AN UNUSUAL CASE OF PROLIFERATIVE SICKLE CELL RETINOPATHY

Case Report

By: *C Tembo, D Kasongole

1Department of Surgery, School of Medicine, University of Zambia, Lusaka-Zambia
2University Teaching Hospitals - Eye Hospital, Lusaka-Zambia
*E-mail Addresses: Chimozi Tembo: chimozitembo@yahoo.com

ABSTRACT

Sickle cell haemoglobinopathies are a group of inherited disorders characterized by quantitative or qualitative malformations of haemoglobin (Hb). Diagnosis of SCD is mainly by haemoglobin electrophoresis. Ocular manifestations are wide, encompassing anterior segment, non-proliferative and proliferative retinopathy. Proliferative sickle cell retinopathy (PSCR) represents a very serious complication and may result in blindness if not diagnosed and treated early. PSCR rarely occurs in patients with sickle cell trait, most times in association with an underlying systemic condition or ocular trauma. We present an unusual case of a healthy young male with no history of systemic illness who presented with proliferative sickle cell retinopathy in both eyes.

INTRODUCTION

Sickle cell trait is thought of as a benign condition in comparison to Sickle cell disease (SCD). Sickling haemoglobinopathies are caused by one or more abnormal haemoglobins that induce red blood cells to adopt an anomalous shape under conditions of physiological stress such as hypoxia and acidosis, with resultant vascular occlusion [1]. This results in distal tissue ischemia and a host of related systemic and ocular complications. SCD is most common among black Africans, due to its protective effect against malaria. It also is found, with much less frequency, in eastern Mediterranean and Middle East populations. Ocular manifestations of SCD are wide. Ocular manifestations can be noted in the anterior segment and in the posterior segment in the form of non-proliferative and proliferative retinopathy [1]. There is an inverse relationship between the severity of systemic disease and the severity of retinopathy in homozygous SS individuals compared to compound heterozygous SC subjects [2].

We present an unusual case of proliferative sickle cell retinopathy in a young male patient who presented with blurred vision in the left eye for 2 weeks who denied any history of sickle cell disease.

CASE SCENARIO

A 41-year-old male patient presented to UTHs - Eye Hospital complaining of blurred vision in the Left Eye (LE) for 2 weeks. He denied any history of trauma or straining.

Past Ocular History revealed that he had been seen two months earlier complaining of loss of vision in the Right Eye (RE) for 3 days of spontaneous onset and was diagnosed with vitreous haemorrhage of the RE. He had received intravitreal Bevacizumab (Avastin) in the RE and was advised that he needed surgery but was lost to follow-up.

The patient had no history of hypertension, diabetes mellitus, sickle cell disease, TB or retroviral disease. Family history was non-revealing. There was no history of alcohol intake or smoking.

On examination, the general condition was good. There was no pallor, jaundice or cyanosis. Visual acuity was hand motion (HM) and 6/18 not improving with pinhole in the right and left eye, respectively. Intraocular pressure measured with Goldmann applanation was 20 mmHg in the right eye and 18 mmHg in the LE. Slit lamp examination of the anterior segment examination was normal in both eyes.

Table 1: Proliferative Retinopathy

| Stage 1 | Peripheral arteriolar occlusion |
| Stage 2 | Peripheral arteriovenous anastomosis |
| Stage 3 | ‘Sea fan’ neovascularization develops at the edge of perfused retina |
| Stage 4 | Vitreous haemorrhage from the new vessels |
| Stage 5 | Rhegmatogenous retinal detachment caused by a retinal break associated with extensive fibrovascular proliferation |

Fundoscopy of the RE revealed pink disc with a CDR of 0.4. The blood vessels were sclerosed infero-temporally and the macula showed a thick epiretinal membrane (ERM) with retinal folds and old vitreous haemorrhage (VH). Fundoscopy of the LE revealed Pink disc with a CDR 0.4. Vessels were Normal. There was subhyaloid haemorrhage (SHH) and Salmon patch was noted supero-temporally.

At this point, an impression of proliferative sickle cell retinopathy both eyes with epiretinal membrane right eye was made.

The Full Blood Count (FBC)/ Differential Count showed thrombocytopenia while all other parameters were normal. Urea/ Creatinine/LFTs were normal. Fasting blood sugar was within normal limits and
so was the chest x-ray. Peripheral Blood Smear showed red cell morphology of normocytic, normochromic. The white cell morphology was mild leukopenia and no blasts were seen. On platelet morphology, thrombocytopenia was noted on film. The sickling solubility test revealed Heterozygous HbS and Haemoglobin electrophoresis AS.

Fundus Fluorescein Angiography (FFA) was done with arm retina time of 15 seconds. Fovea Avascular Zone (FAZ) appeared to be normal in both eyes. Areas of capillary non-perfusion (CNP) were noted in both eyes with areas of leakage only found in the LE.

Consultation was made to the haematologist in view of the thrombocytopenia and blood film picture. Patient was counselled on the guarded visual prognosis. Pan-retinal laser photocoagulation (PRP) was done for both eyes in two (2) sittings, covering the superior and inferior retina. The patient was counselled and planned for surgery both eyes. He was planned for pars plan vitrectomy (PPV) plus membrane peeling (MP), endo-laser (EL) and fluid-air exchange (FAE). He had surgery done on the RE from elsewhere. Unfortunately, he developed post-operative endophthalmitis. He received intravitreal antibiotics at UTH Eye Hospital. Later he developed Hyphaema in the same eye with raised intraocular pressure. Anterior chamber washout was done. Current status, the RE is blind post endophthalmitis with neovascular glaucoma (NVG) while LE has resolving vitreous haemorrhage (VH) in proliferative sickle cell retinopathy (PSCR).

Figure 1: Fundus photos of the right eye showing vitreous haemorrhage taken in 2014

Figure 2: Fundus photo right eye showing epiretinal membrane and tractional retinal detachment taken 2019

Figure 3: Fundus Fluorescein Angiography showing areas of leakage in the right eye
DISCUSSION
SCD is the most common and the most severe haemoglobinopathy [5]. Though sickle cell disease is prevalent in black Africans, routine sickling test is not done in most Zambian hospitals. The result is that very few sickle cell trait carriers know of their genetic condition.
Sickle retinopathy can have devastating consequences and may lead to severe visual impairment and blindness if left untreated.

This is what was noticed with the case under review. Both eyes ended up being blind. PSCR occurs rarely in patients with sickle cell trait. Most cases occur if there is an associated systemic condition such as diabetes, hypertension or sarcoidosis or if there is history of ocular trauma [1,6]. In this case there was no pointer to any co-morbidity systemic condition. While other blood tests were normal, he was positive for sickle cell trait which was confirmed by haemoglobin electrophoresis.

CONCLUSION
Though rare Proliferative Sickle Cell Retinopathy (PSCR) can occur in patients with sickle cell trait. There is need to elicit precipitating factors for patients with sickle cell trait that present with retinopathy. Both Sickle cell disease patients and those with sickle cell trait need regular ophthalmological examination.

LIST OF REFERENCES
2. Bwalya, W.M. (2014) Ocular manifestations of sickle cell disease at the University Teaching Hospital, Lusaka, Zambia