JKAU: Med. Sci., **Vol. 15** No. 1, pp: 15-24 (2008 A.D. / 1429 A.H.)

Phacoemulsification Surgery in Patients with Vogt-Koyanagi-Harada Syndrome

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Abstract. The aim of this study was to evaluate the efficacy of cataract surgery with phacoemulsification technique in patients with Vogt-Koyanagi-Harada syndrome in Saudi Arabia, at the King Abdulaziz University Hospital and Maghraby Eye Centre, Jeddah, Saudi Arabia. A retrospective analysis of the records of patients diagnosed with Vogt-Koyanagi-Harada syndrome that underwent cataract surgery with phacoemulsification technique between March 2002 and September 2007 was carried out. Phacoemulsification surgery was performed in 13 eyes of 7 patients. 6 patients needed bilateral surgery. There were 3 males and 4 females. One patient was left aphakic in both eyes. In the remaining 11 eyes Acrysof foldable intraocular lens was implanted in the bag. The mean age of presentation was 39.57 ± 6.55 years. The patients were followed up for 8.33 ± 3.7 months. Mean improvement of visual acuity was 6.69 ± 2.46 lines. All patients were monitored closely. The time taken for the quieting of anterior chamber was 4.23 ± 1.92 weeks. Two patients developed significant posterior capsular opacity which needed Neodymium-YAG capsulotomy. There were no other complications. The cataract surgery with phaco-emulsification technique was safe and effective in the patients of Vogt-Koyanagi-Harada, provided they were managed effectively during and after surgery.

Keywords: Vogt-Koyanagi-Harada syndrome, Cataract surgery, Phacoemulsification, Acrysof intraocular lens.

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Accepted for publication: 15 March 2008. Received: 22 January 2008.

Introduction

The Vogt-Koyanagi-Harada syndrome (VKH syndrome) is an idiopathic multisystemic disorder, which typically affects pigmented individuals[1]. It is characterized by chronic bilateral progressive panuveitis with extraocular manifestations involving the central nervous, auditory, and integumentary systems[1,2].

VKH syndrome has been reported throughout the world but it is seen predominantly from people of oriental descent.

In Saudi Arabia, it was found as the second most common uveitis entity after Behcet's disease[3]. However, recently Islam and Tabbara found that VKH syndrome is the third most common cause of diffuse uveitis after Behcet's disease and sarcoidosis[4].

VKH syndrome has an immunogenetic cause involving a delicate interaction between both delayed hypersensitivity and humoral responses of T and B lymphocytes as well as other genetic factors as yet undetermined[1].

The mainstay of treatment consists of either high doses of systemic steroids or immunosuppressives if the patient develops steroid intolerance or resistance[5].

Cataract is the most frequent complication[2]. It results from recurrent anterior uveitis and may be as a result of long term steroid therapy[6]. The incidence of cataract in patients of VKH syndrome ranges from 30% to 38%[6]. The other vision threatening complications are glaucoma and subretinal neovascular membrane[5,7].

In view of chronic recurrent inflammation, most cataracts are associated with posterior synechiae. The intraocular pressure initially tends to be reduced. However, with pupillary seclusion and occlusion resulting from posterior synechiae, there is pressure elevation that leads to secondary pupillary block glaucoma with neovascularization of the iris and angle late in the course of the disease[8].

The surgical outcome for cataract extraction in patients with VKH syndrome depends upon the duration and activity of the inflammation[9]. Various post-operative complications like pupillary membranes, pupillary block glaucoma, cystoid macular edema, subretinal gliosis, and phthisis bulbi have been reported[6,9].

The timing and selection of the treatment for these complicated cataracts have been controversial[10]. Equally, the intraocular lens (IOL) implantation and the choice of the IOL are topics for debate[6,9-11].

In this retrospective study the outcome of cataract surgery with phacoemulsification technique with Acrysof foldable IOL in the patient population was evaluated.

