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Ocular Complications of Sickle Cell Disease in Conakry

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Abstract

Introduction Sickle cell retinopathy is the retinal manifestation of sickle cell disease which is due to the alteration of the parameters that influence blood flow in the retinal capillaries. These ocular complications are multiple and affect practically all structures of the eye-ball. The aim was to identify the main ophthalmological complications in patients with sickle cell disease. **Materials and Methods:** This was a prospective descriptive study lasting six (6) months from 25 July to 25 December 2022. The study covered all sickle cell patients who consulted the S.O.S sickle cell center in Nongo and who were referred to the ophthalmology department of the army medical-surgical center during the study period. **Results:** 107 patients (55 males, 52 females) were selected with a ratio of 1.05. Their age ranged from 2 to 45 years. The vast majority of our patients were referred (96.26%). Non-proliferative sickle cell retinopathy was the most common (56.25%). **Conclusion:** Ocular manifestations of sickle cell disease, including sickle cell retinopathy, represent serious complications that can impair vision. Early detection and management are essential to prevent these complications and preserve the visual health of sickle cell patients.

Keywords

Sickle Cell Retinopathy, Guinea

1. Introduction

Sickle cell retinopathy is the retinal manifestation of sickle cell disease, which is due to alterations in the parameters that influence blood flow in retinal capillaries [1].

Sickle cell disease is the most common hereditary haemoglobinopathy S in the world, characterised by multi-systemic complications, among which the eye is not

spared [2].

Its prevalence varies from region to region and it is responsible for a number of complications, including retinal ocular complications that can lead to blindness [3] [4].

Certain ethnic groups (West Indies, Black Africa) are particularly at risk of these conditions. This is a general disease dominated by haemolytic anaemia associated with a variety of clinical manifestations. Retinopathy is common and sometimes severe, more so in heterozygous SC forms than in homozygous SS forms. Retinal vascular occlusions, resulting from the falciformation of red blood cells, are the main ocular complications of the disease [5].

Ocular complications are multiple and affect virtually all structures of the eye-ball, hence the importance of regular and prolonged ophthalmological surveillance of sickle cell patients. [6]

Sickle cell retinopathy selectively affects the periphery of the retina, but the posterior pole may also be affected. There are classically two types of retinopathy: non-proliferative and proliferative. The latter is the most serious form of ocular damage. Fundus examination is often useful for assessing the impact of this condition. This is an important step, enabling macroscopic analysis of the retina in particular. Treatment of proliferative sickle cell retinopathy is based on peripheral confluent Argon laser photocoagulation of areas affected by neovessels, or by surgical methods. The latter is indicated in the most serious complications, requiring special precautions in these patients [7].

In Cameroon in 2018, a study of sickle cell retinopathy in patients with sickle cell disease by Bilong Y *et al.* showed that 27% had non-proliferative sickle cell retinopathy and 35% had proliferative sickle cell retinopathy [8].

In Egypt in 2021, a study of retinopathy in patients with sickle cell disease by Tamer Hassan *et al.* showed that 25% had proliferative retinopathy and 31% had tortuous retinal veins [9].

Thus, the scarcity of previous studies and the high number of sickle cell patients in Guinea, motivated our choice to work on this topic.

General objective:

To study the main ophthalmological complications in sickle cell patients referred to the Nongo S.O.S. sickle cell centre.

Specific objectives:

Describe the sociodemographic profile of sickle cell disease patients with ophthalmological complications.

Identify the clinical characteristics of ophthalmological complications.

Describe the management of ophthalmological complications of sickle cell disease.

2. Methods

The ophthalmology department of the Conakry military hospital and the S.O.S. sickle cell center in Nongo provided the setting for this study.

Type and duration of study:

This was a prospective descriptive study lasting six (6) months from 25 June to 25 December 2022.

2.1. Study Population

The study included all sickle cell subjects referred from the Nongo S.O.S sickle cell center and then to the ophthalmology department of the Conakry military hospital during the study period. A complete ophthalmological examination was performed. We submitted our protocal to the local ethic and all the patient are informed and signed the approuvement before the follow up.

2.2. Selection Criteria

- **Inclusion:**

We had included in our study all sickle cell patients who had been received during the study period at the S.O.S sickle cell center in Nongo.

- **Exclusion:**

We had excluded from our study any patient who had previously undergone laser treatment (PPR: Pan photo retinian), those with a cataract that prevents retinal examination as well as comorbidities that can lead to retinopathies such as diabetes, hypertension or others.

