

VISUAL OUTCOME AFTER LASER PHOTOCOAGULATION FOR STAGE 4 PROLIFERATIVE SICKLE CELL RETINOPATHY

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SUMMARY

Objective: The objectives of this research is to evaluate the visual outcome after peripheral laser photocoagulation for Goldberg stage-4 proliferative sickle cell retinopathy patients seen in Eye Foundation Hospital Lagos between January and December 2002.

Methodology: A hospital-based retrospective case review study was done. The medical records of 10 patients (13 eyes) who were treated with laser photocoagulation for stage 4 proliferative sickle cell retinopathy within a 12 month period were evaluated. World Health Organization (WHO) visual acuity groupings were used for evaluation.

Results: At presentation, 92.3% of the eyes had normal vision while 7.7% had visual impairment. At three months after laser treatment, 84.6% of the eyes had normal vision while 15.4% had visual impairment. At six months after treatment, 76.9% of the eyes had normal vision while 23.1% of eyes were blind due to recurrent uncontrollable vitreous haemorrhage. Out of the 13 eyes which were treated, 7.7% had actually improved from the visual impairment group to the normal vision group.

Conclusion

Peripheral laser photocoagulation is effective in the treatment of selected cases of stage 4 proliferative sickle cell retinopathy; with 76.9% of patients maintaining normal vision 6 months after treatment.

Key words: laser photocoagulation, sickle cell retinopathy, visual outcome

INTRODUCTION

Sickle cell disorder (SCD) is a major multi-systemic disorder in Nigeria. An estimated 2 million Nigerians

have sickle cell disorder and over 25 million are carriers.^{1,2} The homozygous SS genotype is associated with severe systemic complications, while the haemoglobin SC genotype is more commonly associated with ocular complications.³ Both anterior and posterior segments of the eyes can be affected by complications of SCD. Retina complications have been classified into non-proliferative and proliferative.³

Proliferative sickle cell retinopathy (PSR) is the major sight threatening complication in SCD⁴ and may reach frequencies as high as 70% in patients with sickle cell haemoglobin C disease.

Goldberg's classification of PSR into 5 stages is still widely used⁵ in clinical practice today. Stage 1 is characterized by peripheral arteriolar occlusions, leading to arteriovenous anastomosis in stage 2. In stage 3, new vessels arise from the anastomosis; they have a fan-shaped configuration like sea fans (the invertebrate *Gorgonia flabellum*). As the tufts are gradually pulled into the vitreous gel, varying degrees of vitreous haemorrhage occur (stage 4). Stage 5 is characterized by retinal detachment, which may be tractional, rhegmatogenous or both.

Stage 4 is the most variable stage, and often the stage at which visual acuity drops. In our experience most patients present to the hospital at stage 4; some have had initial episodes of a sudden drop in visual acuity from vitreous haemorrhage, which resolved spontaneously. If not well managed, vitreous haemorrhage can lead to irreversible blindness.

In this study, we share our experience of the management of a few patients that presented to our Hospital at stage 4 and were treated with laser photocoagulation.

MATERIALS AND METHODS

A retrospective case review study was done using the medical records of 10 patients (13 eyes) who presented

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and were managed exclusively at the Eye Foundation Hospital within a 12-month period. All patients had both anterior and posterior segment evaluation, with dilated funduscopy. Five patients had fundus photograph documentation, while two patients had fundus fluorescein angiography.

PSR changes were classified according to Goldberg's classification. This cohort of patients had PSR changes at stage 4 with minimal localized vitreous haemorrhage, or previous episodes of vitreous haemorrhage that had resolved spontaneously. Laser photocoagulation was applied to the affected quadrants and done in different sessions usually 2 to 4 weeks apart (depending on each patient's requirement); 1, 2 or 3 sessions were done. Each session involved applying between 250 and 750 pulses of Argon 532 laser photocoagulation burns to the affected quadrant or quadrants. When more than two quadrants were affected, all quadrants were treated. Sea fans were not treated directly, neither were feeder vessels.

Visual acuity classification was done with the WHO classification as follows:

- Group
- 1 $\geq 6/18$ = normal vision
 - 2 $< 6/18 - 6/60$ = visual impairment
 - 3 $< 6/60 - 3/60$ = severe visual impairment
 - 4 $< 3/60$ to light perception (LP) = blind
 - 5 No perception of light = NPL

Post-treatment evaluation was done with dilated funduscopy and bilateral indirect ophthalmoscopy, or Goldmann's 3-mirror lens and other routine anterior segment examinations.

