The Feasibility of 3 Different Uveitis Scoring Systems in the Evaluation of Behcet and Idiopathic Uveitis with Posterior Segment Involvement

Arka Segment Tutulumlu Behçet ve İdiyopatik Üveitlerde 3 Farklı Skorlama Sisteminin Uygulanabilirliği

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ABSTRACT

Purpose: To investigate the feasibility of 3 different scoring systems (uveitis activation score (UAS), fluorescein angiography scoring system (FASS), and the Behcet disease ocular attack score 24 (BOS24)) in the evaluation of Behcet and idiopathic uveitis with posterior segment involvement.

Material and Methods: This study enrolled 85 eyes with panuveitis or posterior uveitis. Forty-five eyes which were associated with Behcet disease constituted Behcet group and 40 eyes which were not associated with any systemic disease constituted idiopathic group. Groups were similar ($p>0.05$) according to age, gender, anatomic localization, duration of disease, duration of follow-up, and best corrected visual acuity (BCVA). Three different uveitis scoring systems including UAS, FASS and BOS24 were compared between the groups. Correlations between BCVA and 3 scoring systems were also performed for each groups.

Results: Bilaterality was significantly higher in Behcet group when compared with idiopathic group (91.1 % vs. 70.0 %, $p = 0.01$). None of the uveitis scoring systems revealed significant difference between groups ($p>0.05$) but the difference was greatest with the BOS24 scoring system ($p=0.19$). BCVA and all 3 scoring systems were moderately and significantly correlated in both groups ($p<0.05$). The highest correlation of BCVA was observed with BOS24 scoring system in both Behcet ($r=-0.63$, $p<0.01$) and idiopathic group ($r=-0.63$, $p<0.01$).

Conclusion: Uveitis grading systems are comprehensive, reproducible, and feasible ways for measurement of inflammation in uveitis with posterior segment involvement.

Key Words: Uveitis activation score, the Behcet disease ocular attack score 24, fluorescein angiography scoring system, Behcet uveitis, idiopathic uveitis.

ÖZ

Amaç: Arka segment tutulumlu Behçet ve idiyopatik üveitlerde 3 farklı skorlama sisteminin (üveit aktivasyon skoru (UAS), floresein anjiyografi skorlama sistemi (FASS) ve Behçet hastalığı oküler atak skoru 24 (BOS24)) uygulanabilirliğini araştırmak.

Gereç ve Yöntemler: Bu çalışma panüveitli veya arka üveitli toplam 85 gözü içermektedir. Behçet hastalığıyla ilişkili 45 göz Behçet grubunu ve hiçbir sistemik hastalıkla ilişkisi olmayan 40 göz idiyopatik grubu oluşturdu. Gruplar yaş, cinsiyet, anatomik yerleşim, hastalığın süresi, takip süresi ve en iyi düzeltilmiş görme keskinliği (EİDGK) açısından benzerdi ($p>0.05$). Gruplar UAS, FASS ve BOS24'ü içeren 3 farklı üveit puanlama sistemine göre kıyaslandı. Her iki grup içinde de görme keskinliği ile 3 farklı puanlama sistemi arasında korelasyon yapıldı.

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Geliş Tarihi - Received: 10.01.2017
Kabul Tarihi - Accepted: 12.01.2017
Ret-Vit 2017; 26: 330-334

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Bulgular: Bilateralite Behçet grubunda idiyopatik gruba göre anlamlı olarak yüksekti (%91.1'e %70.0, p = 0.01). Gruplar arasında üveit puanlama sistemlerinin hiçbirisi farklılık göstermedi (p>0.05) fakat BOS24 puanlama sistemi ile fark en fazla idi (p=0.19). Her iki grupta da EİDGK ile 3 puanlama sistemi orta seviyede ve anlamlı korelasyon gösterdi (p<0.05). EİDGK ile en yüksek korelasyon Behçet grubunda da (r=-0.63, p<0.01) idiyopatik grubunda da (r=-0.63, p<0.01) BOS24 puanlama sistemiyle ile gözlendi.

Sonuç: Üveit skorlama sistemleri arka segment tutulumlu üveitlerde inflamasyonun değerlendirilmesinde kapsamlı, tekrarlanabilir ve uygulanabilir yöntemlerdir.

