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Macular Thickness in Iraqi Patients with Sickle Cell Anemia

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Abstract

Introduction: Sickle cell disease (SCD) is the most common genetic disease in the world. Several authors have described morphological changes from the macula to optical coherence tomography-spectral domain (OCT SD) including the thinning of the predominant inner layers in the temporal area in SCD.

Methodology: This is an analytic study conducted mainly at the specialized ophthalmology department from March 2019 to July 2023. Any SCD patient who is 20 years of age or older were involved and sickle cell patients without retinal pathology (myopia, diabetic retinopathy and vitreo-retinal interface pathology). A full ophthalmologic examination was performed. The visual acuity was taken on the Snellen scale. The near vision acuity on the Parinaud scale. Refraction was done with the Nidek auto-refractometer. Measurement of intraocular pressure by the tonometer. The slit lamp examination of the anterior segment was done. The fundus was made using the Goldman 3-Mirror glass. Retinal thickness measurements were made using a Nidek RS-3000 Advance tomograph. The thickness was measured by the macula map early treatment diabetic retinopathy study [(ETDRS) (macula map)] made of 9 fields composed of 3 concentric circles of 1 mm, 3 mm, and 6 mm in diameter. The OCT test was interpretable when the signal strength index (SSI) was greater than or equal to 7/10.

Results: Here, the median age was 32 years. There is a predominance of female vs male sex ratio = 0.56. 84% of eyes had retinal lesions suggestive of non-proliferative retinopathy in the retina. The solar black spots were the most found retinal lesions (68%). Lesions were more localized temporally. In the OCT measurement, 60% of the eyes showed a decreased retinal thickness SD with 78% concerning the temporal retina. There was no decrease in visual acuity in our patients who had a decrease in retinal thickness at OCT SD.

Conclusion: There is a thinning of the retinal layers in sickle cell patients in the temporal region of the macula. Patients with retinal thinning are asymptomatic with preserved visual acuity.

Keywords: Sickle cells patients, Macular thickness, Optical coherence tomography, Retina, Retinopathy

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Introduction

Sickle cell retinopathy (SCRP) occurs in more than half of patients [1, 2]. SCD is the commonest genetic disorder in the world [1]. Several authors have described morphological changes from the macula to OCT SD including the predominant inner layers thinning in the temporal area. These modifications were found in asymptomatic cases [3].

The WHO estimates that about 7% of the world's population are carriers of hemoglobinopathies and about 300,000 to 400,000 babies are born every year with a severe form of hemoglobinopathy. Hemoglobinopathies are genetic disorders characterized by either abnormal hemoglobin, as in SCD, or insufficient production of hemoglobin chains, as in thalassemia. Proliferative SCRP is the most serious vision-threatening complication of SCD and is reportedly seen in 0.5 percent of patients with HbSS disease, the severe variant of SCD and about 2.5 percent of patients with HbSC disease, a less severe variant of SCD. The frequency of ocular involvement in patients with beta-thalassemia ranges from 41.3 to 85 percent, according to studies. This activity describes the causes, pathophysiology, and presentation of hemoglobinopathy-associated retinopathy and highlights the role of the inter-professional team in the care of affected patients. The ocular manifestations of SCD were described by Cook in 1930 when he noticed retinal hemorrhages in a patient who died of subarachnoid hemorrhage.

SCRP, especially proliferative sickle retinopathy, is considered the commonest cause of loss of vision in SCD. The best method to prevent the development of devastating complications of proliferative sickle retinopathy is regular dilated peripheral retinal examinations. Patients should be educated regarding the importance of regular ophthalmological examinations. Patients should also be instructed to consult an ophthalmologist if they notice any changes in vision. Close follow-up and annual retinal examinations are also required in patients with thalassemia to prevent and detect any complications before permanent damage occurs. Educating the patient will go a long way in this direction [4].

It seems therefore appropriate to assess the central thickness of the retina in SCD patients and to determine links with the clinical parameters.

