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(RESEARCH ARTICLE)

Knowledge of sickle cell patients and attitudes and practices relating to the ophthalmological monitoring at the University Hospital of Bouaké (Côte d'Ivoire)

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Abstract

Introduction: Sickle cell disease is a potentially blinding haemoglobinopathy. The aim of this study was to assess the impact of the knowledge, attitudes and practices of sickle cell patients relating to the disease and its ocular complications on their adherence to ophthalmological monitoring of sickle cell disease.

Material and methods: Cross-sectional study by systematic recruitment of sickle cell patients received for any reason, in the outpatient department of the Bouaké University Hospital, during the study period.

Results: Out of 50 sickle cell patients aged from 8 to 71 years (average of 28.12) the male accounted for 52%. Pupils and students were predominant (60%) and the patients from urban areas represented 92% (n=46). The patients with a high level and secondary level of education represented respectively 42.00% (n=21) and 40.00%. The AS forms and the SC forms were the most represented with respectively 34% (n=17) and 42% (n=21). Those who did not know if sickle cell disease could cause ocular damages represented 52.00% (n=26) and among the 21 (42%) who said they knew, those who did not know that the disease could be blinding by its ocular involvement were 15 accounting for (71.43%). The sickle cell sufferers who had never consulted an ophthalmologist as part of the follow-up of the disease represented 74.00% (n=37) and among them, 33 (89.19%) affirmed that the monitoring consultation had never been prescribed to them.

Conclusion: Poor health education of the sickle cell patients is an obstacle to their adherence to the ophthalmological monitoring and the prevention of blindness due to sickle cell disease.

Keywords: Sickle cell disease; Sickle cell retinopathy; Prevention; Blindness

1. Introduction

Sickle cell disease is a common haemoglobinopathy among populations of African origin, around the Mediterranean, the Arabian Peninsula and the Indian subcontinent. It is hereditary with autosomal recessive transmission. It is a genetic disease of red blood cells caused by a structural abnormality of the globin molecule. This anomaly results from the replacement of glutamic acid by valine at position 6 of the beta chain of globin. Abnormal beta chains formed give hemoglobin "S" while normal beta chains of hemoglobin form hemoglobin "A" [1] About 5% of the world's population carries a sickle cell or thalassemia gene [1].

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This percentage reaches 25% in some regions. This prevalence in Côte d'Ivoire and Mali is around 12% [2]. It involves both the functional prognosis of many organs as well as the vital prognosis in severe forms. The ocular complications of this condition are multiple and affect practically all the structures of the globe. They are particularly serious due to their retinal locations [3]. Sickle cell retinopathy is the vascular consequence of the alteration of red blood cells due to the polymerization of deoxygenated hemoglobin "S". It becomes very insoluble and precipitates in the red blood cell. That one loses its flexibility and takes the shape of a sickle, and then bears the name “sickle". Triggered by local hypoxia or acidosis, sickling can lead to vascular occlusions in the peripheral retinal circulation, primarily in the arterioles and precapillary capillaries [4].

Retinal ischemia results in sickle cell retinopathy with complications such as neovascular proliferation, intravitreal bleeding or even retinal detachment by traction which can lead to irreversible loss of vision [1,4]. The prevalence of sickle cell retinopathy in carriers of the SC phenotype is 60% according to Boni et al [2]. It is frequently described in SS and SC patients and more rarely reported in heterozygous AS and S-Thal forms [5, 6, 7]. However, complications from sickle cell retinopathy are not inevitable. In fact, thanks to screening by ophthalmological monitoring, these complications can be prevented by laser photocoagulation of retinal ischemic areas [8,9]. Screening and early treatment of sickle cell retinopathy are the pillars of the fight against blindness related to sickle cell disease, especially in our context where resources are limited, the technical platform reduced while the prevalence of the disease is high.

However, what useful and necessary information do sickle cell patients have about the disease and its ocular complications? Furthermore, what impact could this knowledge have on their attitudes and practices relating to the ophthalmological surveillance of the disease or even on the incidence of blindness related to this disease?

To answer these questions, our study aimed to assess the impact of knowledge, attitudes and practices of sickle cell patients related to the disease and its ocular complications on their adherence to ophthalmological monitoring of sickle cell disease and, consequently, on prevention of blindness related to this disease.

