

Pattern of Presentation of Sickle Cell Retinopathy in Ibadan

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Abstract

Background: Sickle cell retinopathy is not uncommon in Nigeria. Most cases of retinopathy occur in patients with the hemoglobin SC genotype. A significant proportion present late.

Methods: A review of Thirty three (33) cases of sickle retinopathy seen over 3years (2008-2010) that presented to the retinal unit of the department of Ophthalmology, University College Hospital, Ibadan were reviewed. Demographics and pattern of presentation were recorded in the pro forma prepared for the study.

Results: HbSC produces most of the presentations. Male: Female ratio was 3:1. About 2/3 of the patients were below 40 years of age. 24/33 (70%) of the patients presented with proliferative retinopathy. About half of them were blind at presentation. Pan retinal laser photocoagulation is the commonest mode of treatment. The role of AntiVEGF intravitreal injection in the management of sickle retinopathy is discussed.

Conclusion: General/Family physicians are to refer patients with sickle retinopathy for regular ophthalmic examinations to identify treatable lesions amenable to intervention.

Keywords: Sickle retinopathy; Blindness; Laser photocoagulation; AntiVEGF; Screening

Introduction

The prevalence of the hemoglobin S gene in Nigeria is between 20 and 25% [1]. About 25% of adult's population in Nigeria have the sickle cell trait, AS, while the Hb C trait is largely confined to the Yoruba people of southwestern Nigeria in whom it occurs in about 6%. Other variant hemoglobin including beta thalassemia is rare, but alpha thalassemia occurs in 39% (32% with 3 alpha-globin genes; 7% with 2 alpha-globin genes [2].

In Nigeria, HbSC produces most of the retinopathic changes with previous studies showing patient's presentation at the late stages of the disease [3-13].

In the presence of oxidative stress, the red blood cells of sickle cell patients take on a sickle shape instead of the normal bi-concave disc. They become trapped in the small vessels leading to ischemia, hypoxia, and tissue necrosis. The hypoxia leads to more sickling and a cycle is created.

SS patients have the worst systemic complications while SC and Sthal patients have the most severe ocular problems. This is because the blood is more viscous in the latter group as small retinal arterioles occlude more easily.

Pre-proliferative retinal findings include salmon-patch hemorrhages and black sunburst pigments. These are retinal pigment epithelial layer reaction to hemorrhage and choroidal infarction [14]. Others are intraretinal refractile bodies, and silvering of peripheral arterioles. Proliferative findings include sea-fan neovascularization, vitreous hemorrhages, and retinal detachments. Other associated findings include venous tortuosity, retinal holes, Central retina artery occlusion, and angioid streaks.

Treatment consists of pan retina laser photocoagulation (PRP) or cryotherapy applied to the peripheral areas of retinal ischemia, usually anterior to the sea fans. Treatment will lead to involution of the new vessels due to blockage of their feeder vessels. Recently, antiVEGF agents have proven to be useful in the treatment of sickle retinopathy. They block VEGF thereby causing involution of sea fans, helped

cleared vitreous hemorrhage, thus allowing adequate laser treatment. Vitrectomy is reserved for non clearing vitreous hemorrhage and tractional retina detachment. The present study describes the pattern of presentation of sickle retinopathy in Ibadan.

Methods

Thirty three case notes of patients with sickle retinopathy seen at the retinal clinic of the department of ophthalmology, University College Hospital, Ibadan over the last 3 years (2008-2010) were retrieved. Pattern of presentations were recorded in the study pro forma. Results were analyzed using proportions and percentages.

Results

Thirty three (33) cases of HbSC and 14 cases of HbSS were seen. Age Sex distribution of patients with HbSC is shown in table 1. Two (2 out of 14) cases of HbSS presented with central retinal artery occlusion, while the remaining twelve (12) showed retinal vascular dilation and tortuosity. Treatment offered included Laser/Cryotherapy ±

Sex/Age	21-30	31-40	41-50	>50	Total (%)
Male	9	8	4	4	25 (76%)
Female	1	5	1	1	8 (24%)
Total	10	13	5	5	33 (100%)

Table 1: Age/Sex distribution of patients with SC retinopathy.