Method

This is an analytic study conducted mainly at the specialized ophthalmology department from March 2019 to July 2023. Any SCD patient who is 20 years of age or older were involved and sickle cell

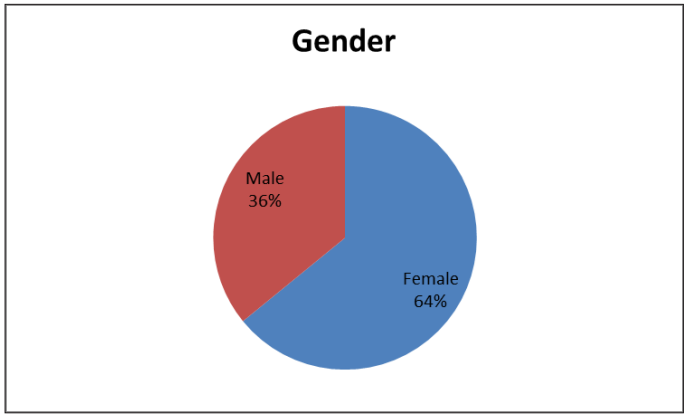

Figure 1: Gender distribution of the study.

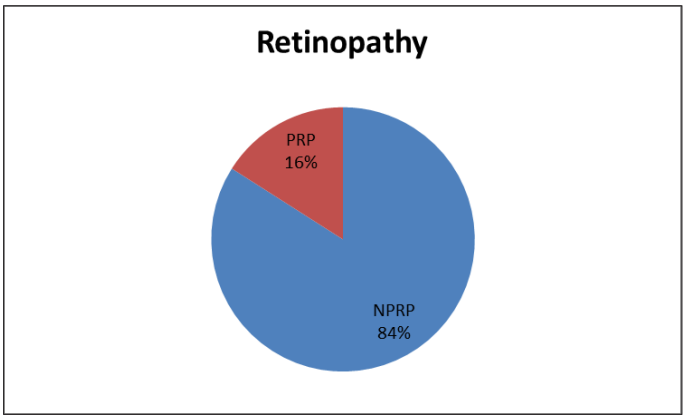

Figure 2: Retinopathy distribution of the study.

patients without retinal pathology (myopia, diabetic retinopathy and vitreo-retinal interface pathology). A full ophthalmologic examination was performed.

- The visual acuity was taken on the Snellen scale.
- The near vision acuity on the Parinaud scale.
- Refraction was done with the Nidek auto-refractometer.
- Measurement of intraocular pressure by the tonometer.
- The slit lamp examination of the anterior segment was done.
- The fundus was made using the Goldman 3-mirror glass.
- Retinal thickness measurements were made using a Nidek RS-3000 advance tomograph.
- The thickness was measured by the macula map ETDRS (Macula map) made of 9 fields composed of 3 concentric circles of 1 mm, 3 mm, and 6 mm in diameter.
- The OCT test was interpretable when the SSI was greater than or equal to 7/10.

Biological data from patient records were hemoglobin level (the patients were considered to have severe anemia for a hemoglobin level < 7 g/l, a moderate anemia between 7 and 10 g/dl, a mild anemia at more than 10 g/l according to WHO). The percentage of hemoglobin S after quantification and fragmentation by electrophoresis was noted.

The data were entered into and analyzed by SPSS ver. 24 software. To establish the correlations between the data, we used CI around the OR to estimate the association between the data and the tests. Pearson and Fischer are exact to determine the significance for a $p < 0.05$.

Results

Here, the median age was 32 years. There is a predominance of female vs. male sex ratio = 0.56 (Figure 1). 84% of eyes had retinal lesions suggestive of non-proliferative retinopathy in the retina (Figure 2). The solar black spots were the most found retinal lesions (68%) (Figure 3). Lesions were more localized temporally. In the OCT measurement, 60% of the eyes showed a decreased retinal thickness SD (Figure 4) with 78% concerning the temporal retina (Figure 5). There was no decrease in visual acuity in our patients who had a decrease in retinal thickness at OCT SD.

Discussion

During the OCT measurement, 60% of eyes presented a reduced SD retina thickness with 78% concerning the temporal retina. This predominant localization of the reduction in the thickness of the retina was found by Mathew et al. [5] reported 107 sub-Saharan African and Afro-caribian patients whose age was between 18 and 74 years old. This thinning is more found in the temporal macular region in African and Caribbean patients [5].