2. Material and methods

This was a prospective descriptive cross-sectional study that lasted two and a half months (August 2 to October 17, 2018) conducted at the outpatient department of the Bouaké University Hospital (Côte d'Ivoire).

We systematically recruited patients with sickle cell disease, all forms combined, who came on their own or were accompanied for consultation for whatever reason. We excluded patients diagnosed and followed for less than one year, patients for whom the language barrier prevented understanding the survey sheet, patients whose sickle cell status was not biologically established.

After explaining the interest of the study, reassured as to the confidentiality of the answers by the anonymity of the survey sheets, the informed consent of the respondents was obtained verbally. The following data was studied:

- Socio-demographic data (age, sex, level of education and socio-professional activity) and the phenotype of the disease.
- age at diagnosis and time elapsed since diagnosis;
- the patient's knowledge of the disease, on the one hand and of its ocular complications on the other;
- The attitudes and practices of patients with sickle cell disease relating to the prevention of blindness linked to the disease through ophthalmological monitoring.

Quantitative variables were expressed as means with standard deviation and extreme values. Qualitative variables were expressed as proportions. The data was presented in the form of tables when necessary.

3. Results

3.1. Sociodemographic characteristics, phenotypes and time elapsed since diagnosis

From 70 patients identified, 50 were retained for the study. Out of those 50 patients, 52.00% (n=26) were male (sex ratio of 1.08). The average age of the patients was 28.12% years with extremes of 8 and 71 years with a standard deviation of 14.92. The age groups of 10 to 20 years and 20 to 40 years were the most represented with proportions of 32% each. School children and students represented 60% of the patients (n=30) (Table 1) and the education levels for

all the patients were respectively high for 42% (n=21), secondary level for 40% (n=20) and primary level for 8% of cases (n=4) Five patients (10%) had no education level.

Table 1 Distribution of sickle cell patients by occupation categories

Occupation categories	Size	Percentage (%)
Actors in the agricultural sector	3	6
Craftsmen	3	6
Merchants and service providers on duty	7	14
Schoolboys/girls and students	30	60
Public servants	6	12
Retired	1	2
Total	50	100

Schoolboys/girls and students accounted for 60% of the sickle cell patients

The SC and AS forms were the most represented with respectively 42% (n=21) and 34% (n=17). The SS and SFA2 forms accounted for 10% and the SAFA2 form accounted for 4%. Patients whose diagnosis dated back at most 5 years were the most represented with 48% (n=24), followed by those whose diagnosis dated back between 5 and 10 years with 24%. Patients with a diagnostic delay of between 10 and 15 years numbered 6, or 12%. Eight patients (16%) had been diagnosed for more than 20 years.

3.2. Knowledge about sickle cell disease

Thirty-eight sickle cell patients (76%) claimed that sickle cell disease was a blood disease while 12 patients (24%) considered it as a bone disease. Concerning the hereditary character, 45 patients (90%) agreed that sickle cell disease was a hereditary disease, 4 patients (8%) affirmed to have no idea and 1 patient (2%) affirmed that it is not in no way a hereditary disease. About the curability of the disease, 28 patients (56%) answered that sickle cell disease was not curable at the current stage of advances in medicine. Fifteen patients (30%) claimed that it was curable by medicine. Seven sickle cell patients (14%) said they had no idea.

Regarding their sources of information on sickle cell disease, 39 patients (78%) replied that they had received this information from health personnel, 6 patients (12%) replied that they had received it during talks with relatives or friends. Five patients (10%) claimed to have received them from the media (radio, television, and internet).

3.3. Knowledge about ocular complications of sickle cell disease

Concerning the possibility of an ocular attack by sickle cell disease in its evolution, 26 patients (52%) affirmed not to know it when 21 patients (42%) affirmed to know it. However, 15 of these 21 patients (71.43%) said they did not know if the ocular complications of sickle cell disease could lead to blindness. Three patients (6%) said that sickle cell disease has no ocular complications and the term “sickle cell retinopathy” had never been heard by 41 sickle cell patients (82%).