Material and Methods

This study was a retrospective review of the records of patients who underwent cataract surgery with phacoemulsification technique by the author in King Abdulaziz University Hospital and Maghraby Eye Center, between March 2002 and September 2007. All of the patients had a confirmed diagnosis of complete VKH syndrome based on the criteria suggested by the international committee on nomenclature at the First International Workshop on Vogt-Koyanagi-Harada Disease in October 1999[12].

Pre-operatively, all of the patients had a complete ophthalmic examination comprising best-corrected visual acuity (BCVA) using Snellen charts. Anterior segment evaluation was done with a Haag-Streit slit lamp (1.6 mm × 1.0 mm beam) to determine the presence and absence of anterior chamber (AC) and vitreous inflammation, extent of posterior synechiae, pupil dilation, type of cataract and intraocular pressure (IOP) by applanation tonometry. Fundus evaluation was done by slit lamp biomicroscopy with 78 D lens and indirect ophthalmoscopy. In patients with dense cataract with hazy media, ultrasonography was done to assess the posterior segment.

The patients' records were reviewed for the patients' details such as: age, sex and the laterality of cataract, duration of VKH syndrome, pre-operative medications, interval between the last inflammatory episode and surgery, duration of post-operative follow-up, pre- and post-operative visual acuities, intra- and post-operative complications, and post-operative Fundus findings.

The patients who did not have immediate post-operative follow-up evaluation were excluded from the study.

The patients who were free of inflammation for 3 months before surgery were included for this study. Inflammation was controlled by

systemic, periocular and topical steroids and Azathioprine (Imuran) 1.5-2.0 mg/kg/d.

The absence of anterior chamber inflammation was confirmed at the last pre-operative visit *i.e*., 3 d before surgery.

All of the patients were started on prednisolone 1 mg/kg/body weight for 3 d before surgery. The prednisolone was tapered slowly over 4 to 6 wk post-operatively depending on the degree of ocular inflammation. Topical prednisolone acetate was started for 3 ds before surgery and slowly tapered depending upon the degree of inflammation.

All of the surgeries were performed by standard phaco-emulsification technique under peribulbar anaesthesia (Xylocaine 2% and Marcaine 0.5%). Surgery involved making a clear corneal incision, synechiolysis with Iris spatula under Healon, stretch pupilloplasty to dilate the pupil if necessary, capsulorhexis was done with forceps, nucleus sculpting and emulsification by the divide and conquer technique. The cortical material was aspirated by Simcoe cannula after which posterior chamber IOL was implanted in the capsular bag. At the conclusion of surgery sub-conjuctival injection of Methyl prednisolone (40 mg/ml) and gentamycine (25 mg/ml) was given to all patients.

Post-operatively, all patients were treated with topical antibiotics and topical and systemic steroids, which were tapered over 4 to 6 wk. The frequency of the topical and the dose of systemic steroids were increased when a fibrinous reaction was noted.

Patients were examined at 1 and 7 d, 4 wk, 8 wk and 3 and 6 mths post-operatively. At the initial post-operative visits, in addition to visual acuity and IOP, the integrity of corneal section, corneal status, AC inflammation and the presence or absence of posterior synechiae and posterior capsular opacification were noted. Subsequent post-operative visits comprised a complete ophthalmological evaluation including BCVA, slit lamp evaluation, applanation tonometry and a detailed Fundus evaluation by indirect ophthalmoscopy.

Results

Thirty-five patients with VKH syndrome were examined between March 2002 and November 2007. Among them, 12 patients had visually significant cataract. Nine of the 12 patients underwent cataract surgery.

Two patients did not have sufficient follow-up and hence was excluded from the study. Among the remaining 7 patients, 6 had bilateral cataract surgery. Hence, we had 13 eyes in 7 patients. There were 3 males and 4 females.

The mean age of presentation was 39.57 ± 6.55 (range 29-46) years. The patients were followed up for 8.33 ± 3.7 (range 2-15) mths.

