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Study of the Retinal Impact of Sickel Cell Disease in Homozygous SS Patients Aged over 15 Years in a Hospital Environment in Kinshasa

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Abstract

Context: Sickle cell disease is a widespread disease throughout the world with 2.3% of the world's population. The DRC being the 2nd country in Africa and the 3rd in the world. The objective was to study the retinal lesions found in SS sickle cell patients, which is the form found in the DRC.

Methods: A descriptive cross-sectional multicenter study was conducted from December 2021 to March 2022 at the University Clinics of Kinshasa. The study looked for retinal lesions in homozygous sickle cell patients over the age of 15. A complete ophthalmological examination was carried out, including a direct fundus examination after pupil dilation. Optical coherence tomography was performed when needed.

Results: 55 patients (110 eyes) were seen during the study period. The mean age of these patients was 20.6 ± 7.1 years. The female gender predominated with 50.9% with a sex ratio of 0.96. Blurred vision was the most common eye complaint at 23.6%. Visual impairment and blindness were found respectively in 2.7% and 0.9%. Retinal lesions were found in 29 eyes or 26.3%. These lesions were: Vascular tortuosity in 17.3%; peripheral retinal pallor in 3.6%; Solar black spots in 2.7%; salmon hemorrhage, RCAO, peripheral arteriolar occlusion, proliferative sickle cell retinopathy in one eye each.

Conclusion: Retinal lesions of sickle cell disease are quite common and should be sought regularly in patients with sickle cell disease.

Keywords: Homozygous SS; Sickle cell retinopathy

Introuduction

Sickle cell anemia or sickle cell anemia is a very competent hereditary disease throughout the world and particularly in the Democratic Republic of Congo (DRC) [1]. The disease is recognized as a major public health problem worldwide as approximately 2.3% of the world's population is affected. Each year, about 500,000 children with sickle cell disease are born in the world, more than half of them (300,000) in Africa. According to the WHO, the mortality of these children is very high before the age of 5: 230,000 children in sub-Saharan Africa, 2,600 in North America and 1,300 in France [2]. It is most performed in the African region where the proportion of the population carrying the sickle cell gene is estimated at 10 to 40%. The prevalence rate of sickle cell disease is estimated at 2% [3]. The DRC is the 3rd country in the world after India and Nigeria and the 2nd country in Africa to be affected by the disease [3]. And in this same country, epidemiological data showed

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that with an estimated population of 90,000,000; 2 to 3% of newborns are homozygous for hemoglobin S; while in the adult population, the carriage of the trait stands at around 25-30%. This figure is significant from an epidemiological point of view, yet the disease remains little known, resulting in high mortality in a country with limited resources. The Center for Mixed Medicine and SS Anemia (CMMASS) emphasizes that the treatment of a patient requires around US$1,000 per year. Despite significant progress in reducing the mortality of children under 5 and pregnant women, the situation of sickle cell disease remains very worrying in our country because of poor care and ignorance of both the population than nursing staff [3,4]. During this disease, chronic hemolytic anemia, acute complications by vasoocclusion of microvessels, chronic visceral complications of ischemic origin can also affect all the organs, including the eye, of which the retinopathy is one. Retinopathy is a complication of sickle cell disease; because the retina is one of the potentially damaged sites in sickle cell disease [5]. The analysis of the fundus, refined by fluorescein angiography, makes it possible to determine several types of lesions. The most serious is the proliferative form of retinopathy corresponding to stages 3,4 and 5 of the Goldberg classification. The latter concerns 54.6% of people with a heterozygous form (SC) and 18.1% of homozygous patients (SS) [2,5,6]. Clinical retinal involvement is asymptomatic in the early stages, hence the need for preventive ophthalmological management. The severity of retinopathy is independent of the severity of general manifestations. There are factors that are known to be protective, such as high fetal hemoglobin. Visual alterations and functional complaints are reported in only 10 to 20% of patients with sickle cell disease, and this when they become at the stage of complications such as intravitreal hemorrhage or retinal detachment [2,7]. The peak prevalence of onset of proliferative retinopathy would be around 25-39 years for the SS forms, described at the age of 8 years for the SC forms and 15-24 years for the others. In addition, the authors found the proliferative forms, evaluated at 8% of the SC forms and 0.6% of the SS forms; hence the interest in organizing screening from the age of 9 for the SC forms and from 13 years for the SS forms [2,7]. Other teams have found lesions in 24% of cases including 5.6% of proliferative forms in patients aged 5 to 36 years and recommend a systematic examination from the age of 10 years [2,8]. We want through this study, to determine the retinal lesions in SS sickle cell patients in our environment in order to consider management, and encourage ophthalmological screening of SS sickle cell patients. Hence the importance of a systematic ophthalmological examination for early detection of retinal lesions.