When asked whether ophthalmological follow-up could prevent the onset of sickle cell retinopathy, 36 patients (72%) answered yes, 1 patient (2%) answered no and 13 patients (26%) answered that they had no idea about the issue. In addition, 31 patients (62%) said that early detection of sickle cell retinopathy could prevent blindness, while 16 patients (32%) said they had no idea. Three sickle cell sufferers (6%) answered that early detection could not prevent progression to blindness.

3.4. Attitudes and practices relating to ophthalmological monitoring

As part of the follow-up of their disease, 37 patients (74%) said they had never had an ophthalmological consultation compared to 13 patients (26%) who said they had already done so at least once.

From these 13 patients who had already had an ophthalmological consultation, the average was 4.61 consultations with extremes of 1 and 15 consultations with a standard deviation of 4.70. From the 37 patients who had never had an ophthalmological consultation as part of the follow-up of their disease, those who stated that they had never received the prescription for an ophthalmological consultation were 33 and represented 89.19% (Table 2).

Table 2 Distribution of sickle cell patients' responses relating to the reason for which they have never carried out a monitoring ophthalmological consultation

Patients' responses	Size	Percentage (%)
Monitoring ophthalmological consultation never prescribed	33	89,19
Monitoring ophthalmological consultation prescribed but absence of functional symptoms	2	5,41
Monitoring ophthalmological consultation prescribed but not carried out due to negligence	1	2,70
Monitoring ophthalmological consultation prescribed but distant eye care centre	1	2,70
Total	37	100

Among the sickle cell patients who had never had a monitoring ophthalmological consultation, those for whom it had never been prescribed accounted for 89.19%.

4. Discussion

4.1. Sociodemographic characteristics, phenotypes and time elapsed since diagnosis

Male patients were predominant with a sex ratio of 1.08. Our result is like those of Diarra Y et al [10] as well as Diagne et al [11] who reported a male predominance. On the other hand, female predominance has been found in other studies on populations with sickle cell disease [12, 13, 14] and within the general population in urban areas in Togo [15]. These discrepancies are explained by the fact that genetically, the transmission of Hb S is not linked to sex. The most represented age groups were those of 10-20 years and 20-30 years with 32.00% each. The average age was 28.12 years, close to that found by Cheucheu et al which was 31.5 years [16]. Similar averages were found in studies by Balo in Togo [15] and Asnani in Jamaica [17]. The relative youth of this population could be explained by the structure of the Ivorian population as described by the National Institute of Statistics, namely a predominantly young population [18]. With regard to the level of education, the high level was the most represented with 42.00%, followed by the secondary level 40.00%.

The predominance of a high level of education could be explained by the urban setting of the study. This level of education could be an advantage in assimilating health information disseminated. This trend was also observed for the professional categories for which more than half (60%) of the sample was made up of schoolchildren and students followed by traders and service providers. A study carried out in Nigeria [19] showed that young people who are single or have graduated from higher education institutions could be the most appropriate target in the prevention and control of sickle cell disease [20] Does the high representation of the paucisymptomatic forms SC and AS, in general, in our series dominated by pupils and students, confirm such a finding? Indeed, as pointed out by some authors [21], absenteeism and school delays linked to morbidity constitute a real obstacle to the education and instruction of carriers of chronic diseases in general, and sickle cell patients in particular. So, this social category could be more sensitive to awareness messages. Sickle cell patients whose diagnosis dated back at most 5 years were the most represented with 48.00%. Early diagnosis could help reduce morbidity and mortality related to sickle cell disease as reported by Cheucheu et al in their study [16].

4.2. Knowledge about sickle cell disease

The proportion of correct answers relating to sickle cell disease concerning, definition in relation to the tissue concerned (76.00%), heredity (90.00%), and curability of (56.00%), was relatively high. These rates reflect a good general knowledge of sickle cell disease by affected subjects. This could be explained by the fact that the sources of information on sickle cell disease were health personnel in 78.00% of cases.

Could the low rate of patients having received information through the media be indicative of a lack of awareness-raising policy on sickle cell disease, which nevertheless constitutes a public health problem in our regions?

If we want to reduce the rate of transmission of the disease, it will absolutely be necessary to resort to all modes of communication, in this case, mass media.