Anahtar Sözcükler: Üveit aktivasyon skoru, floresein anjiyografi skorlama sistemi, Behçet hastalığı oküler atak skoru 24, Behçet üveiti, idiyopatik üveit.

INTRODUCTION

Uveitis is infectious or noninfectious immune-mediated inflammation of the uvea and adjacent structures of the eye.[1] Behcet disease is a multisystem vasculitis of unknown origin, which is most prevalent in Mediterranean countries, Asia, the Middle East, and Japan.[2,3] Behcet disease constitutes 26% of all uveitis in Turkey.[4] Ocular involvement, which occurs mostly as a recurrent nongranulomatous panuveitis or posterior uveitis occurs in approximately 60–80% of patients with Behcet disease.[5,6]

Evaluation of ocular inflammatory activity in uveitis is mainly based on frequency of the inflammatory attacks,[7-9] anatomic location of uveitis (anterior uveitis, panuveitis, posterior uveitis),[8,9] severe ocular inflammation signs (hypopyon, involvement of retina, macula, or optic disc),[8] physicians' impression of severity (mild, moderate, severe),[10] or best corrected visual acuity (BCVA).[11] Such a descriptive data on uveitis do not allow precise monitoring of intraocular inflammation or comparison between clinical studies. Scoring systems may help to quantify the disease activity, treatment choices, prognosis, and comparison between studies more objectively. To understand a precise and easily used system for evaluation of disease activity in uveitis, different scoring systems emerged. Since 1991, uveitis activation score (UAS) was used.[12] Then, fluorescein angiography scoring system (FASS) was introduced in 2010.[13] Recently, the Behcet disease ocular attack score 24 (BOS24) came to the area.[3]

In this study, we investigated the feasibility of 3 different scoring systems in the evaluation of Behcet and idiopathic uveitis with posterior segment involvement.

MATERIALS AND METHODS

This retrospective study enrolled medical records of a total of 85 eyes of panuveitis or posterior uveitis, including 45 eyes which were associated with Behcet disease (Behcet group) and 40 eyes which were not associated with any systemic disease (idiopathic group). The study was performed at a tertiary referral center, and the ethics approval was approved by the Institutional Review Board. The research followed the tenets of the Declaration of Helsinki.

All patients underwent a complete ocular examination including BCVA, slit lamp biomicroscopy, dilated fundus examination, and fundus fluorescein angiography (FFA - Heidelberg Retina Angiograph system-2; Heidelberg Engineering, Heidelberg, Germany).

Posterior uveitis was diagnosed as the inflammation of the choroid or the retina. Panuveitis was diagnosed when the inflammation included the anterior chamber, vitreous, retina or choroid. All Behcet patients met the diagnostic criteria for Behcet disease after referral to rheumatologists.[14] Idiopathic uveitis was diagnosed after ruling out any associated systemic diseases. Exclusion criteria of the study were: 1. Patients with any systemic disease associated with uveitis except Behcet, 2. Patients with diabetic retinopathy, 3. Patients with severe media opacity that prevented to visualize posterior segment, 4. Patients with solely anterior uveitis.

The uveitic eyes were scored in the acute phase of the disease according to UAS,[12] FASS,[13] BOS24[3] as described below:

Uveitis Activation Score (0-29 points)

1. Macular edema (0-1)
2. Papillitis (0-3)
3. Neovascularization of the optic disc (NVD) (0-1)
4. Chorioretinal lesion (0-8, 1 point for each of eight section)
5. Retinal vasculitis (0-8, 1 point for each of eight section)
6. Neovascularization elsewhere (NVE) (0-8, 1 point for each of eight section)

Fluorescein Angiography Scoring System (0-40 points)

1. Optic disc hyperfluorescence (0-3)
2. Macular edema (0-4)
3. Retinal vascular staining/leakage (0-7)
4. Capillary leakage (0-10)
5. Retinal capillary nonperfusion (0-6)
6. NVD (0-2)
7. NVE (0-2)
8. Pinpoint leaks (0-2)
9. Retinal staining/pooling (0-4)