Methods:

The present study is a multicenter descriptive cross-sectional study which took place from December 2021 to March 2022 at the University Clinics of Kinshasa with patients from the Mixed Center for Anemia SS Mabanga (CMASS) and hospital centers of the BDOM (Bureau Diocésain des Œuvres médical) : C.H. Lisungi ; C.H. René de Haes ; C.H. Saint Joseph de Tyrain ; C.S Saint Clément. This study concerned SS homozygous sickle cell patients over 15 years of age who had consulted the medical training courses mentioned above. Our sampling was exhaustive which took into account any sickle cell patient who had given his free consent to participate in the study and who had carried out all the ophtalmology examinations meeting our parameters of interest. A total of 55 sickle cell patients met the inclusion criteria and constituted our sample size. We personally contacted the doctor in charge of the CMASS sickle cell care structure and the nurse in charge of sickle cell patients in the BDOM structures who agreed to send us the patients to the CUK for ophthalmological examinations. The data was collected using pre-established sheet. The anonymity of our results was guaranteed.

Results:

Sociodemographic parameters

Table 1: Distribution of patients according to socio-demographic data

Demographic data	Number (n=55)	%
§ Age		
- 15 – 24	44	80,0
- 25 – 34	8	14,5
- 35 – 44	2	3,6
- 45 – 54	1	1,8
§ Sex		
- Male	27	49,1
- Femelle	28	50,9
Total	55	100,0

The average age of the patients was 20.6 ± 7.1 years with extremes of 15 and 48 years. The majority of patients were between 15 and 24 years old. The female sex represented 50.9% with a sex ratio of 0.97.

Data anamnestic

Taking hydroxyurea was recorded in 7.3% of patients.

Ophthalmological parameters

Visual acuity

Table 2: Distribution of patients according to visual acuity

AV	Raw		Corricted	
	Number	%	Number	%
Normal	98	89,1	108	98,2
Subnormal	8	7,3	-	-
Visual impairment	3	2,7	1	0,9
Partial blindness	-	-	-	-
Total blindness	1	0,9	1	0,9
Total	110	100,0	110	100

1 eye showed total blindness and 3 eyes showed visual impairment. After correction, low vision and blindness were found in 1 eye each, i.e. 1.8%.

AS lesions

Iridocrystaline synechiae and lens opacity were AS lesions found in this study with 1.8% each.

Fundus

Table 3: Distribution of patients by fundus results

Fundus eyer	Number	%
Normal	81	73,6
RDNP	28	25,5
RDP	1	0,9
Total	110	100,0

It results from this table that 26.4% of eyes had retinal lesions, of which 25.5% had RDNP, 1 eye (0.9%) had RDP.

Table 4: Fréquencies of retinal manifestations in SS homozygous sickel cell patients

Parameters	Number	%
§ RDNP		
Vascular tortuosity	19	17,3
Retinal palor	4	3,6
Solar black spots	3	2,7
OACR	1	0,9
Salmon haemorrhages	1	0,9
Occlusion arteriolar peripheral	1	0,9
§ RDP		
Retinal detachment	1	0,9

Vascular tortuosity was the most found retinal lesion with 17.3%, followed by retinal pallor and solar black spots with respectively 3.6 and 2.7%.

Table 5: VA of SS sickle cell patients according to the different retinal lesions parameters Visual acuity Total

Paramèters	Acuité visuelle								Total	
	Normal		Subnormal		Visually impaired		Blindness totale			
	n	%	n	%	n	%	n	%	n	%
§ RDNP										9
Vascular tortuosity	15	13,6	3	2,7	1	0,9	0	0,0	19	17,2
Peripheral retinal pallor	2	1,8	2	1,8	0	0,0	0	0,0	4	3,6
Solar black spots	3	2,7	0	0,0	0	0,0	0	0,0	3	2,7
OACR	0	0,0	0	0,0	1	0,9	0	0,0	1	0,9
Solmon haemorrhages	1	0,9	0	0,0	0	0,0	0	0,0	1	0,9
OAP	1	1,9	0	0,0	0	0,0	0	0,0	1	1,9
§ RDP										
Retinal detachment	0	0,0	0	0,0	0	0,0	1	0,9	1	0,9

Out of 19 eyes or 17.2% which had vascular tortuosity, 15 eyes or 13.6% showed normal VA, 3 eyes or 2.7% with subnormal VA, one eye or 0.9% was visually impaired. Retinal pallor was found in 4 eyes including 2 eyes or 1.8% with normal AV and 2 eyes or 1.8% with subnormal AV. The DR was also found in one eye with total blindness.

