PATTERN OF PRESENTATIONS SEEN IN SICKLE CELL RETINOPATHY PATIENTS AT EYE FOUNDATION HOSPITAL LAGOS, NIGERIA

A O HASSAN *FRCS, FRCOph, FWACS*
O ODERINLO *FRCSEd, DRCOphth*
O OKONKWO *FRCSEd, DRCOphth*
FO OLUYADI *FWACS*
AO OGUNRO *FWACS*
SA OKE *DO (WACS)*

Eye Foundation Hospital, 27B Isaac John Street, Ikeja GRA, Lagos

ABSTRACT

Objective: To describe the pattern of presentation of sickle cell retinopathy patients who presented at the Eye Foundation Hospital Lagos, Nigeria between January 2002 and March 2003.

Materials and Methods: The medical records of 27 patients who presented at the Eye Foundation Hospital with retinal changes due to sickle cell disease within a 15-month period were reviewed retrospectively.

Results: A total of 27 patients were evaluated, 67% were male while 33% were female. The mean age at presentation was 36.18 years with female patients tending to present earlier than male patients.

The most common complaint at presentation was a sudden drop in vision seen in 63% of the patients evaluated. The dominant genotype among the patients was SC with 81.5%; 7.4% were SS and 11.1% were AS.

The duration from onset of symptoms to presentation was evaluated. The median duration at presentation was greater than 12 weeks after onset of symptoms; 85% of patients presenting had proliferative retinal changes.

Proliferative sickle retinopathy (PSR) changes were classified according to Goldberg's classification of 1971. Stage 4 PSR was the most common stage seen, occurring in 48% of patients.

Conclusion: Sickle cell retinopathy patients seen at the Eye Foundation Hospital generally presented after 12 weeks of onset of sudden drop in vision. They were mostly in the SC genotype group and mostly had stage 4 proliferative retinopathy according to Goldberg's classification.

Key words: sickle cell retinopathy, pattern of presentation, genotype

INTRODUCTION

Sickle cell disease (SCD) is the most common haemoglobinopathy[1] and it is associated with multi-organ complications which may include avascular necrosis of the head of the femur, splenic infarction, and retinopathy.[2] Retinopathy may be proliferative or non-proliferative. Proliferative sickle cell retinopathy (PSR) is often associated with visual loss in the later stages. The incidence of PSR in SCD patients varies from 5 to 10% depending on the genotype, being commoner in SC, than SS and S-thal.[3] Since sickle cell patients,[4] are now living longer, many are seen in the fourth and fifth decades of life with sudden onset of visual symptoms mainly due to vitreoretinal complications such as vitreous haemorrhage and retinal detachment[5]

Prevention of this visual loss is dependent on early detection and treatment. We must understand the pattern of presentation to be able to proffer solutions to this peculiar problem.

In this study, we share our experience of the pattern of presentation seen in sickle cell patients presenting at our hospital between January 2002 and March 2003, with complaints due to retinopathy.

MATERIALS AND METHODS

The medical records of 27 patients who presented at the Eye Foundation Hospital with retinal changes due to sickle cell disease within a 15-month period were reviewed. Relevant information obtained formed the

* *Author for correspondence*

database for the analysis. All patients were seen within the study period of January 2002 and March 2003,all patients history were taken to include symptoms at presentation, genotype and duration of symptoms. patients who did not know their genotype were sent to the laboratory to have haemoglobin electrophoresis determination of their genotype.

Examination included visual acuity assessment, extraocular muscle motility, pupillary reaction. Slit lamp assessment of the anterior segment, intraocular pressure by applanation tonometry, and dilated funduscopy with binocular indirect ophthalmoscope and 78D lens assessment of the macula. Fundus photographs were taken in 14 patients and 4 patients had fundus flourescein angiography.

RESULTS

A total of 27 patients were evaluated 67% (18 patients) were male while 33% (9 patients) were female. The mean age at presentation was 38.18 years. Female patients tended to present earlier than the males with a mean age at presentation of 34.7 years compared to 38.7 years for the males. In view of the difference in number of male and female patients in our cohort of patients, this difference was not considered statistically significant

The most common complaint at presentation was a sudden drop in vision seen in 63% of the patients evaluated, 22% of the patients presented with a history of seeing floaters while 4% presented with other complaints not indicative of retinal disease but which were discovered after dilated funduscopy which was done for all new patients. The remaining patients (11%) did not have complaints and only presented at the hospital for routine check (figure 1).

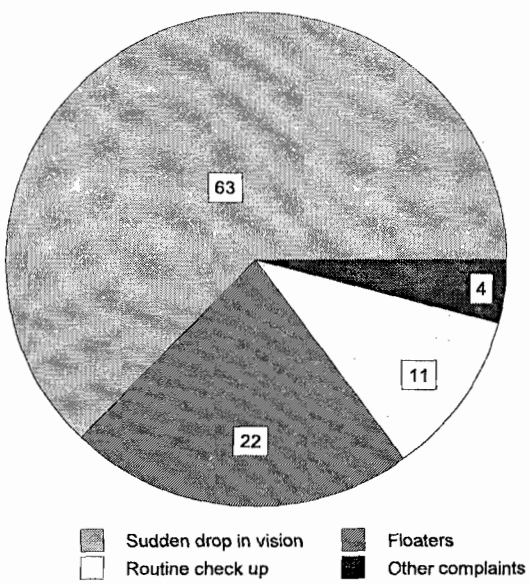

Figure 1. Complaint at presentation

Patients were also evaluated according to their haemoglobin genotype. The most common genotype seen in the patients was SC (81.5%); the others were 7.4% SS and 11.1% AS.

