

Full Length Research Paper

Eye manifestations of children with homozygous sickle cell disease in Nigeria

I.O. George*¹ and S.A.H.Cookey²

¹Departments of Paediatrics, University of Port Harcourt Teaching Hospital, Port Harcourt, Nigeria

²Departments of Ophthalmology, University of Port Harcourt Teaching Hospital, Port Harcourt, Nigeria

Abstract

Sickle cell disease can damage blood vessels in the eye and cause scarring and detachment of the retina, which can lead to blindness. The purpose of the study is to determine the eye manifestations in Nigerian children with sickle cell anemia. Children (aged 6months to 16years) with sickle cell anaemia were examined for ocular abnormalities from 1st January to 31st June 2010 at the Ophthalmology Clinic of the University of Port Harcourt Teaching Hospital, Nigeria. Routine ophthalmic examination included measurement of visual acuity, inspection of the adnexa and cornea, refraction, slit-lamp examination and dilated fundoscopy. 94 children examined, 64(68.1%) were males and 30 (31.9%) were females. Ocular abnormalities were found in 32 (34.0%) children. Conjunctival signs were present in 8(8.5%) of the children, retinal vascular tortuosity in 13(13.8 %), arteriovenous crossing in 1(1.1%), optic nerve pallor in 10(10.6%) and glaucoma 1(1.1%). Ocular abnormalities are common. However, no evidence of proliferative retinopathy was found in this study which is in tandem with those previously published in Africa.

Keywords: Eye manifestations, sickle cell anaemia, children, Nigeria.

INTRODUCTION

Sickle cell disease is one of the commonest genetic disorders (Stuart and Nagel, 2004). It has variety of pathological conditions resulting from the inheritance of sickle haemoglobin either in a homozygous state (SS) or in a heterozygous state (SC) (Stuart and Nagel, 2004).

The ocular manifestations of sickle cell disease (SCD) result from vascular occlusion, which may occur in the conjunctiva, iris, retina, and choroid (Akinsola and Kehinde, 2004). The eye effects vary and the degree of ocular involvement is not necessarily based on the severity of the systemic disease (Babalola and Wambebe, 2005). The conjunctiva may show the sickling sign (Kaimbo et al., 2000); the anterior segment may show iris atrophy with anterior or posterior synechiae secondary to occlusions in the iris circulation (Kaimbo et al., 2000). The posterior segment may show optic, macular, nonproliferative and proliferative retinal changes (Kaimbo et al., 2000).

Sickle cell retinopathy can be further divided into

nonproliferative and Proliferative. The nonproliferative ocular signs include: venous tortuosity, silver wire arterioles, salmon patch hemorrhages (oval pale retinal hemorrhages), intraretinal hemorrhages, black sunbursts (melanin deposits on the retina due to hemorrhages following small branch retinal artery blockages), macular arteriole occlusions, retinal vein occlusions, angiod streaks, and dark without pressure (Sayag et al., 2008).

Proliferative retinopathy involves the growth of abnormal vascular fronds that place patients at risk of vitreous hemorrhage and retinal detachment. The initiating event in the pathogenesis of proliferative disease is thought to be peripheral retinal arteriolar occlusions. Local ischemia from repeated episodes of arteriolar closure is presumed to trigger angiogenesis through the production of endogenous vascular growth factors, such as vascular endothelial growth factor and basic fibroblast growth factor (Joussen et al., 2007).

Although peripheral vaso-occlusion may be observed as early as 20 months of age (Akinsola and Kehinde, 2004), clinically detectable retinal disease is found most commonly between 15 and 30 years of age (Goldberg, 1982). Sickle retinopathy is found more often and earlier

*Correspondence Author E-mail: geonosdemed@yahoo.com

in SCD-SC, but is also common in SCD-SS and sickle thalassemia.

Treatment is reserved for eyes that have progressed to proliferative retinopathy, and patients are thus at risk for severe visual loss from bleeding and retinal detachment. Given the high rate of spontaneous regression and lack of progression of neovascularization in some eyes, the indications for treatment of retinal neovascularization are not always clear. Therapeutic intervention usually is recommended in cases of bilateral proliferative disease, spontaneous hemorrhage, large elevated neovascular fronds, rapid growth of neovascularization, and cases in which one eye has already been lost to proliferative retinopathy.

In view of the potential effect of sickle cell anaemia on the eyes, we deemed it necessary to determine the frequency of ocular abnormalities in our patients. The findings will serve as tools for proper management and advocacy in Nigeria.

