve Nd-YAG lazer, kapsül kontraksiyon sendromu ve ön kapsüler kesafet için etkili tedavi yöntemleri olarak görülmektedir.

Anahtar Kelimeler: Kapsül kontraksiyonu, kapsüler kesafet.

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INTRODUCTION

Capsule contraction syndrome (CCS) is progressive shrinkage of a capsulectomy opening and exaggerated reduction in capsuler bag diameter which is caused by proliferation and fibrous metaplasia of residual lens epithelial cells on the inner surface of continous curvilinear capsulorhexis (CCC).[1,2] Etiology of this clinical entity supposed to relationship an imbalance between centrifugal and centripetal forces and usually occurs in eyes with weakened zonules such as pseudoexfoliation syndrome (PEX), uveitis, pars planitis, high myopia, myotonic dystrophy, retinitis pigmentosa, diabetes mellitus. Furthermore surgical tecniques such as small capsulorhexis size and inadequate residual lens epithelial cells removement and certain intraocular lense (IOL) materials and/or designs have been reported as risk factors of CCS.[3-6] Capsuler bag shrinkage causes angulation of the haptics or optic edges and even entire IOL and this angulation lead to decentralization of the IOL.[7-9] In this article, we described two cases that developed CCS after the uncomplicated phacoemulsification surgery.

The first case with PEX has extensive traction on the zonules due to severe capsular contraction and this contraction lead to IOL subluxation four years later after the uncomplicated phaco surgery. This report describes one of the latest developed CCS that cause IOL dislocation. The second case was 8 years old boy that developed CCS one year later after the surgery without posterior capsular opacification (PCO) and the best of our knowledge this was one of the youngest case in literature.

CASE REPORTS

Case 1

A sixty-five years old male patient with PEX was underwent uncomplicated phacoemulsification surgery and posterior chamber in the bag IOL implantation. During the phacosurgery after a clear corneal incision, nearly 5.0 to 5.5 mm diameter continous curvilinear capsulorhexis (CCC) and hydrodissection was done. Phacoemulsification, cortical aspiration and posterior capsule polishing were performed respectively. A single piece hydrofobic acrylic, optic size of 6.5 mm and overall diameter of 13.75 mm foldable IOL was implanted in the capsular bag. In both, preoperative examination and during the surgery there was not any suspicious signs of zonules instability such as phacodonesis. Postoperatively topical fluoroquinolon and dexamethasone eyedrops were prescribed 5 times a day for a week. Topical antibiotic was stopped at the end of the first week and steroid were tapered gradually during the first month.

Four years later the surgery, the patient complained with distortion and gradually reduction in visual acuity. In slit lamp examination was revealed a severe fibrosis throughout the CCC and diminished equatorial diameter of capsular bag caused by peripheral fibrotic thickenning combined with tilted IOL and pseudophacodonesis. Severly inferior dislocated IOL due to dehiscens of superior zonules was detected without any history of trauma or any signs of other eye diseases.

To prevent IOL dislocation into the vitreous, IOL extraction combined with complete capsulectomy, anterior vitrectomy and anterior chamber IOL implantation were planned. Preventing total IOL dislocation into the vitreous, superior border of capsular bag was grasped by IOL forceps and gentle, side to side lateral movement was done to take out all pieces of adhesive IOL with contracted capsuler bag complexn (Figure 1).

Anterior vitrectomy was performed and pupillary miosis was achieved via injection of miostate (0.01% carbachol, ALCON). After the fulfilling in anterior chamber with VSD, anterior chamber IOL implantation and peripheric iridectomy was performed. The following day visual acuity was increased from 20/200 to 20/60 with minimum central part of corneal edema without anterior chamber reaction.

At the end of the first month, visual acuity was 20/25 with correction. During the follow up period of 6 months, whatever ophthlamic problems were not detected like IOL dislocation, iris distortion and pupillary reaction.

Case 2

An 8 years-old boy is complaining with progressive visual detoriation in his rigth eye for a few year without any history of trauma, and/or ocular inflammation.

