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Optical Coherence Tomography Changes in Patients with Sickle Cell Disease
Running title: Imaging in Sickle Cell Disease

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Abstract:

Aim: Sickle cell disease can cause retinopathy, resulting in vision loss and vitreous hemorrhages in patients. The purpose of study is to compare the optical coherence tomography (OCT) changes of the control group and patients with sickle cell disease.
Methods: This study was conducted between December 2020 and March 2021 in Department of Ophthalmology, Faculty of Medicine, Mersin University, Mersin, Turkey. This study included 48 eyes of 24 sickle cell disease patients and 40 eyes of 20 healthy controls. The changes seen were analyzed with a scale divided into 9 sectors according to the Early Treatment Diabetic Retinopathy Study (ETDRS) protocol.
Results: Of the 24 patients, 7 (29,2%) were male and 17 (70,8%) were female. Four (20%) of the 20 patients in the control group were male and 16 (% 80) were female. Changes in temporal inner, superior outer and macular volume were found to be significantly lower in the control group. ($p = 0,011$, $p < 0,001$, $p = 0,010$, respectively)
Conclusion: Consequently, sickle cell disease can cause blindness that may be accompanied by retinopathy. Those with this disease need careful examination of the retina and use of imaging methods.
Keywords: optical coherence tomography; retinopathy; sickle cell disease

Introduction

Sickle cell disease is a disorder that affects hemoglobin in erythrocytes. People with this disease have atypical hemoglobin molecules called hemoglobin S, which can distort red blood cells into a sickle or crescent shape. Sickle cell disease affects millions of people worldwide. It is most common among people whose ancestors come from Africa, Mediterranean countries such as Greece and Turkey[1].
Various pathologies are seen in the anterior and posterior segments of the eye due to sickle cell disease. One of the reasons for these complications is the changes caused by sickled cells in the microvascular structures[2].The aim of this study is to compare retinal thickness in different regions between sickle cell patients and healthy controls.

Material and Methods

The protocol of this study was approved by Mersin University Clinical Research Ethics Committee. Written informed consent was obtained from all participants included in this study. In addition, the Declaration of Helsinki was adhered to throughout this study. Statistical analysis of study data was done with SPSS 23.0 package program. Categorical variables were summarized as numbers and percentages, and continuous variables as mean ±standard deviation. The control of the normal distribution of continuous variables was checked with the Shapiro Wilk test. Student's t test was used to compare the means of two independent groups. Relationships between categorical variables were investigated by chi-square analysis. Statistical significance level was taken as $p < 0.05$ for all comparisons.

The optical coherence tomography (OCT) examinations (OCT Spectralis, Heidelberg Engineering GmbH, United Kingdom) performed on 48 eyes of 24 patients with sickle cell diseases who admitted to the Department of Ophthalmology, Faculty of Medicine, Mersin University between December 2020 and March 2021 were retrospectively

screened. None of the patients had proliferative and/or non-proliferative retinopathy. The changes seen were analyzed with a scale divided into 9 sectors according to the ETDRS protocol and compared with a control group (40 eyes of 20 patients) of completely healthy individuals with similar age and gender distribution. The macula was divided into three concentric circles with diameters of 1 mm, 3 mm, and 6 mm, named fovea, pericentral ring, and peripheral ring, respectively. Also, according to the ETDRS, the pericentral and peripheral rings were equally divided into four regions: superior, nasal, inferior, and temporal. In total, 9 sectors were involved in the macular area.