4.3. Knowledge about ocular complications of sickle cell disease

Patients' knowledge of “eye-sickle cell disease" relationship can be considered very low. As in the study conducted by Mukinayi et al. in 50 Congolese families, people had a low level of knowledge about the possibility of complications related to sickle cell disease [22]. This could be explained by the silent evolution of the ocular involvement of the disease which promotes the occurrence of potentially blinding lesions discovered late and sometimes revealing the condition, unlike occasional disabling symptoms of other locations, particularly bones.

This poor level of information for sickle cell patients could also be explained by a failure in the health education of sickle cell patients and even in their overall care, which is the responsibility of health personnel, as Cheucheu et al have already pointed out [16]. In fact, if the general practitioner or the hematologist prescribed the ophthalmological consultation more often, even without explanation, some patients could assume the possibility of ocular complications without knowing which ones: “if my doctor asks me to go see the ophthalmologist then ocular complications linked to sickle cell disease may be possible”.

This prescription could then arouse the curiosity of patients and motivate research or questions on the relation between their disease and ocular health. Ignoring the risks of ocular complications or even blindness to which patients with sickle cell disease are exposed, combined with the financial difficulties that some of them are sometimes confronted with, favors the occurrence of complications that can lead to blindness. In general, the overall lack of knowledge concerning the ocular impact of sickle cell disease could be attributed to the lack of communication between the patient and the medical professionals (general practitioners, hematologists, ophthalmologists, etc.).

However, the lack of curiosity on the part of the patients, who for the most part still had an education level higher than or equal to secondary school, could also be blamed. The fight against morbidity and mortality related to sickle cell disease requires a multidisciplinary approach and must call on the commitment of the patient himself. This was mentioned by Tanabé P et al [23] in a qualitative analysis focusing on the best self-practices in the management of sickle cell disease and Animasahun et al. [24], including improving the knowledge of the various health actors concerned.

4.4. Attitudes and practices relating to ophthalmological monitoring

In our study, nearly 3 out of 4 sickle cell sufferers (74%) stated that they had never consulted an ophthalmologist as part of the follow-up of their condition, yet the interest of such follow-up in sickle cell sufferers, all types combined, is clearly established [25]. Moreover, in addition to adequate medical care, the maintenance of a disciplined lifestyle, stable or sedentary occupation and family support are determining factors for the survival of the patient with sickle cell disease. To this end, health education of patients and their parents, in particular by raising awareness of treatment and hospital follow-up from diagnosis, are the main contributing factors [26].

From a total of 13 sickle cell patients who had undergone consultations, the average number of consultations was 4.61 with extremes ranging from 1 to 15 consultations and a standard deviation of 4.70. This finding highlights the lack of regularity in the ocular monitoring of the condition. From 37 people suffering from sickle cell disease who had never had an eye monitoring consultation, 33 (89.19%) said that this consultation had never been prescribed for them, which means that it had been prescribed only for 4 people suffering from sickle cell (10, 81%) who despite this prescription never made it.

Once again, the lack of awareness on the part of the medical staff could be at the source of the poor health education of sickle cell patients with the consequence of poor adherence to ophthalmological monitoring of sickle cell patients.

5. Conclusion

From this study it appears that people suffering from sickle cell disease representing a slight majority of men with a good level of education, overall, had a relatively low level of knowledge about sickle cell disease and even lower about its ocular complications. This entails the risks of misunderstanding the need for ophthalmological monitoring of their disease. In addition to this, the attitude of general practitioners and hematologists characterized by the non-prescription of monitoring ophthalmological consultations constitute real obstacles to the fight against blindness linked to ocular complications of sickle cell disease.

The attitudes and practices of sickle cell patients related to the prevention of blindness due to sickle cell disease, which is quite widespread in our environment, reflect insufficient health education for sickle cell patients among our populations. It appears from this study that any action aiming at reducing the incidence of blindness related to sickle

cell disease requires better involvement of first contact health personnel, patient associations and the media in raising awareness among sickle cell patients.

Compliance with ethical standards

Acknowledgments

Recognition to sickle cell patients who agreed to participate in the study.

Disclosure of conflict of interest

As co-authors of this work, we declare a complete absence of conflict of interest in the production of this article

Statement of informed consent

We obtained verbal consent from all survey participants, for adult patients. For minor patients (the majority in Côte d'Ivoire is 18 years old), we obtained this agreement from the adult parent who accompanied the patient.

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