

Case Report

Bilateral Simultaneous Macular Infarction with Spontaneous Visual Recovery in Genotype SS Hemoglobinopathy Patient

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ABSTRACT

To report the rare and dramatic event of bilateral macular infarction in a sickle cell hemoglobinopathy (SS genotype) patient, resulting in bilateral severe reduction in visual acuity. Without any intervention, the patient's vision gradually improved over the follow-up period. Central visual field defects however persisted. A 21-year-old male Nigerian, presented with a 1-week history of bilateral sudden painless loss of vision. His symptom was associated with fever, feeling of heaviness in the chest and head, and a dizzy spell. Visual acuity was reduced to 20/200 in both eyes and near acuity was; right eye: N24, left eye: N36. Funduscopy showed a pale, milky white, thickened retinal patch superotemporal to the fovea in both eyes. Fluorescein Angiograph: revealed features consistent with occlusion of the parafoveal terminal arterioles in both eyes. Although he did not receive any ocular treatment, and exchange blood transfusion was not done, he regained near-normal visual acuity in both eyes over a 17-month follow-up period, central visual field defects persisted in both eyes. Visual recovery in this patient demonstrates that macular function could improve over time following macular ischemia, without any treatment. Patients and caring physicians should be aware of this possibility.

KEYWORDS: *Macular infarction, macular ischemia, sickle cell, visual loss*

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INTRODUCTION

The occurrence of vaso-occlusive disease (VOD) is a known feature of sickle cell hemoglobinopathy.^[1,2] VODs can involve the eye and may be associated with sudden severe loss of vision. Sudden loss of visual acuity in an SS Genotype patient may result from vaso-occlusion involving the central retina artery^[2,3] or involving the terminal arterioles near the foveal avascular zone (FAZ).^[2,4] Cases of unilateral and bilateral central retinal artery occlusion (CRAO) and macular arteriolar occlusion have been reported.^[1,3,4] In some case reports, visual loss in sickle cell disease with ocular VODs has been reversible^[3,5] whereas in other reports visual loss was noted to be irreversible.^[4]

Furthermore, there have been only few published reports of bilateral simultaneous macular infarction occurring in a sickle cell disease patient.^[6,7]

Preservation of central acuity, despite lack of normal macular perfusion, has been well described in cases

of ischemic diabetic maculopathy, hypertensive maculopathy, and even in densely diffuse macular infarction arising from sickling.^[2] Mechanism for this is not well understood.

We report this rare case and further illustrate that macular function can improve over time bilaterally following a bilateral sudden vascular insult to the macular. However, central visual fields may not reflect the improvement in visual acuity.

CASE REPORT

A 21-year-old male of African descent diagnosed by a pediatrician since childhood (after genotype testing) to have a hemoglobinopathy SS disease, presented with a

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Figure 1: Fundus picture of the right eye showing a parafoveal white patch, which arises as a result of retina infarction with parafoveal arteriolar narrowing

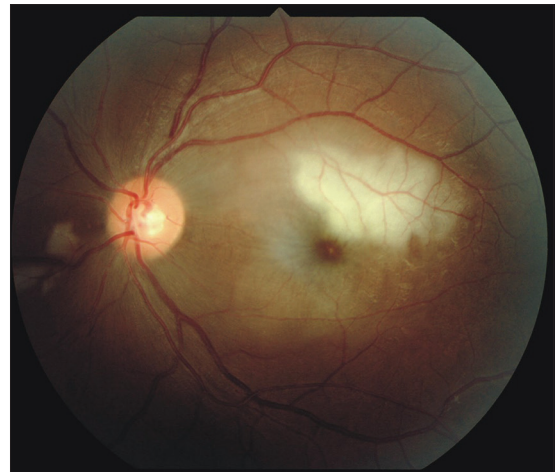


Figure 2: Fundus picture of the left eye showing a parafoveal white patch, which arises as a result of retina infarction with parafoveal arteriolar narrowing