RESULTS

The study cohort comprised 6 men and 4 women. Their age range was between 29 and 53 years. The average age of the female patient was 40.75 years, compared to 42.16 years in male patients. The genotype SC patients (8) accounted for 80% of patients treated, while AS (1) and SS (1) were 10% each.

At presentation 92.3% of the eyes (12 eyes) had normal vision, while 7.7% (1 eye) had visual impairment in group 2 (fig. 1).

At three months after laser photocoagulation, 84.6% of the eyes (11 eyes) had normal vision while 15.4% (2 eyes) had visual impairment. One eye had a minor episode of vitreous haemorrhage causing visual acuity to drop to group 2 (fig. 2).

At six months after treatment, 76.9% (10 eyes) had normal vision, while 23.1% (3 eyes) had become blind from recurrent vitreous haemorrhage. One of the eyes in group 2 (visual impairment) at 3 months actually improved to group 1 (normal vision). Two eyes from group 1 dropped to group 3 (fig. 3). These patients were advised to have 3 port pars plana vitrectomy.

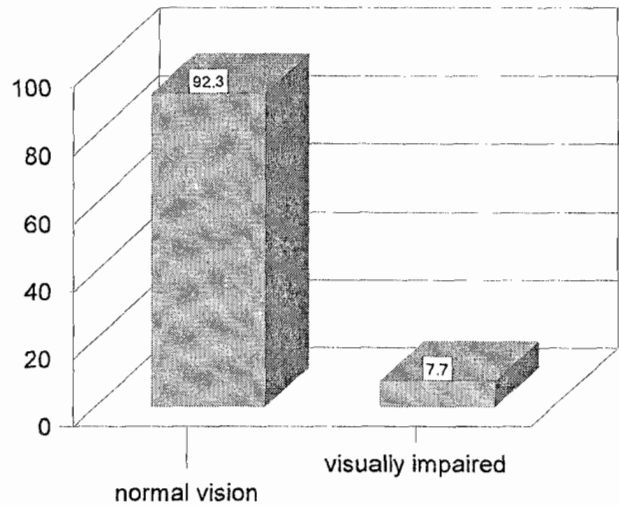


Figure 1. Visual assessment of 13 eyes at presentation

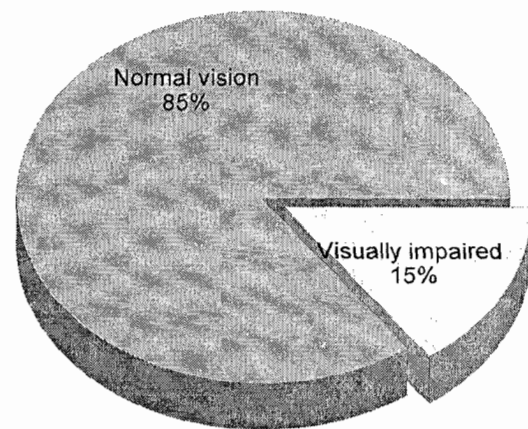


Figure 2. Visual acuity of study group 3 months after laser treatment

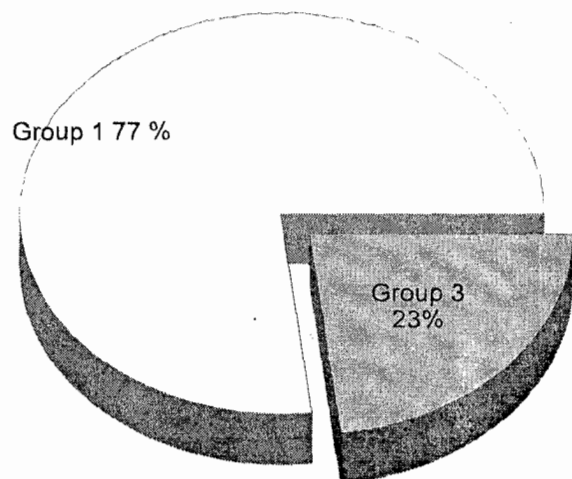


Figure 3. Visual acuity groups, 6 months after laser treatment

Laser treatment was successful in 76.9% of the eyes (10 eyes) at 6 months as 69.2% (9 eyes) maintained normal vision while 7.7% (1 eye) improved from visual impairment to normal vision.