MATERIALS AND METHODS

This was a prospective, cross-sectional study of Children with sickle cell anaemia seen at the sickle cell clinic of the University of Port Harcourt Teaching Hospital (UPTH), Nigeria. The patients were referred to the ophthalmology department of UPTH for eye examination from January 1 to June 31, 2010. Their ages ranged from 6 months to 16 years. Ninety six children were examined, 2 were excluded because of poor collaboration at eye examination. For this study, the records of 94 children were considered. Diagnosis of sickle cell anaemia was made based on clinical features and confirmed by haemoglobin electrophoresis. Children with diabetes were excluded from the study.

All patients underwent measurement of visual acuity, refraction, slit lamp examination and dilated fundus examination. Visual acuity was estimated by using illiterate E, or Snellen visual acuity charts among older children. The ability to fixate and follow a light, and a judgment made as to whether the child was sighted or not was used for other children. All children were dilated with 0.5 or 1% tropicamide collyrium and were examined once. Fluorescein angiography was not performed. On ocular examination, the presence or absence of the conjunctival sign, anterior or posterior segment changes were considered. The conjunctival sign consisted of the presence of regular venous dilatations, or other signs such as capillary micro aneurysms, telangiectatic dilatations and comma-shaped vessels. Retinal findings were classified in nonproliferative retinopathy and proliferative retinopathy. Nonproliferative retinopathy consisted of retinal venous tortuosity (occasionally arteriolar tortuosity), a silver wire appearance of the retinal arterioles, CRAO or CRVO with haemorrhages, angiod streaks, salmon-colored patches. Proliferative

retinopathy consisted of retinal artery occlusions, choroidal vascular occlusions, retinal haemorrhages, and neovascularization. Data was entered into a Microsoft Excel Spread sheet and analysed using descriptive statistics. Results were presented in text and tables.

RESULTS

We examined a total of 94 children with haemoglobin SS genotype during the study period. Of these 64(68.1%) were males and 30 (31.9%) were females giving a male: female ratio of 2.1:1. Details of age and sex distribution are shown in Table 1. None of patients presented with subjective ocular symptoms. Ninety two (97.9%) children had normal vision while 2 (2.1%) had mild ametropia (Table 2). Ocular abnormalities were found in 32 (34.0%) children. Conjunctival signs were present in 8(8.5%) of children, retinal vascular tortuosity in 13(13.8 %), arteriovenous crossing in 1(1.1%), optic nerve pallor in 10(10.6%) and glaucoma 1(1.1%). All the ocular lesions increased with age as shown in Table 2.

DISCUSSION

In this report 34.0% of children with homozygous SS had ocular abnormalities. This rate is lower than previous Nigerian study which reported a prevalence rate of 90% (Elebesunu-Amadasu and Okafor, 1985).

Ocular manifestations of sickle cell eye disease can be severe enough to lead to blindness which could even be of sudden onset. Majority of our patients had good vision. Abnormalities of the conjunctival vessels are common in patients with sickle cell disease.

Conjunctiva signs were present in 8(8.6%) of our patients reviewed and this is at variance with a Nigerian study (Obikili et al., 1990) in which 77% of their 78 homozygous sickle cell HbSS patients had conjunctiva signs. This may be due to the fact that parents are now more informed about the disease and its complications and also tend to have better health seeking behaviour than many years ago. Demonstration of conjunctival abnormalities is a useful clinical finding and may determine clinical severity. The earliest published observations reported widespread slugging of the blood and saccular dilatations in the conjunctival vessels (Serjeant et al., 1972). The presence of saccular and sausage-like dilatations which were packed with red cells was confirmed by Goodman and colleagues and also noted that they were transient in nature (Serjeant et al., 1972). Paton (1964) described multiple, short, comma shaped capillary segments which appeared pathognomonic of sickle cell disease.

Tortuosity of major retinal vessels was found in 13.8% of patients with SS disease in our study. The mechanism by which tortuosity develops is obscure. Our finding is at

Table 1. Age and Gender Distribution of 94 Children with Homozygous Sick Cell Disease

Age(Years)	Male (%)	Female (%)
<5	14(14.9)	6(6.4)
5-<10	26(27.7)	18(19.1)
10-16	24(25.5)	6(6.4)

Table 2. Age Distribution of ocular abnormalities in Homozygous Sick Cell Disease

	Age (year)			Ocular Abnormalities		
	CS	RVT	A-V crossing	A-V crossing	ONP	Ametropia
<5	1	2	0	0	2	0
5-<10	3	4	0	0	3	1
10-16	4	7	1	1	5	1

Key: CS- Conjunctival sign, RVT- Retinal vascular tortuosity, A-V(Arterio-venous), ONP-Optic nerve pallor.

tandem with that reported previously (Majekodunmi and Akinyanju, 1978; Akinsola and Kehinde, 2004; Traore et al., 2006)

Sickle cell retinopathy usually occurs in patients over 20 years of age (Eruchalu et al, 2004). Proliferative retinopathy was not found in our patients. This is not surprising since this condition occurs commonly in children above 20 years and our targeted population were children 16 years and below. Akinsola and Kehinde (2004), Lagos, Nigeria and a Cohort study from Jamaica also did not report any case of proliferative retinopathy. However, Abiose, 1972, in a study of Nigerian children with sickle cell disease (Hb SS), found one out of 92 children with signs of neo-vascularization.