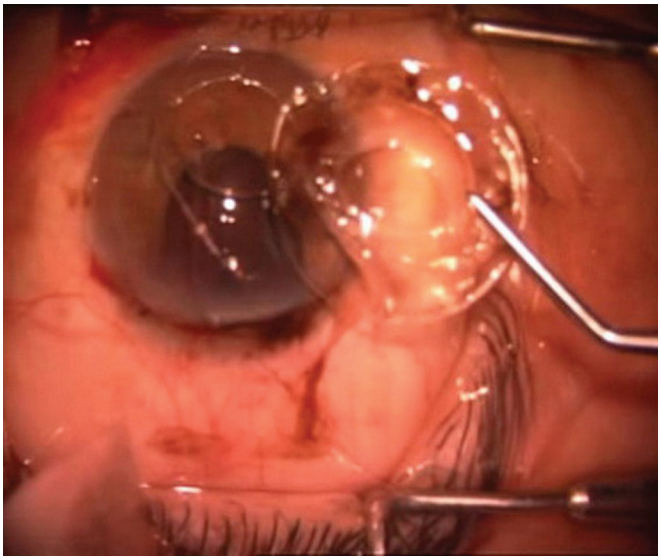

Figure 1: *IOL inside of contracted capsuler bag.*

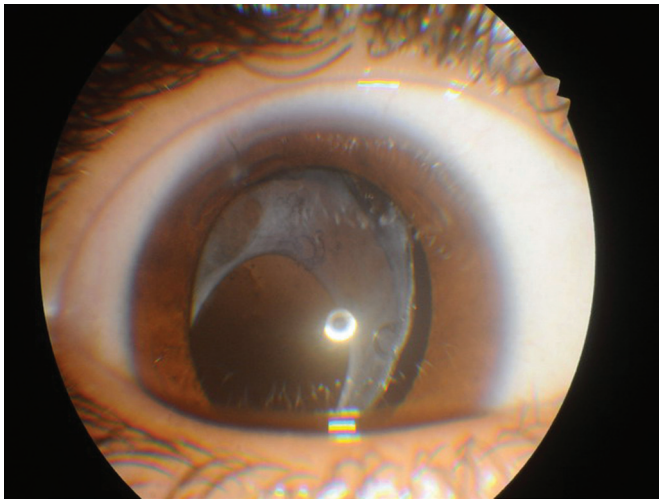

Figure 2: *Anterior capsular contraction without posterior capsular opacification.*

In slit lamp biomicroscobic evaluation was revealed that cortical cataract and ectopic lense with tendered temporally side without phacodonesis and ectopic pupilla on his right eye (RE) although both anterior and posterior segment findings were normal in the left eye (LE). No systemic associations were detected. Visual acuities were 20/200 and 20/20 in right and left eye respectively. To improve visual acuity, phacoemulsification surgery was planned in elective surgical condition. capsular tension ring (CTR) with the total size of 13 mm and made by poly methyl methacrylate (PMMA) was inserted in the capsular bag, than phacoemulsification, irrigation and aspiration of cortical material and posterior capsule polishing were performed and one piece hydrophobic acrylic foldable IOL with an optic size of 6.5 mm and overall diameter of 13.75 mm was implanted in the capsular bag. With the help of CTR, phaco surgery and IOL implantation performed without any complication and IOL position was more centralized than comparing the preoperative cristalline lens position. Visual acuity (VA) was improved from 20/200 to 20/32 at postop first week and 20/20 with correction of +1.25-1.50 axis of 130° at the end of the first month.

Postoperative period was uneventfull during the six months except precence of moderate fibrosis from equatorial part of capsular bag to capsular openning on the nasal side. Although visual acuity was still 20/20, one year later after the surgery patient was complained with reduction in visual acuity and detoriation of vision in his operated eye. On slit lamp evaluation was revealed that a significant anterior capsular fibrosis and decreased in size of capsular openning combined with peripheral fibrotic thickenning, reduction of total equatorial diameter of capsular bag, banding of haptics over the optics and banding over both ends of CTR each other without posterior capsular opacification (Figure 2).

Furthermore pseudophacodonesis, dehiscens of zonules and IOL dislocation was not detected. To open capsular closure and improve visual acuity anterior capsulotomy by Nd-YAG laser was performed VA improved from 20/200 to 20/40. To prevent anterior chamber reaction and elevation of intraocular pressure during the absorbtion of fibrotic capsuler particles, topical steroid (Loteprednol etabonat 5 mg/ml) was administered 5 times a day for a week. VA was improved 20/20 at first week, capsulotomy opening side persisted without anterior chamber reaction and normal IOP during the follow up period of one year.

DISCUSSION

CCS develops due to excessive shrinkage of the anterior capsule and reduction of the equatorial diameter of capsular bag. Histopathologic and ultrastructural evaluations are shown that this syndrome caused by proliferation and fibrous metaplasia of residual lens epithelial cells that result of an increased reperative reaction to the lens disruption. Several chemical factors including prostoglandine, basic fibroblast growth factor, interleukins such as IL-1 and IL-6, which are produced by the residual lens epithelial cells, increase in humor aqueous after cataract surgery and these chemical factors stimulate proliferation of lens epithelial cells, which mainly located in the equatorial region, by alterations in cell to cell contact interactions and induce fibrous metaplasia. Transforming growth factor-βs (TGF-βs) which are related to scar formation are present in the aques humor too.[2-5]

Pseudoexfoliation syndrome which has lead to impair the blood-aques barrier and subsequently heavy leakage of protein and other chemical substances from the iris vessels to humour aqueous that increased the risk of capsular fibrosis.[2] The process of CCS is progress (or progressses?) in four stages, the most advanced stage (Stage 4) characterized with eccentric displacement of the CCC and IOL decentration due to excessive and asymmetric shrinkage of capsular bag as we observed in our patient with PEX.[5] Even the intensity of fibrosis is maximum in the early repairement phase of capsular damage during the early postoperative period, the process may continue for several months.[3,10-14] Although the earliest contraction of CCC in postop. second week in eyes with uveitis have been reported, significant decrease in capsulorhexis size usually occures during the first 3 months postoperatively. [2,7] Cochener at al.,[15] were repoted that contraction was significantly greater at 30 to 150 days with a slowly progression 5 months after surgery. Gallagher and Pavilack at al.,[3] reported a total of 6 case who developed CCS, four of these eyes had preoperative PEX and 5 patietnt of them were involved preopertive zonular laxity.