The Behcet Disease Ocular Attack Score 24 (0-24 points)

1. Cells in the anterior chamber (0-4)
2. Vitreous haze (0-4)
3. New inflammatory changes (exudates or hemorrhages) in the peripheral retina: 2 points for each quadrant (0-8)
4. New inflammatory changes (exudates or hemorrhages) in the posterior pole of retina (0-4)
5. New inflammatory changes (exudates or hemorrhages) in the fovea (0-2)
6. New inflammatory changes (papillar edema usually accompanied by hemorrhages, exudates, retinal edema surrounding the optic disc) in the optic disc (0-2)

STATISTICAL ANALYSES

The SPSS 16.0 software package was used in data analysis. For the descriptive statistics, discontinuous variables were shown as numbers and percentages (%); continuous variables that distributed normally were shown as mean ± standard deviation; continuous variables that did not distribute normally were shown as median (minimum-maximum). Normality of the data was evaluated with the Kolmogorov Smirnov test. A chi square test was used for categorical values. Student's t-test was used for continuous variables that distributed normally. The Mann-Whitney U-test was used for continuous variables that did not distribute normally. Spearman's correlation test was used for correlations. A P value less than 0.05 was considered significant.

RESULTS

Baseline characteristics (age, gender, BCVA, anatomic localization, duration of the disease, duration of follow-up, and bilaterality) are shown in Table 1. The age, gender, BCVA, anatomic localization, duration of the disease, and duration of follow-up were similar among the groups ($p > 0.05$). Bilaterality was significantly higher in Behcet group when compared with idiopathic group (91.1 % vs. 70.0 %, $p = 0.01$).

None of the uveitis scoring systems revealed significant difference between the groups, but the difference was greatest with BOS24 scoring system (Table 2).

BCVA and all 3 scoring systems (UAS, BOS24, and FASS) were moderately and significantly correlated in both Behcet group and idiopathic group (Table 3). The highest correlation of BCVA was observed with the BOS24 scoring system

Table 1. *Baseline characteristics (BCVA, age, gender, anatomic localization, duration of uveitis, duration of follow-up, and bilaterality) of the study groups are shown.*

Group	Behcet Group (n = 45)	Idiopathic Group (n = 40)	P Value
Age (years)	24 (min 20 - max 68)	22.5 (min 20 – max 51)	0.36*
Gender	42 male, 3 female	40 male	0.1**
BCVA (Snellen)	0.53 ± 0.34	0.48 ± 0.31	0.84***
Anatomic localization	32 panuveitis (71%), 13 posterior uveitis (29%)	32 panuveitis (80%), 8 posterior uveitis (20%)	0.34**
Duration of uveitis (months)	10 (min 1 – max 39)	6 (min 1 – max 168)	0.13*
Duration of follow-up (months)	3 (min 1 – max 26)	5 (min 1 – max 168)	0.84*
Bilaterality	41/45 (91.1%)	28/40 (70 %)	0.01**

BCVA = best corrected visual acuity, min-max = minimum-maximum
*Mann Whitney U test, **Chi square test, ***Student T test

Table 2. *Comparisons of Behcet group and idiyopathic group according to uveitis scoring systems UAS, BOS24, and FASS.*

	Behcet Group	Idiopathic Group	P
UAS (0-29)	9.91 ± 3.49	9.90 ± 4.54	0.48*
FASS (0-40)	18.27 ± 7.85	17.00 ± 7.86	0.95**
BOS24 (0-24)	13.69 ± 5.54	12.55 ± 4.49	0.19**

UAS = Uveitis Activation Score, FASS = Fluorescein Angiography Scoring System, BOS24 = The Behcet Disease Ocular Attack Score 24
*Mann Whitney U test, **Student T test

Table 3. *Correlations between BCVA and 3 uveitis scoring systems (UAS, FASS, and BOS24) in Behcet and idiopathic group.*

	BCVA - Behcet group	BCVA - Idiopathic group
UAS	p < 0.01, r = -0,529	p < 0.01, r = -0,615
FASS	p < 0.01, r = -0,571	p < 0.01, r = -0,471
BOS24	p < 0.01, r = -0,625	p < 0.01, r = -0,633

Spearman's correlation test was used.
BCVA = best corrected visual acuity, UAS = Uveitis Activation Score, FASS = Fluorescein Angiography Scoring System, BOS24 = The Behcet Disease Ocular Attack Score 24.

in both Behcet group (r=-0.63, p<0.01) and idiopathic group (r=-0.63, p<0.01).