We found glaucoma in 1.1% of the cases studied. Individuals with sickle cell disease are prone to neovascular glaucoma (Bergren and Brown, 1992). This may be secondary to obstruction of the anterior chamber outflow tract. Every Black patient presenting with traumatic hyphaema should be screened for sickle cell disease (Jeffers, 1995). Neovascular glaucoma can be treated with retinal laser photocoagulation (Bergren and Brown, 1992).

Sickling is a process that generally affects all organs. One of the major negative effects of the disease is associated with retinal damage. Sickle cell retinopathy is a condition that develops due to insufficient oxygen delivery to the retina. This is the reason why all patients suffering from sickle cell disease are supposed to undergo regular eye examination. By doing so, the disease may be diagnosed on time and irreversible damage to the eye successfully prevented.

The main limitations of this study include the relatively small sample size, non-utilization of fluorescein angiography thus some posterior segment changes could have been missed.

CONCLUSION

Eye abnormalities are common in children with sickle cell anaemia in Port Harcourt; Nigeria. There is need for early detection and treatment so as to prevent progression of lesions. Hence, it is recommended that a yearly examination be carried out in all children with sickle cell disease, irrespective of age or electrophoretic pattern.

REFERENCES

- Abiose A (1978). Ocular findings in children with homozygous sickle cell anaemia in Nigeria. *J Paediatr Ophthalmol Strab*; 15: 92-95.
- Akinsola FB, Kehinde MO (2004). Ocular findings in sickle cell disease patients in Lagos. *Niger. Postgrad. Med. J.* 11(3):203-6.
- Bergren RL, Brown GC (1992). Neovascular glaucoma secondary to sickle-cell retinopathy. *Am. J. Ophthalmol*; 113(6):718-719.
- Babalola OE, Wambebe CO (2005). Ocular morbidity from sickle cell disease in a Nigerian cohort. *Niger. Postgrad. Med. J.* 12(4):241-4.
- Elebesunu-Amadasu M, Okafor LA (1985). Ocular manifestations of sickle cell disease in Nigerians; experience in Benin City, Nigeria. *Tropical and Geographical Medicine*; 37(3): 261-263
- Eruchalu UV, Pam VA, Akuse RM (2004). Sight threatening retinopathy in an eight year old Nigerian male with sickle cell β^0 thalassaemia: Case report. *Annals of African Medicine Vol. 3, No. 3: 141 – 143.*
- Goldberg M (1982). Sickle cell retinopathy in *Clinical Ophthalmology*. Ed. Duane T and Jeager E. Harper and Row publishers, Philadelphia; 3(17):1-45.
- Jeffers JB (1995). Traumatic hyphaema management. *Curr Concepts Ophthalmol*; 3:34-38.
- Joussen AM, Gardner TW, Kirchhof B and Ryan AJ (2007). Sickle cell retinopathy and haemoglobinopathies. *Retinal vascular disease Springer Berlin Heidelberg*; 700-34.
- Kaimbo WA, Kaimbo DI, Ngyulu MR, Dralands L, Missotenn L (2000). Ocular findings in children with homozygous sickle cell disease in the Democratic Republic of Congo. *Bull Soc Belge Ophthalmol*; 275:27-35.
- Obikili AG, Oji EO, Onwukeme KE (1990). Ocular findings in homozygous sickle cell disease in Jos, Nigeria. *Afr. J. Med. Med. Sci.* 19(4):245-50.
- Majekodunmi SA, Akinyanju OO (1978) Ocular findings in homozygous sickle cell disease in Nigeria. *Can. J. Ophthalmol.* 13 (3):160-162.

- Paton D (1962). Conjunctival sign of sickle cell disease. Further observations. Arch Ophthalmol; 68: 627-32.
- Serjeant GR, Serjeant BE, Condon PI (1972). The conjunctival sign in sickle cell anemia. A relationship with irreversibly sickled cells. JAMA; 219: 1428-31.
- Sayag D, Binaghi M, Souied EH, Querques G, Galacteros F, Coscas G (2008). Retinal photocoagulation for proliferative sickle cell retinopathy: A prospective clinical trial with new sea fan classification. Eur. J. Ophthalmol. 18(2): 248 – 254
- Traore J, Boitre JP, Bogorey IA, Traore L, Diallo A (2006). Sickle cell disease and retinal damage: a study of 38 cases at the African Tropical Institute (IOTA) in Bamako. Med. Trop. (Mars); 66(3):252-4.