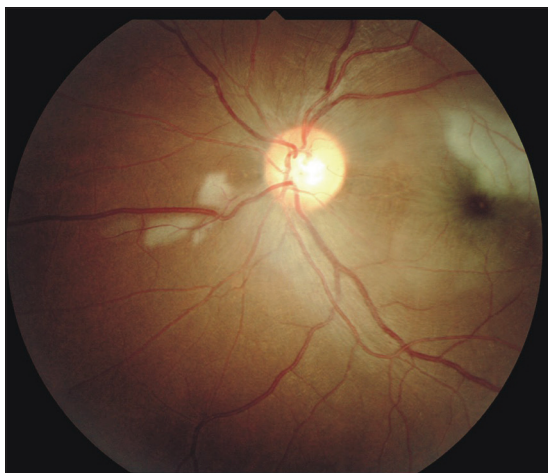


Figure 3: Fundus picture of the left eye also showing area of white patch nasal to the disc with arteriolar narrowing

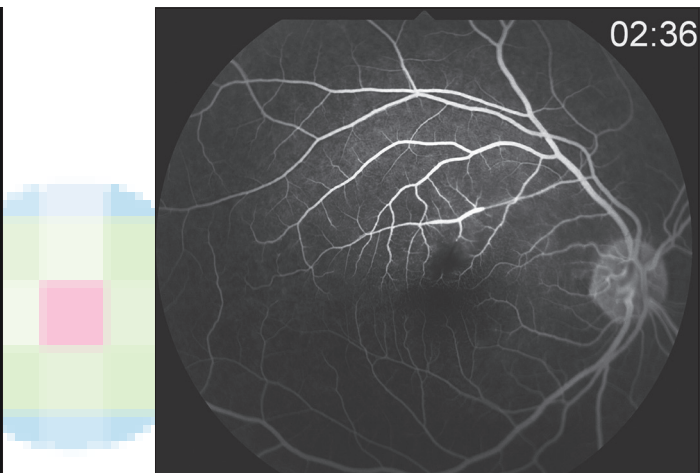


Figure 4: Right eye fluorescein angiogram showing area of arteriolar nonperfusion in the parafoveal area within the area of retina infarction

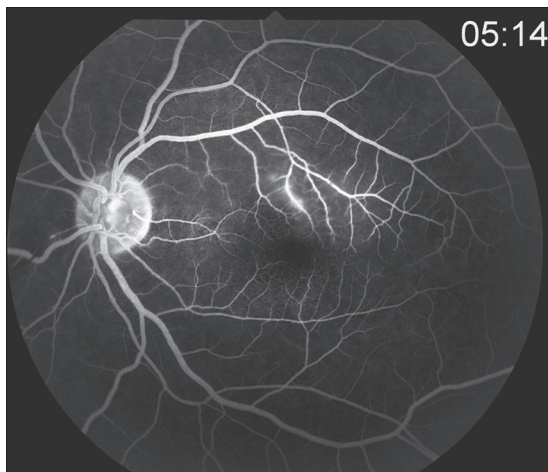


Figure 5: Left eye fluorescein angiogram showing area of arteriolar nonperfusion in the parafoveal area within the area of retina infarction

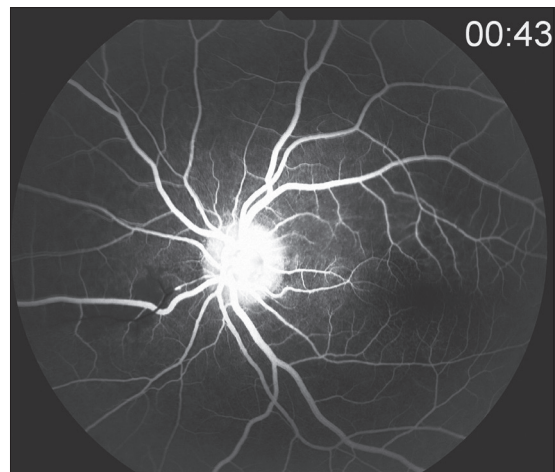


Figure 6: Left eye fluorescein angiogram showing area of arteriolar nonperfusion nasal to the disc

1-week history of sudden severe painless loss of vision in both eyes. This was associated with a systemic illness

characterized by feverish feeling, heaviness of the chest and head, and a dizzy spell.