Only one patient, without zonular laxity, did not noticed a bluring in vision until postoperative 5 months whereas most of others noticed a significant visual detoriation in 3 months after surgery. In their series, patient without zonular laxity was developed CCS later than others who had preoperatively zonular laxity.

Deokule at al.,[5] were observed severe CCS and performed Nd: YAG laser capsulotomy up to 24 month after cataract operation with the mean duration time of 14±2.5 months. Similarly, our second case which is 8 year-old, developed significant fibrosis and CCS later than 6 months postoperatively even with high potential risk of epithelial cells proliferation due to young age so we performed Nd: YAG laser capsulotomy in 12 months postoperatively.

Our first patient without any obvious preoperative risk factor rather than PEX, did not noticed visual detoriation until postoperative four years. It is a well known information that following years later IOL with fibrotic capsular bag may fall in vitreus cavity with contraction mainly in patient with already compromised zonules such as high myopia reported by Fernández-Buenaga R et al.,[16] According to the our observation, slowly progressive condition of CCS may continue longer than few months as its reported before and symptomps may occur later even with risk factors such young age or PEX.

A markedly decreasing in the capsulorhexis diameter and decentration usually occurs in eyes with weakened zonules such as pseudoexfoliation, uveitis, advanced age and eyes had history of trauma.[3-5] The weakened zonules can not resistant, which comes to the excessive contraction force, created from microflaments of myofibroblasts that cause shrinkage of the capsular openning. Asymmetric extreme contraction of capsular openning may cause tilting or decentration of the IOL and in severe cases IOL dislocation may occure due to complete segmental zonular dehiscence rings as we observed in our first case with PEX.

To achieve the correct position of ectopic lense in second case, we implanted CTR without scleral fixation. Nonabsorbable sutures in scleral fixation have potential risk of scleral erosion during long term follow up especially in patietnts with long life expectancy such as our 8 year-old case. Subluxation and/or phacodonesis were not observed in our case, therefore ectopic lense may not lead to zonular weaknes or dehiscens and because of potential risk of CTR fixation in to the sclera we didn't prefer to use it. Even CTR may increase resistance to centripedal tractional force from capsular contraction, it does not invariably preventing capsular shrinkage as we obsereved in our case.[14,17]

In our first patient, zonular weakening may become obvious in time both due to increased in age and also progressively increasing destructive effect by pseudoexfoliation material. IOL subluxation in case one was due to these imbalance between slowly progressive increased contraction effect of fibrotic capsule and progressively decreasing zonular tension which occured later than expected postoperative period compared to different reports by other authors. According to our observation the postoperative follow up period should be longer and patient had to be informed about long term risks of CCS.

CCS can occur with a variety of IOLs but lower rates were reported with lenses made by acrylic materilas. Adhesive reaction between the acrylate optic and the both anterior and posterior capsule give rise to posterior capsular opacification (PCO) as well as CCS.[2,8,9] Patient with young age had increased risk of PCO due to high proliferative potential of equatorial cells. But in our case that age of 8 year, PCO did not develop with the help of tight adhesion between hydrophobic acrylic IOL and posterior capsule.

Developement of CCS without PCO may explaine with diffusion of cytokines and other proliferative substance from free edge of anterior capsule openning to equatorial region and to stimulate fibrouse proliferation in potential space between IOL and anterior capsule. As far as we know ours is one of the first case who developed CCS wihout PCO in a young age group in the literature.

Because of their adhesive nature, manupilation of acrylate lenses could be difficult in the capsuler bag. Our first patient had severe adhesion between acrylic IOL and both anterior and posterior capsule and significant reduction on capsuler bag diameter result of advanced capsular fibrosis. Therefore surgical capsulectomy combined with scleral fixation on defective zonular segment may have additional damage to already compromised zonules. So we prefered to remove the IOL with the fibrotic capsular bag and performed IOL implantation in to the anterior chamber.

In conclusion, to our knowledge our second case is the one of the youngest case who had CCS even with hydrophobic acrylic lens and CTR and also without development of PCO. So both CTR and acrylic IOL may not prevent CCS when it prevents PCO in young patients and also as far as we know our first case is one of the latest case who developed CCS and presented with IOL subluxation.

We suggest that closely follow young patients and eyes with appearent or suspected zonular compromise such as PEX in not only early but also longer postoperative period.

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