Results

Of the 24 patients, 7 (29,2%) were male and 17 (70,8%) were female. The average age was 38,7 ±10,1 years (min - max: 23 - 58 years). Four (20%) of the 20 patients in the control group were male and 16 (% 80) were female. The average age was 37,5 ±8,5 years (min - max: 25 - 60 years). The OCT measurements for both groups were evaluated using a grid divided into 9 sectors (nasal inner, nasal outer, temporal inner, temporal outer, superior inner, superior outer, inferior inner, inferior outer and fovea) according to the ETDRS protocol. Full thickness measurements of the patients: nasal inner 334,25 ±24,30 µm, nasal outer 313,5 ±19,01 µm, temporal inner 304,75 ±28,85 µm, temporal outer 275,66 ± 16,51 µm, superior inner 327,58 ±27,53 µm, superior outer 268,25 ±11,85 µm, inferior inner 322,41 ±19,65 µm, inferior outer 288,6 ±17,67 µm and fovea was 274,08 ±41,27 µm. Macular volume was 8,32 ±0,54 mm³. The mean macular volume in the control group was 8,61 ±0,49 mm³. Changes in temporal inner, superior outer and macular volume were found to be significantly lower in the control group ($p = 0,011$, $p < 0,001$, $p = 0,010$, respectively) (Table 1).

	Sickle Cell	Control	p
	mean±SD (µm)	mean±SD (µm)	
nasal inner	334,25 ± 24,30	343,58 ± 21,20	0,071
nasal outer	313,50 ± 19,01	317,26 ± 12,88	0,286
temporal inner	304,75 ± 28,85	321,94 ± 33,05	**0,011**
temporal outer	275,66 ± 16,51	280,65 ± 14,83	0,139
superior inner	327,58 ± 27,53	328,71 ± 25,03	0,842
superior outer	268,25 ± 11,85	323,67 ± 24,10	**< 0,001**
inferior inner	322,41 ± 19,65	330,29 ± 36,62	0,202
inferior outer	288,60 ± 17,67	292,74 ± 21,33	0,322
fovea	274,08 ± 41,27	290,68 ± 39,17	0,058

Table 1: Optical coherence tomography measurements of participants

Discussion

In this study, we demonstrated that there is a difference in thickness of retinal segments visualized by optical coherence tomography between patients with sickle cell disease and healthy controls.

In a study published by Grego et al. in 2020, found that optical coherence tomography can detect early signs of retinopathy and maculopathy in children[3]. In our study, even in patients without advanced disease, different results were found compared to healthy controls. Therefore, sickle cell disease changes retinal vascularity and retinal thickness regardless of its stage.

In a similar study, the importance of optical coherence tomography was emphasized in the detection and numerical follow-up of temporal thinning in the pediatric patient group, especially in subclinical patients[4]. In our study, the temporal inner, superior outer segments and macular volume were significantly lower. This shows that the damage to the retinal vessels affects the temporal retina more prominently.

In a study conducted in our country, patients with both sickle cell disease and beta-thalassemia were investigated and early structural changes were shown in patients who did not develop proliferative or non-proliferative retinopathy for either disease[5].

In a study in the literature, determined that there were quantitative changes in OCT imaging in patients with asymptomatic sickle cell disease, and macular thinning was more prominent especially in patients with proliferative retinopathy[6]. In our study, in addition to retinal segment thinning, macular volume was found to be decreased even if the patients did not have proliferative retinopathy. This result shows us that retinal changes will show earlier in OCT imaging, even if retinopathy is not at a level that cannot be recognized clinically.

Lim and Cao, performed retinal thickness analysis on sickle cell patients and healthy controls in a study. In this study, the macula in sickle cell eyes was thinner than in control eyes[7]. In our study, although the central foveal thickness was not different from the control group patients (274,08 vs 290,68 µm, respectively), the macular volume was significantly lower. This is thought to be due to the relatively small number of patients.

The limitations of the study were that the vitreous hemorrhage seen in patients with advanced disease did not allow imaging studies, so imaging was performed in milder patients. In addition, larger patient populations and multicenter studies are needed for more specific proof of the changes that occur.

In conclusion, sickle cell disease can cause blindness, which may be accompanied by retinopathy. The posterior segment examinations of patients with this disease should be carefully examined and imaging methods should be used.

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