DISCUSSION

Proliferative sickle cell retinopathy (PSR) at stage 4 is one of the major causes of blindness in sickle cell patients⁶ and when not well managed can lead to irreversible blindness. This stage is the most variable of the stages of PSR; patients may present with relatively mild vitreous haemorrhage and with good visual acuity, or very dense vitreous haemorrhage with very poor acuity. The sea fans responsible for this haemorrhage could be single elevated fans or have multiple feeder vessels. A study to evaluate the frequency of blindness (ability to count fingers or less) in sickle cell patients found a frequency of 4% in the entire sickle cell population and about 6% among haemoglobin SC patients although bilateral blindness was rare. If this situation is true in our environment, then about 1 million Nigerians are presently blind in one or both eyes from PSR.⁷

The challenges that the management of stage 4 PSR therefore presents are enormous. Various treatment options exist – laser photocoagulation, diathermy, cryopexy and posterior vitrectomy – in cases where dense vitreous haemorrhage preclude a view of the retina. Laser photocoagulation is preferred by many surgeons as it is associated with less pigment dispersion; it is easier to apply and less painful to the patient. In the past, laser photocoagulation was applied to close feeder vessels of the neovascular foci, but this was found to be associated with the complications of retinal and choroidal haemorrhages, breaks in Bruch's membranes and choroidal neovascularization with extension into the

vitreous, retinal breaks, and retinal detachment.⁸

In this study, we applied peripheral laser photocoagulation to adjacent retina in the quadrant(s) affected. Sea fans were not treated directly, neither were feeder vessels. Other investigators that have advocated similar treatment protocols have recorded equally good results.⁹⁻¹¹

The age range of patients in this study (29-53 years) corresponds to expectations that it takes about a decade to progress through each stage of PSR.¹² Hence most patients with PSR present during their active working life, when visual loss has greater implications. PSR is one of the earliest presenting proliferative retinopathies. As expected, patients were predominantly of the SC genotype (80%) further emphasizing the fact that ocular complications of sickle cell retinopathy are more common in these patients.

One of the difficulties encountered with PSR patients in our environment is late presentation. Most patients still have good visual acuity even with PSR changes at stage 3 and early stage 4, hence, they are unaware of the impending danger. In our study, 92.3% of the eyes treated still had normal vision, although they already had variable degrees of vitreous haemorrhage. The young man whose fundus photograph is shown in figure 4 only presented because he noticed a recent onset of floaters. His vision had not changed yet. However, once the neovascular foci begin to bleed, vision can drop rapidly, and if not treated, will lead to irreversible blindness.

The likelihood of recurrent haemorrhages is emphasized in our study by two patients whose vision dropped to group 3 vision from group 1, due to recurrent vitreous haemorrhage. This represents 23.1% of the cohort, and indicates the considerable risk these patients live with; they should, therefore, be well monitored even after laser photocoagulation for recurrences. In some studies, the rate of recurrence was even higher at 47%,¹³ the recurrence could be from newly developing sea fans or from previously treated ones that did not regress completely. An improvement in vision is also possible as seen in one eye (7.7%), at six months.

Fundus fluorescein angiography (FFA) only became routinely available in our hospital during the follow-up period of some of the patients, hence the low number² of patients that were thus investigated. We, however, found FFA useful for differentiating between fully infarcted lesions, auto-infarcted lesions and lesions that were still active. The patient whose fundus photograph