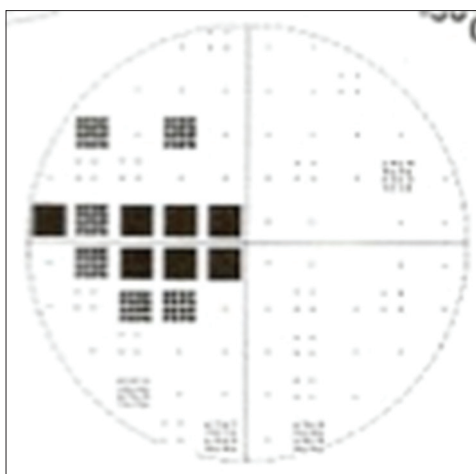


Figure 7: Right eye central visual field showing a nasal defect

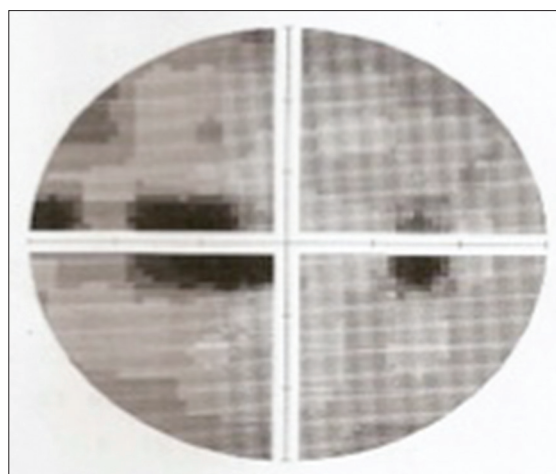


Figure 8: Right eye central visual field showing a nasal defect (Grey Scale)

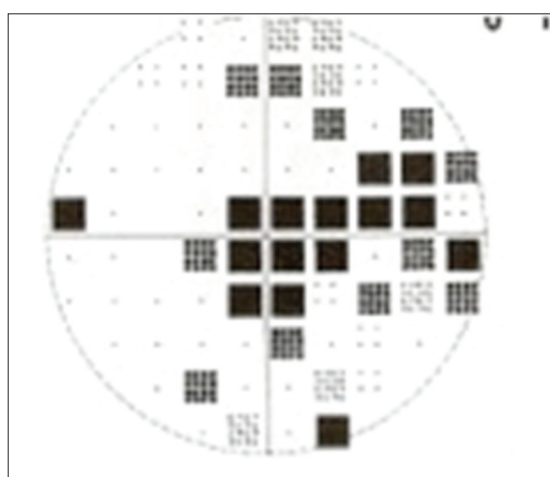


Figure 9: Left eye central visual field showing a cecocentral defect

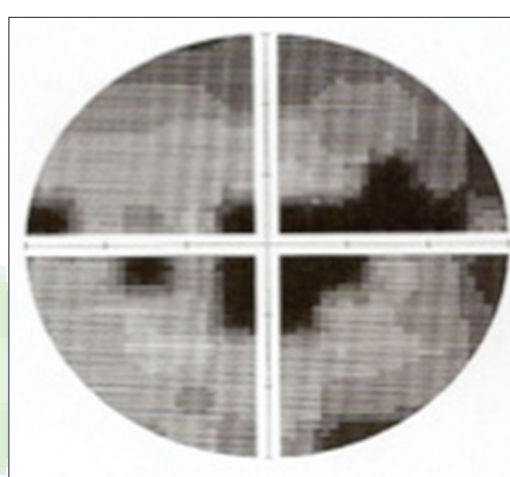


Figure 10: Left eye central visual field showing a cecocentral defect (Grey Scale)

There had been no previous ocular symptoms, and he had been seeing the hematologist for care relating to his genotype and suffered an occasional vaso-occlusive crisis, otherwise, he had no other known problems.

His presenting best-corrected visual acuity was 20/200 in both eyes and near visual acuity was OD: N24 and OS: N36. Anterior segment examination findings were entirely within normal limits.

Main findings were seen on dilated funduscopy using an indirect ophthalmoscope. This revealed a pale, milky white, thickened retinal patch superotemporal to the fovea in both eyes as seen in Figures 1 and 2. A similar but smaller area of white patch was also present a disc diameter from the nasal edge of the disc, along the inferonasal arcade in the left eye as seen in Figure 3.