Minvielle et al. [6] in a series of cases of 9 sickle cell patients aged 19 to 54 years noted during the examination by the OCT-A the existence of more marked microvascular anomalies of the temporal peri foveolar region concerning the superficial and deep capillary plexus.

Indeed, OCT SD and OCT-A are the two recent non-invasive retinal imaging tools used that can demonstrate subclinical macular damage secondary to ischemia [7].

All of our patients had a hemoglobin S percentage greater than 80%. We did not find a correlation between the presence of retinal lesions of non-proliferative SCRP, and the study done in 2015 notes that a discreet thinning of the macular proliferative SCRP.

We did not note a decrease in visual acuity in our patients who presented a reduction in the thickness of the retina at OCT SD. Indeed, the etiology of retinal thinning found at OCT SD is not fully understood and the functional consequences have not yet been elucidated [8].

Conclusion

There is a thinning of the retinal layers in sickle cell patients in the temporal region of the macula. Patients with retinal thinning are asymptomatic with preserved visual acuity.

Acknowledgements

None.

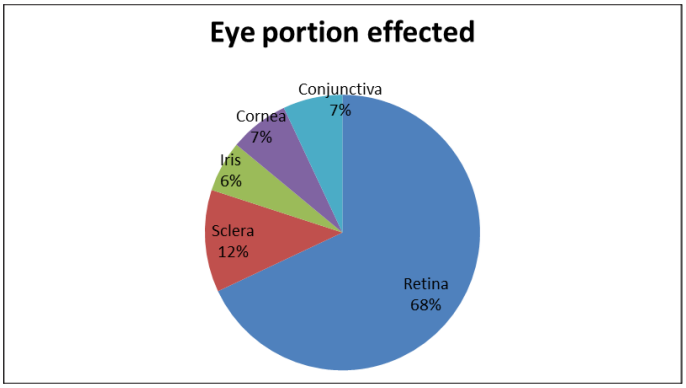

Figure 3: Eye portion effected by SCD distribution of the study.

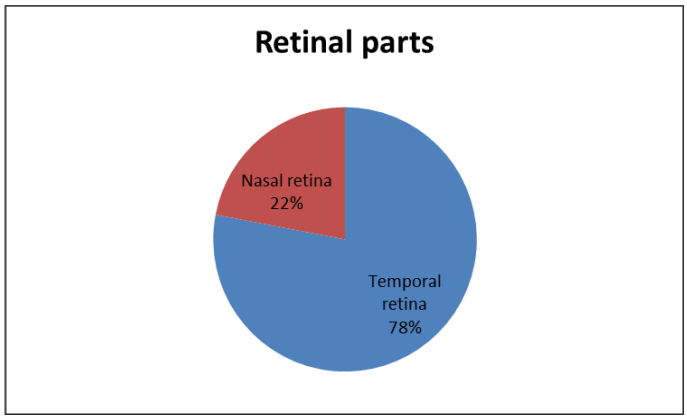

Figure 4: Retinal parts effected by SCD distribution of the study.

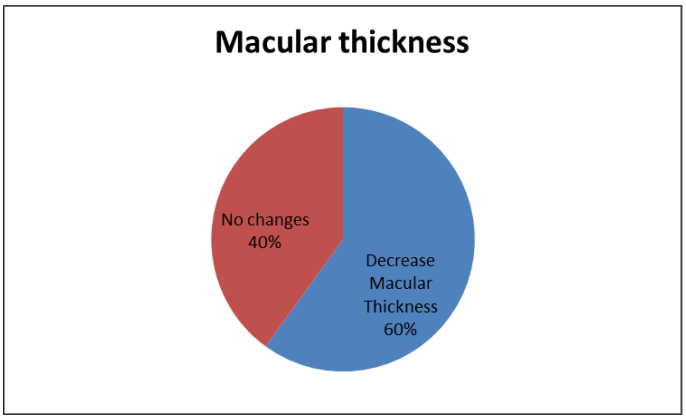

Figure 5: Macular thickness of the study.

Conflict of Interest

None.

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