Only 20 patients were evaluated for duration of symptoms before presentation due to inadequacies in the records of the other patients.

The patients who presented after 12 weeks of onset of symptoms formed the largest group (40.7%); 22.2% presented before 4 weeks of onset of symptoms, 3.7% presented between 5-8 weeks of onset while 11.2% presented between 8 and 12 weeks of onset. The median duration at presentation was greater than 12 weeks.

Goldberg's classification stages were employed in the evaluation of proliferative retinopathy changes. Goldberg's classification of 1970 [6] is as follows:

Stage 1: peripheral arteriolar occlusion
Stage 2: peripheral arteriovenous anastomoses
Stage 3: neovessel growth from arteriovenous anastomoses which have a fan shaped configuration (sea fans)
Stage 4: varying amounts of vitreous hemorrhage
Stage 5: vitreous traction and retina detachment

Of the eyes presented, 44.2% (19) were at stage 4, compared to 24.3% at stage 5 and 27.9 % at stage 3. Only 2.3% of eyes were at stages 1 and 2 (table 1). Hence PSR at stage 4 was the most common stage at presentation.

Table 1. Stage of proliferative sickle cell retinopathy at presentation

Stage of PSR	Right eye	Left eye	Total number	Percentage of total
1	-	1	1	2.3%
2	-	1	1	2.3%
3	8	4	12	27.9%
4	9	10	19	44.2%
5	5	5	10	24.3%
Total	22	21	43	100.0%

The remaining 11 eyes had non-proliferative changes; 6 eyes (54.5%) had black sunbursts, while 3 eyes (18.3%) had retinal holes. Black sunbursts were, therefore, the most common non-proliferative sickle cell retinopathy change in our cohort of patients (table 2).

DISCUSSION

Patients with retinal changes as a result of sickle cell disease often present at the Eye Foundation Hospital, Ikeja, Lagos. This is not unexpected as an estimated 2 million Nigerians have sickle cell disorders and over 25 million are carriers. Since our hospital is located in southwest Nigeria, we fall within a belt that has a high prevalence rate for sickle cell disorders[7]

On cursory evaluation we realized that most patients only presented when they had symptoms. The most common presenting complaint was a sudden drop in vision seen in 63% of patients evaluated; this often occurred in advanced proliferative retinopathy stages when vitreous haemorrhage or retinal detachment occurs. Even with this sudden drop in vision, a majority of the patients evaluated (40.7%) still waited until after 12 weeks before presenting at the hospital.

Only 3 patients presented for routine check up; 2 of these had only non-proliferative changes of black sunburst in both eyes while the last one had stage 3 proliferative retinopathy in one eye (left eye), the seafans were still flat on the retina and were easily treated.

Proliferative retinopathy changes were evaluated using Goldberg's classification of 1970. As expected, visual acuity usually drops at stage 4 due to vitreous hemorrhage and in stage 5 due to retinal detachment. Out of the 43 eyes with PSR 30 eyes (69.8%) had a sudden drop in vision; 19 of these (44.2%) were already at stage 4 and had vitreous haemorrhage (table 2).

Table 2. Symptoms and ocular findings

Ocular findings	Symptoms	Number of eyes		
Stage of proliferative sickle cell retinopathy	Sudden drop in vision	Floaters	Routine check up	Other complaints
1	1(1.85%)c	-	-	-
2	1(1.85%)c	-	-	-
3	5(9.26%)c	6(11.1%)	1(1.85%)	-
4	16(29.6%)	3(5.55%)	-	2(3.7%)
5	7(12.9%)	1(1.85%)	-	-
Non proliferative sickle cell retinopathy				
Black sunburst	1(1.85%)	-	5(9.26%)	-
Retina holes	2(3.7%)	-	-	-
Salmon patch hemorrhages	3(5.55%)	-	-	-

Table shows relationship between ocular findings in each eye and symptoms. Patients presented with c appears in eyes where the complaint was basically for the contralateral eye.

Some investigators[8] have noted that it takes about a decade for progression of retinal changes through each of Goldberg's stages of PSR. Hence, most of these changes would have started 30-40 years before the patients in our cohort presented, supporting the average age at presentation of 36.18 years.

The difficulty with the management of stage 4 PSR in our environment was emphasized in our previous article[9] seafan neovascularizations that were elevated with multiple feeder vessels were more difficult to control with only laser photocoagulation resulting in a 23.1% incidence of recuurent vitreous hemorrhage .Patients who presented in earlier in stage 4 had better visual outcomes after laser treatment in our cohort of patients we usually observed patients at stages 1 and 2, offered laser photocoagulation for stage 3 and early stage 4 who had only localized mild vitreous hemorrhage, patients with pronounced vitreous hemorrhage and stage 5 were offered surgery posterior vitrectomy usually.