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intravitreal AntiVEGF Bevacizumab in 23 (70%). Vitrectomy was done to clear vitreous hemorrhage in 10 (30%) cases (Tables 1-4).

Discussion

Sickle cell retinopathy is not uncommon in Ibadan. More males present with HbSC retinopathy in the present study. Age of presentation is middle age, but the trend is now lower, about 10% of cases seen are below 30years of age. This is similar to the study from Lagos [4].

HbSC retinopathy account for most presentations with increased ocular morbidity in this study. Almost half of the patients seen were blind at presentation. Retinopathy is uncommon in patients with HbSS, but a few of them presented with retinal artery occlusion while most presented with increased retina vascular tortuosity. A study from Ghana showed that retinopathy is confined to HbSC patients [15]. Ongoing study in children with HBSS showed increased vascular tortuosity (personal communication).

Even though HbSS patients have a larger number of circulating sickled red cells, their overall lower hematocrit may provide relative protection from vaso-occlusion in the small-caliber vessels of the retina [16].

An alternative theory proposes that the retinal vascular occlusions in HbSS disease may actually be so complete that total infarction and retinal necrosis occur; with no viable tissue remaining that is capable of initiating an angiogenic VEGF response. In contrast, the occlusions in HbSC disease may be less severe, resulting in chronic ischemia, but less complete infarction, and therefore with continuous secretion of angiogenic substances by the damaged tissues [17].

Some workers argued that HbSS do not live long enough to develop retinopathy when compared to the HbSC.

Most patients in this study presented at the late stage of the disease. Late presentation was reported in earlier studies [3,6]. This is not unconnected with late diagnosis and referral.

Most patients require laser photocoagulation to the peripheral ischemic retina to destroy VEGF producing cells and improve retinal oxygenation. Cryotherapy is reserved for patients with hazy media

Symptoms	No (%)
Sudden visual loss	20 (61.0%)
Floaters	7 (21.0%)
Flashes	2 (6.0%)
Gradual visual loss 1 3	1 (3%)
Others 39	3 (9%)
Total	33 (100%)

Table 2: Presenting symptoms.

Visual Acuity	No of eyes (%)
Normal (>= 6/18)	30 (45%)
Low Vision (<6/18-3/60)	5 (8%)
Blind (<3/60)	31 (47%)
Total	66 (100%)

Table 3: Presenting visual acuity in 66 eyes.

Retinopathy findings	Nos (%)
Vitreous hemorrhage	24 (73%)
Sea fan neovascularization	19 (58%)
Tractional retinal detachment	5 (15%)
Black sunburst	5 (15%)
Complicated cataract	2 (6%)

Note: Some eyes presented with more than one finding

Table 4: Retinopathy findings in patients with sickle retinopathy in Ibadan.

from vitreous hemorrhage preventing adequate laser. Vitrectomy was carried out in patients presenting with non clearing vitreous hemorrhage. The AntiVEGF, bevacizumab is used with increasing frequency in our center. It helps in clearing vitreous hemorrhage, thereby allowing proper laser treatment. Bevacizumab has been used similarly in proliferative diabetic retinopathy [18]. These facilities are necessary in ophthalmic facilities that want to cater for this group of patients.

Screening

Why screen? Sickle retinopathy is a public health problem; it is symptomless in early stages, treatable and preventable. The role of primary physicians in referring patients with hemoglobinopathy for regular ocular exams cannot be overemphasized. All newly diagnosed patients with hemoglobinopathy should have ophthalmic evaluation before irreversible damage occur.

Sickle retinopathy is a cause of avoidable blindness in Ibadan, greater communications between primary physicians and ophthalmologists will aid in identifying those patients at risk of developing visual loss and therefore urgent candidates for the available treatments.

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