Table 1. Patient demography, anterior chamber inflammation, post-operative complications.

Patient no.	Age	Eye	AC cells				Complications	Follow-up
			Day 1	Week				
				1	4	8		
1	29	OD	1+	1/2+	rare	Quite	nil	12
		OS	1+	rare	rare	Quite	nil	9
2	46	OD	4+	2+	rare	Quite	nil	12
		OS	1+	rare	rare	Quite	nil	2
3	46	OS	3+	1+	rare	Quite	nil	12
		OD	3+	1/2+	rare	Quite	PCO ++	6
4	36	OD	2+	1/2+	rare	Quite	nil	15
5	40	OD	3+	2+	rare	Quite	nil	12
		OS	2+	1/2+	rare	Quite	nil	6
6	35	OS	4+	2+	1+	Rare	PCO ++	8

Pre-operatively, posterior synechiae involving in 3 quadrants were noted in 11 eyes. Both eyes of one patient had posterior synechiae in 4 quadrants (Patient No.1). Mixed cataract was noted in 11 eyes (posterior sub capsular and nuclear); the other 2 eyes had total cataract (Patient No. 1). All of the patients were free of inflammation for 3 months pre-operatively. The de-pigmented Fundus lesions suggesting of VKH syndrome was seen in 11 eyes. In the remaining 2 eyes, Fundus view was not possible which necessitated ultrasound evaluation.

Intra-operatively, synechiolysis was performed by iris spatula. The pupils were dilated using stretch pupilloplasty. All of the patients underwent cataract surgery with phacoemulsification technique. One patient (No. 1) with dense cataract with occlusio-pupillae, the IOL

implantation was differed upon the patient decision. Subsequently his fellow eye was also left aphakic. In the remaining 11 eyes, Acrysof foldable IOL was implanted in the bag. Peripheral iridectomy was done in the eyes which were left aphakic. Visual acuity improved in all patients (Tables 2, 3, and 4). Mean improvement of visual acuity was 6.69 ± 2.46 (range 2-11) lines (Table 3). 62.5 % patients had visual acuity better than 20/30 (Table 3). There were no cases without improvement of visual acuity.

Table 2. Pre-operative visual acuity.

Preoperative VA	No. of eyes
<20/200	7
20/100-20/80	6
20/70-20/40	0
>20/30	0

Table 3. Post-operative visual acuity.

Post-operative final	No. of eyes
<20/200	0
20/100-20/80	0
20/70-20/40	8
>20/30	5

Table 4. Post-operative visual lines improvement.

Visual lines improvement	No. of eyes
2 to 5	4
6 to 9	7
>10	2

Two patients (Patients No. 3 and 10, Table 4) had fibrinous reaction in the immediate post-operative period, which was resolved with aggressive treatment. The time taken for the quieting of AC was 4.23 ± 1.92 (range 1 to 8) wk. Two patients (Patients No. 6 and 10, Table 4) developed significant posterior capsular opacity (PCO), which needed neodymium-doped yttrium aluminum garnet (ND:YAG) laser capsulotomy. There were no other complications.

Discussion

The outcome after cataract surgery in VKH syndrome depends upon various factors like strict pre-operative and post-operative control of inflammation and meticulous surgical planning and technique[6]. The improvements in the quality of surgical technique and the IOL material and design have changed the management of cataract in the uveitis patients[9].

Medline search revealed few studies which evaluated the surgical outcome of cataract in VKH syndrome. At present, this is the first study which has evaluated phacoemulsification surgery in VKH syndrome in the Middle East. In one retrospective study from Saudi Arabia, Tabbara *et al.* compared the course and visual outcome of VKH syndrome in children versus adults[13]. In their study the ocular complications and visual loss were found to be more severe in children than adults despite surgical and medical therapy.

The challenges in operating such high risk eyes are the presence of dense posterior synechiae, non-dilating miotic pupil, post-operative pupillary membranes, potential risk for recurrent and prolonged post-operative uveitis, secondary glaucoma and phthisis bulbi.