3. Results

We conducted a prospective descriptive study lasting six (6) months from 25 June to 25 December 2022.

The study included 107 patients aged between 2 and 45 years with an ophthalmological complication. (See Tables 1-3 and Figures 1-5)

Table 1. Breakdown of 107 cases of ophthalmological complications in sickle cell disease patients according to age.

Age range (years)	Workforce	(%)
0 - 9	42	39.2
10 - 19	36	33.6
20 - 29	22	20.5
≥30	7	6.5
Total	107	100

Average age = 14.1682 ± 9.4195, Extreme 2 to 45 years.

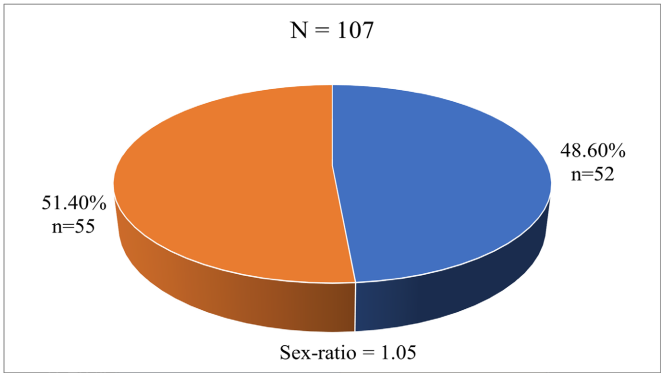

Figure 1. Repartition of 107 sujet of sickle cell disease by sex.

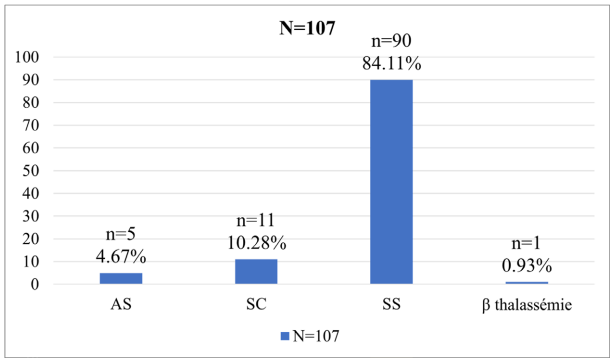

Figure 2. Repartition of 107 case ophtalmologic complication in génotypique form sickle cell disease.

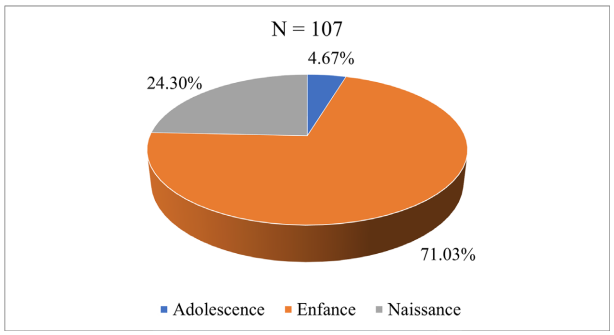

Figure 3. Repartition of 107 case ophtalmologic complication by date screening sickle cell disease.

Table 2. Repartition of 214 eyes complications ophtalmologic by acute visual.

AVSC	Number	Pourcentage (%)
1/10 à 5/10	6	2.8
6/10 à 9/10	42	19.63
10/10	164	76.63
PL+	1	0.47
PPL	1	0.47
Total	**214**	**100**

NB: **PL+** = light perception (if patient percept the light), **PPL** = not light perception.

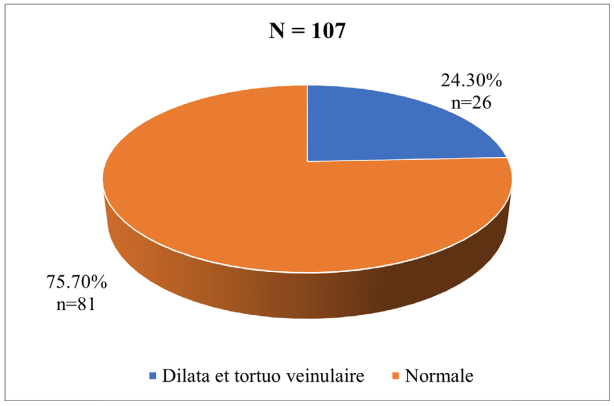

Figure 4. Repartition of 107 cases by fundus.