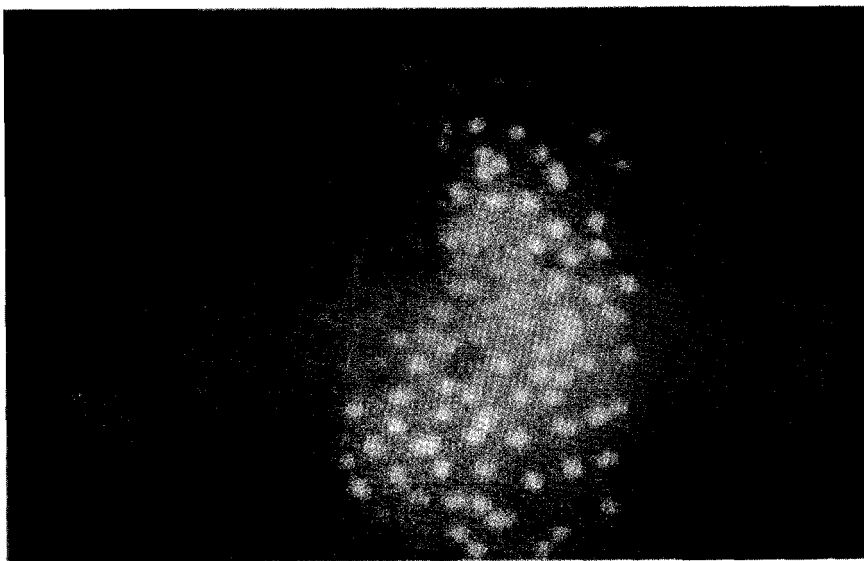


Figure 4. Fundus appearance after laser photocoagulation to peripheral stage 4 sickle cell retinopathy in a 35-year old patient

is shown in figure 5 had lesions that appeared to have infarcted (black arrow), but FFA revealed extensive hyper-flourescence from dye leakage. FFA is now routinely done for all PSR patients to be treated with laser photocoagulation at the Eye Foundation Hospital.

We believe the treatment outcomes would be better if patients presented earlier. Photocoagulation is better done at stage 3, when the lesions are flat on the retina, with no significant vitreous traction.

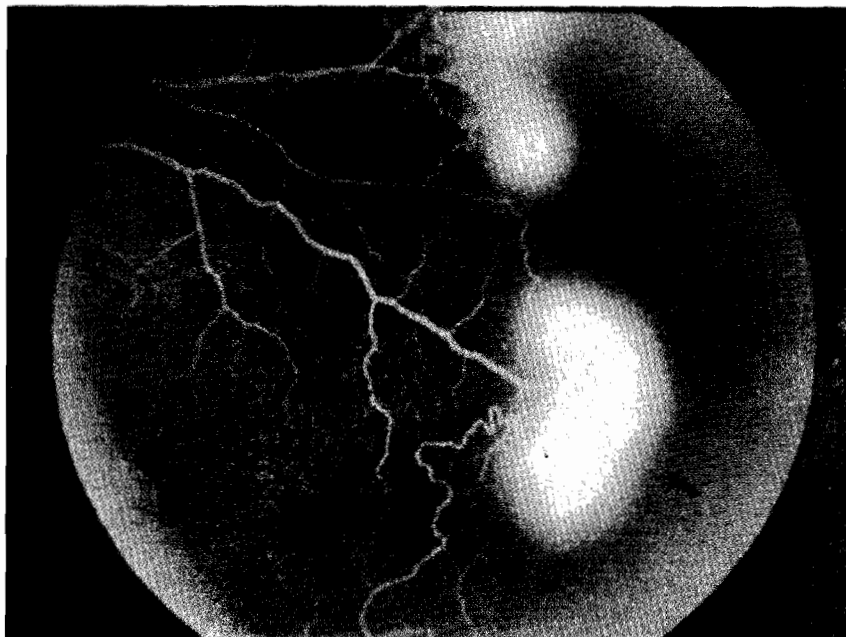


Figure 5. Flourescein angiopathy findings in stage-4 sickle cell retinopathy

A Jamaican study¹⁴ reported that peripheral retinal vessel closure was seen quite commonly in SCD-affected children at 12 years, hence the need to begin screening early. A dilated funduscopy should be done every two years after the 10th birthday for all persons who have sickle cell disorder – HbSC, AS and SS – to ensure early detection and prompt management.

To prevent unnecessary vision loss to an estimated 25 million sickle cell patients in Nigeria it is imperative that more information on PSR is routinely presented. Paediatricians, haematologists, physicians, and obstetricians need to be further enlightened about the ocular complications of PSR so they can send people with sickle cell disorder for routine evaluation.

CONCLUSION

In this study, peripheral laser photocoagulation was shown to be effective in the management of selected cases of stage 4 proliferative sickle cell retinopathy, with 76.9%

of patients maintaining normal vision 6 months after treatment.

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