DISCUSSION

In the present study, descriptive results were similar with the previous literature in which bilaterality was significantly higher in Behcet group (91.1%) when compared with idiopathic group (70.0%), 71% of the Behcet uveitis and 80% of the idiopathic uveitis presented as panuveitis, and the remainder were posterior uveitis. Kozakoglu et al.[15] reported that, in Turkey, the most common type of uveitis was anterior uveitis (52.5%) followed by panuveitis (28.1%), posterior uveitis (12.7%) and intermediate uveitis (6.7%). Etiological classification was established in 56.8% of these patients, in which the most common etiology was Behcet disease (32.1%). Ozdal et al.[16] reported that Behcet uveitis was bilateral in 87% of cases, fundus changes were observed in 82.9% and vitritis was present in 93% of eyes.

Grading systems are comprehensive and reproducible ways for measurement of inflammation in uveitis because assessing solely BCVA is insufficient and does not give enough information for the severity of uveitis. The uveitis scoring systems may be useful for diagnosing uveitis, assessing the inflammation, monitoring progression, and following response to treatment. Standardizing the clinical data in uveitis have been recommended by different uveitis scoring systems; UAS, FASS, and BOS24.[3,12,13] Our study results revealed that none of the uveitis scoring systems were evidently different in scoring Behcet and idiopathic uveitis when the posterior segment was involved, but the difference was greatest with BOS24 between the groups. The highest correlation of BCVA with any scoring system was observed with BOS24 in both Behcet group and idiopathic group.

BOS24 has subcriterias about the vitreus haze and anterior chamber cells which UAS and FASS do not have and these subcriterias are mandatory in assessing panuveitis apart from posterior uveitis. BOS24 is scored by only new emerging inflammatory signs. Flaring in the anterior chamber, chronic vitreous opacity and macular edema are not involved in BOS24. On the other hand, UAS and FASS scores macular edema. Retinal inflammatory signs, especially the posterior pole of the retina and fovea has high scores in BOS24 and posterior segment involvement, which can lead to blindness with recurrent inflammatory attacks, has a very important prognostic value in Behcet uveitis.[16] There are some limitations of the BOS24. It is difficult to score retinal lesions if ocular fundus lesions are invisible due to vitreous opacity, vitreous hemorrhages, or a hard cataract. In such situations, scoring systems based on FFA, FASS, remain a better choice in evaluation of ocular disease activities in uveitis.

Tutkun et al.[13] introduced FASS in order to standardize the interpretation of angiographic findings in uveitis. FFA is useful in diagnosis of uveitic entities for assessing retinal inflammation, complications such as macular edema, monitoring disease progression, and following response to therapy.[13,17] Diffuse vascular leakage and optic disc hyperfluorescence on FFA is the hallmark of retinal vasculitis and is one of the most frequent findings in patients with Behcet uveitis.[16,18-19] Because of the limitations of FFA in imaging of the choroid, Tutkun et al.[13] has also recommended indocyanine green angiography (ICGA) as a useful tool in studying choroidal inflammatory processes.

There are some limitations about this study. First, this study was analyzed retrospectively which restricts the study to previously recorded information. Second, ICGA was not performed. Our study data did not include ICGA because in our study cohort, we used ICGA only when needed, the number of ICGA data was not enough to make comparisons.

CONCLUSION

In summary, uveitis scoring systems are useful for diagnosing, monitoring, following uveitis in order to standardize the clinical findings when the posterior segment of the eye is involved. Behcet and idiopathic uveitis with posterior segment involvement can be assessed with any of the UAS, FASS or BOS24 scoring systems. Our findings demonstrate that the BOS24 system seems to better among the others. On the other hand FFA is the gold standard of evaluating the posterior segment in uveitis and if ocular fundus lesions are invisible, FASS is a better way in the evaluation.

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