A fundus fluorescein angiogram (FFA) showed bilateral parafoveal occlusion with an absence of dye transit in terminal arterioles close to the FAZ, and within the opaque areas (also involving the area nasal to the disc in the left eye). This is depicted in Figures 4–6. As history

of the current illness was at this time a week old, it was our impression that exchange blood transfusion may not offer benefit to outcome. There was, therefore, no attempt at exchange blood transfusion nor was any treatment given for his ocular symptoms. Nonetheless, his vision in both eyes improved steadily as he was followed. At 17-month follow-up, visual acuity had improved to 20/20 and N5 in both eyes. At this visit and with the visual acuity improved, he had a visual field assessment in which a central 24–2 threshold test was performed using an AP-5000C (Kowa model) visual field analyzer. His improvement in visual acuity occurred despite the presence of significant visual field defects and a repeat FFA still showing areas of nonperfusion of parafoveal arteriolar network in both eyes. His bilateral visual field defects are seen in Figures 7-10.

DISCUSSION

The posterior segment findings in sickle cell disease can be broadly divided into proliferative and nonproliferative

changes. In a study in central Nigerian, Nigerians with sickle cell disease were reported to have nonproliferative sickle cell retinopathy and proliferative sickle cell retinopathy in 21% and 5%, respectively, of the 90 reviewed patients.^[8] The proliferative changes are a cause of vision loss through the formation of sea fan neovascularization at the junction of the nonperfused and perfused retina, resulting in vitreous hemorrhage and tractional retina detachment. The mechanism for this neovascularization is peripheral vasocclusion resulting in areas of peripheral retina ischemia and release of vasoproliferative substances^[9] which promote the formation of neovascularization. Neovascularization appears to be more commonly seen in Hb SC and less common in Hb SS. Although peripheral vasocclusion and neovascularization has been commonly reported in several Nigerian studies,^[10-13] vasocclusion of the macular and more posterior vessels has not been so described.

The spontaneous vaso-occlusive disease is a known complication of sickle cell hemoglobinopathies and has been reported in association with SS genotype patients.^[2] CRAO, retinal arteriolar occlusion, and occlusion of the terminal macular vessels resulting in macular infarction have been well reported and described. Macular infarction can be unilateral as well as bilateral. Both unilateral and bilateral cases have been reported to respond to treatment with hydration and blood transfusion with some degree of success.^[1,3] The bilateral disease is often associated with significant reduction in vision, which may be permanent. There have been reports of visual improvement, which have occurred following treatment with hemodilution using exchange blood transfusion.^[1,3]

In the patient presented in this case report, steady improvement in vision occurred over a 17-month period, and he did regain a vision of 20/20 in both eyes and could read N5 slowly at near in both eyes without any therapeutic intervention. There have been similar reports of spontaneous improvement in vision following retinal VOD in sickle cell disease patients without any therapeutic intervention.^[2,7]

He was however noted to have binasal visual field defects, which still persisted as at his last follow-up visit. The visual field defects were nasal in the right eye and nasal with central extension (cecocentral) in the left eye. The larger size of the field defect in the left eye may be related to a larger area of infarction in this eye in comparison to that in the right eye.

It is well recognized that despite obvious areas of macular nonperfusion, sickle cell disease patients may have normal central acuity.^[2] Although the visual recovery in this patient demonstrates that macular function could improve over time following macular

ischemia and or macular infarction, visual field defects do persist. A similar finding of binasal visual field defects following simultaneous bilateral retinal infarctions has been reported previously.^[7] Therefore, visual field testing could be helpful, in serving as a useful tool when seeking to determine the previous occurrence of healed macular infarction.

A visual field test is simple and inexpensive and can be used as an adjunct to the optical coherence tomography scan^[7] which has been proposed for the measurement of retinal thinning to document retinal infarction and its repair in patients with a known VOD such as in sickle cell.

CONCLUSION

A central visual field test should, therefore, be part of the standard workup in a sickle cell disease patient in who previous macular infarction and repair is been suspected.

This case report demonstrates that following macular infarction in a sickle cell hemoglobinopathy patient, near-normal vision can be reestablished over time. This can occur even in bilateral cases without any exchange blood transfusion or alternative treatment. This information is important for the patient and the treating ophthalmologist who is confronted with such a case.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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