Discussion

Sex

In relation to the most represented sex, our study reports that out of 55 SS patients, 50.9% were female and 49.1% male with a sex ratio of 0.9. Dohvoma et al., [9] in their study of 88 sickle cell patients, reported 55.7% female with a sex ratio of 0.8. The two studies note a predominance of the female sex which could be explained by the female Hb level which is low and would be the basis of the symptomatic manifestations in sickle cell patients. The latter would push them to visit hospitals.

Age

In relation to the most represented age group, this study reported 80.0% of patients aged 15 to 24 years with an average age of 20.6 ± 7.1 years. Kueviakoe M et al in Togo [10], had reported a predominance of the age group from 18 to 27 years old, i.e. 43.9% in his study of patients over 18 years old. This would be explained by the irregularity in medical consultations in patients of advanced age following the different occupations and this would expose them to the

risk of sickle cell retinopathy. Indeed, the literature confirms that the older a sickle cell patient gets, the more susceptible he is to retinal lesions [2].

Taking hydroxyurea

Our study found 4 patients on hydroxyurea, none of whom had sickle cell retinopathy. This would be explained by the presence of the fetal hemoglobin produced which plays a protective role of the vessels

Visual acuity

In connection with the measurement of visual acuity in sickle cell patients, our study found a high frequency of normal visual acuity in 110 eyes, 98 or 89% of the eyes; it corroborates the study by Issa Abdi which reported on 76 eyes, 53 or 69.7% of eyes with normal visual acuity[11]. This confirms the low frequency of proliferative sickle cell retinopathy in SS.

Eye background

Frequency of retinopathy

In Cameroon, Dohvoma et al, found sickle cell retinopathy in 18 patients, or 20.5% out of 88 SS patients [9]. In Belgium, Nzimeni Y. reported 6 patients or 8.4% with sickle cell retinopathy out of 71 SS patients examined [12]. Our study found a higher frequency than those of the aforementioned studies in the SS form. Indeed, out of 55 SS patients, 16 or 29% had sickle cell retinopathy including 15, or 27.2% of RDNP and 1, or 1.8% of RDP. This could be explained by the quality of care which is not good in our environment Age group most affected by retinopathy

Compared to the age group most affected by retinopathy, our study showed that that of 15 to 24 years, was the most affected with 23.6%. Unlike K.P. Ballo et al in 1997 [13] who reported a predominance of sickle cell retinopathy in the age groups of 25 to 34 years at 23.8% and 15-24 years at 20.6% with extremes ages from 5 to 50 years old. The predominance of the age group from 15 to 24 years in our series could be justified by the high number of patients in the said age group with 80%.

The onset of proliferative retinopathy is around 25-39 years old for the SS forms, at the age of 8 years for the SC forms and from 15 to 24 years old for the other forms; the proliferative forms are evaluated at 8% in SC and 0.6% in SS; hence the interest in organizing screening from the age of 9 for the SC forms and 13 years for the SS forms; as confirmed by the Lemee Gilda study in France and Estepp JH et al. [2.7]. The confirmation of the latter corroborates our results which reported a prevalence of proliferative retinopathy for SS at 0.9% between the age of 35 and 44 years.

Conclusion

SS homozygous sickle cell disease affects approximately 2 to 3% of the population in the Democratic Republic of Congo, with minor retinal involvement. Given that it is a vascular disease, and given the seriousness of the ocular repercussions (particularly retinal) that the said disease can present; joint care by ophthalmologists and hematologists must be required in order to prevent blindness. We confirm that the older a sickle cell patient gets, the more susceptible he is to retinal lesions; hence it must be taken into account in order to check the vascular status of the sickle cell patient by performing a systemic fundus examination at least once a year. The eye being the mirror of the human body, the fundus will allow hematologists to have an idea of the installation of stenotic vasculopathy at the systemic level in order to improve management.

Declaration of interests

The Authors declare that they have no competing financial interests or known personal relationships which might appear to influence the work reported in this article.

Contribution of the authors

1. Bukasa Kizito Deogratias: funding, data collection, patient examination, essay and article writing
2. Kayembe Lubeji David: work supervision and coordination
3. Kelekele Kandjapa Joseph Theodor: examination of patients and correction of work
4. Kabedi Ngoy Nelly: production of retinophotos as needed and correction of the work
5. Nsambay Lukusa Delux: examination of patients
6. Gini Ambunga Yannick: registration of patients

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