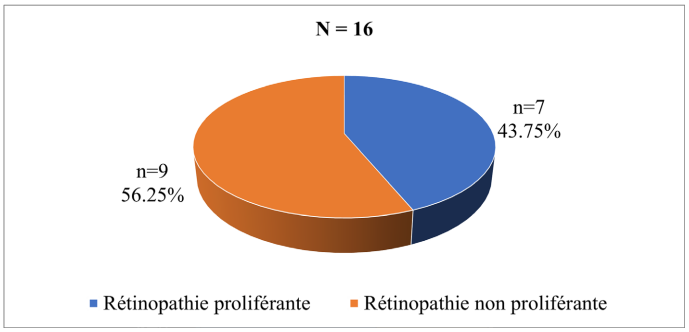

Figure 5. Répartition of 16 cases by retinopathy in sickle cell patient with ophtalmologic complication.

Table 3. Répartition of patient by stade of retinopathy sickle cell disease.

Variable	Number	Pourcentage
Proliferant Rétinopathy	7	47.75
Stade I	2	12.5
Stade II	2	12.5
Stade III	1	6.25
Stade IV	1	6.25
Stade V	1	6.25
Non proliferant retinopathy	9	56.25

4. Discussion

Sickle cell anaemia is a haemoglobinopathy with the highest frequency and severity of retinal damage. The mean age was 14.16 ± 9.41. Our results is different from those of Kassoum, who reported a predominance of patients between 21 and 30 years, with a proportion of 33.3% in this study of sickle cell retinopathy in SC [10]. This difference can be explained by the fact that our study covered all genotypic forms of sickle cell disease. It should also be noted that according to the literature, proliferative retinopathy in most cases only observed after the age of 10 years in Sickle Cell composite heterozygotes.

The risk seems to be the same for men and women, although a higher frequency was observed in our study. Our results are similar to those of Inoussa *et al* who, in their study of the knowledge, attitudes and practices of sickle cell patients in relation to sickle cell (SC) retinopathy, reported that the predominant sex was male, with a proportion of 54% and a sex ratio of 1.17 [11]. This male predominance can not be explained as the transmission of sickle cell disease is in no linked to sex.

We have recorded several genotypic forms of sickle cell disease and the most common form is the homozygous SS form with a rate of 84.11%, followed by the composite heterozygous SC form with a rate of 10.28%. This could be explained by the frequency of the SS homozygous form in our context and by the fact that vaso-occlusive crises are more common in these patient.

Our study revealed that the majority of our patient had a visual acuity of 10/10.

Our results are similar to those of **Ouattara**, who reported in his study on the evaluation of the quality of ophthalmological follow-up of patients with sickle cell disease that patients with a bilateral visual acuity of 10/10 were the most frequent, with a rate of 61.8%.

During our study, a fundus examination was performed in all our SC patients. Our results are inferior to those of **Kassoum**, who reported that 97% of his patients had a normal anterior segment, compared with 3% with an abnormal fundus. He also reported that 94% of his patients had a transparent vitreous compared with 4.5% with intravitreal haemorrhage and 1.5% with posterior vitreous detachment.

According to the type of retinopathy, our results are similar to those of **Kassoum** who, in his study of sickle cell retinopathy in SC subjects, reported a proportion of 63.6% of proliferative retinopathy compared with 36.4% of non-proliferative retinopathy. These results are similar to those of **Dohvoma *et al.*** who found in their study of the prevalence of sickle cell retinopathy in SS homozygotes a proportion of 21.6% of non-proliferative retinopathy and 0.6% of proliferative retinopathy. This could be explained by the fact that our study covered all forms of sickle cell disease and, according to the literature, heterozygous composite SC subjects develop more proliferative retinopathy.

Among our patients who had developed proliferative retinopathy, the majority were at stage I and II with the same proportion. Our results are similar to those of **OUATTARA** who reported in his study on the evaluation of the quality of ophtalmological follow-up of sickle cell disease patient a proportion of 22.2% of stage II proliferative retinopathy [12]. In our study, some have no insurrancy to the extra examination for more investigation, in the future study in perspective we want more material like angiography, the tomography by coherence optic for more investivation of sickel cell retinopathy.

5. Conclusion

The ocular manifestations of sickle cell disease, particularly sickle cell retinopathy, represent serious complications that can impair vision. Early detection and management are essential to prevent these complications and preserve the visual health of patients with sickle cell disease.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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