In this study, all patients were free of inflammation 3 months before surgery. Azathioprine (Imuran) 1.5-2.0 mg/kg/d was used in all patients, in addition to systemic and local steroids while controlling the inflammation.

Among 13 eyes, 11 eyes had posterior synechiae involving 3 quadrants and in 2 eyes there were 360 degrees synechiae. Intra-operatively, synechiolysis was performed by Iris spatula and pupillary dilation achieved by stretch pupilloplasty.

Phacoemulsification has advantages in dealing with these complicated cataracts as it leads to less breakdown of blood-brain barrier. A continuous curvilinear capsulorhexis ensures in-the-bag IOL placement in the majority of cases[9]. In this study, all patients underwent phacoemulsification surgery.

The use of highly bio-compatible IOLs such as foldable acrylic and heparin surface modified (HSM) IOLs can lessen the chances of post-operative posterior synechiae and PCO[9]. In this study we implanted

Acrysof foldable IOL in 11 eyes of 6 patients. None of our patients developed posterior synechiae post-operatively. Two patients developed PCO, which needed YAG capsulotomy later.

Pre-operatively, all patients (13 eyes) had visual acuity less than 20/80 and 7 eyes (53%) had visual acuity less than 20/200 (Table 1). Post-operatively, all patients improved visual acuity better than 20/70. 5 eyes (38%) had visual acuity better than 20/30 (Table 2). Mean improvement of visual acuity was 6.69 ± 2.46 lines. Two eyes showed visual acuity improvement by 10 lines (Table 3). All of the results were comparable to previous studies[6,9].

All of the patients had post-operative inflammation ranging from 1+ to 4+ cells in the immediate post-operative period. The time taken for the quieting of AC ranged from 1 to 8 wk. (M 4.23 ± 1.9 wk) (Table 4).

Being a retrospective study, this study also suffers from its inherent drawbacks. However, this is a major study dealing with cataract surgery with phacoemulsification technique in VKH syndrome, in this part of the world.

To conclude, cataract surgery with phacoemulsification technique is safe and effective in patients of VKH syndrome with quiet eyes provided they were managed efficiently during intra-operative and post-operative periods.

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جراحة استحلاب العدسة عند مرضى متلازمة فوجت كويناجي هارادا

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المستخلص. تقييم طريقة استحلاب العدسة عند مرضى متلازمة فوجت كويناجي هارادا في السعودية. مستشفى الملك عبدالعزيز الجامعي ومركز مغربي للعيون – جدة السعودية.

المرضى وطريقة البحث: تحليل ومراجعة ملفات المرضى بمتلازمة فوجت كوينا جي هارادا، والذين أجريت لهم جراحة لاستئصال المياه البيضاء بطريقة استحلاب العدسة في الفترة الواقعة بين مارس ٢٠٠٢ وحتى سبتمبر٢٠٠٧م.

نتائج البحث: تم إجراء جراحة استحلاب العدسة في (١٣) عين عند ٧ مرضى، ٦ مرضى احتاجوا الجراحة في كلتا العينين. كان منهم ٣ ذكور و٤ إناث. أحد المرضى تم تركه بدون زرع عدسات فى العينين، وباقى العيون الإحدى عشر، تم زرع عدسات أكريسوف مطوية في محفظة العدسة. متوسط الأعمار كان (٣٩,٥٧ + ٦,٥٥) سنة. تم متابعة المرضى لمدة (٨,٣٣ + ٣,٧) شهر. وكان متوسط تحسن حدة البصر (٦,٦٩ + ٢,٤٦) خط من لوحة سنلن. استغرق صفاء الخزانة الأمامية للعين (٤,٢٣+١,٩٢) أسبوع بعد الجراحة. مريضان فقط حصل لديهم كثافة على المحفظة الخلفية للعدسة. لم يكن هناك أية اختلافات أخرى.