With the availability of fundus flourescein angiography in our facility, PSR changes can be identified at stages 1 and 2 before they become ophthalmoscopically visible. The fundus photograph in figures 2a and 2b show peripheral arteriovenous anastomosis with the distal retina largely avascular and non-perfused, the changes being more obvious on flourescein angiography. Both photographs in figure 2a and 2b are of the same patient showing the same area of the fundus. Early detection and prompt management will prevent visual loss from late complications.

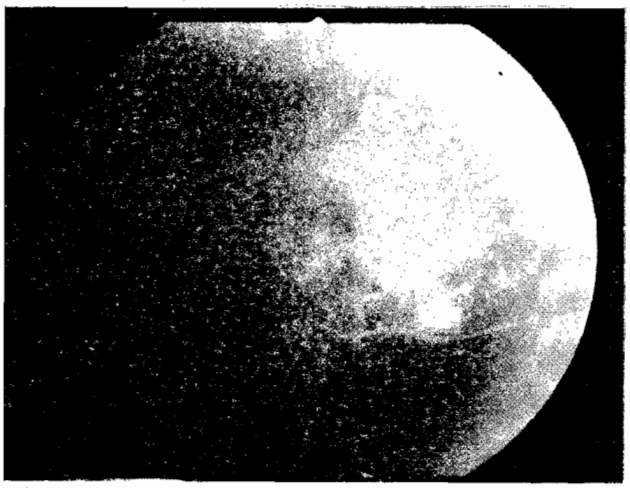

Figure 2a

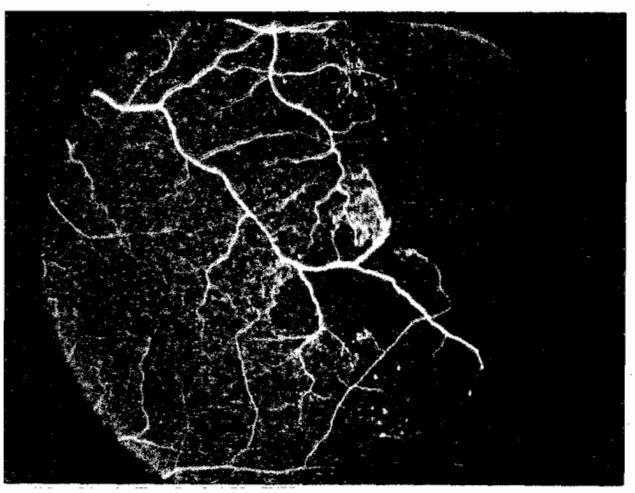

Figure 2b

Early proliferative sickle retinopathy changes in a young patient presenting for routine evaluation. Note the peripheral areas of retina ischaemia and vascular occlusion seen as peripheral areas of capillary non perfusion on fundus flourescein angiography.

It is important to improve the awareness of the populace through 'Know Your Genotype' programmes with emphasis on genotype SC and its ocular complications, as this is the most common genotype with ocular complications, and least likely to be detected because of paucity of systemic manifestations.

Physicians, obstetricians, paediatricians and haematologists have to be better educated about ocular complications of sickle cell disease to encourage routine referrals for ocular evaluation so as to enable early detection.

CONCLUSION

Sickle cell retinopathy patients seen at Eye Foundation Hospital typically presented after 12 weeks of onset of sudden drop in vision, they were mostly of genotype SC and had mostly proliferative retinopathy at stage 4 of Goldberg's classification.

REFERENCES

1. Morel C. Retinal involvement in haemoglobinopathy. *J Fr Ophthmol* 2001; **24(9):** 987-92.
2. Koduri P R, Agbemedzo B. , Nathana. Hemoglobin S-C disease revisted: Clinical study of 106 adults. *Am J Hemataol* 2001; **68(4):** 298-300.
3. Babalola O E, Wambebe C O. When should children and young adults with sickle cell disease be referred for eye assessment. *Afr J Med Med Sci* 2001; **30(4):** 261-3.
4. Mckerrel T D, Cohen H W, Billet H H. The older sickle cell patient. *Am J Hematol* 2004; **76(2):** 101-6.
5. Penman A D , Serjeant G R. Recent advances in the treatment of proliferative sickle cell retinopathy. *Current Opinion in Ophthalmology* 1992; **3(3):** 379-88.
6. Goldberg M F. *Sickle Cell Retinopathy: Clinical ophthalmology* Harper and Row, 1976, 1-44.
7. Akinyanju Olu. Control of sickle cell disorders: the way forward. *Sickle Cell Bulletin.* January 2001; **1.**
8. Steve T Charles, Henry M Clayman. *Retinal Surgery: Contemporary ophthalmic surgery.* 1st ed. Moshy Company. 1990: 715-728.
9. Hassan A O, Oderinlo O, Okonkwo O. Visual outcome after laser photocoagulation for stage 4 proliferative sickle cell retinopathy. *Nigerian Journal of Ophthalmology* 2004; **12(1)**: 19-22.