

## Unusual presentation of ocular trauma in sickle cell trait

Nidhi Pandey

Sickle cell trait is usually considered as a benign condition. However under certain adverse circumstances, it can give rise to vaso-occlusive features as in sickle cell disease. We present here two cases, both involving healthy young males, who developed retinal vaso-occlusive features following blunt ocular trauma. There was a rapid progression of the retinopathy with the development of proliferative changes in both patients and also vitreous hemorrhage in one patient, within 2 months of the trauma. The development of retinopathy was independent of raised intraocular pressure. Both patients were found to have sickle cell trait.

**Key words:** Proliferative sickle retinopathy, raised intraocular pressure, sickle cell trait, trauma, vaso-occlusion

Sickle cell trait is usually considered as a benign condition, as the complications are uncommon and mild.<sup>[1,2]</sup> However, under certain circumstances, such as hypoxia, acidosis, and dehydration, traits can be transformed into a syndrome resembling sickle cell disease with vaso-occlusive features, resulting in great morbidity and mortality. We present here two cases of unilateral proliferative sickle cell, retinopathy developing after ocular trauma in healthy young males having sickle cell trait.

### Case Reports

#### Case 1

An 18-year-old, healthy male presented with complaints of diminished vision in the right eye (RE) following trauma to RE and chest, 15 days back due to fall from height. A few hours after sustaining trauma, he noticed diminished vision in RE, though no ophthalmic consultation was sought. The vision is gradually improved spontaneously by the time he presented to us 15 days following the trauma.

On examination, RE had 15° exotropia; best corrected visual acuity on presentation was 6/24. Anterior chamber showed 2+ cells and flare. Intraocular pressure (IOP) was normal. Left eye was normal with normal IOP. Fundus examination

showed normal disc and macula with multiple intraretinal, and superficial retinal hemorrhages all around the posterior pole, more marked on the temporal side. Underlying retinal edema was also visible [Fig. 1]. Vessels beyond the area of hemorrhages appeared normal. However, fluorescein angiography was done on the same day, showed an abrupt cessation of fluorescein entry in the retinal vessels beyond the areas of retinal hemorrhages [Fig. 2]. The hyperfluorescent columns were visible in some arteries at the junction of perfused and non perfused retina [Fig. 3]. Blood examination including coagulation profile was normal. No systemic abnormality was noted. The patient was started on topical and systemic steroid therapy in tapering doses.

On subsequent follow-up, after 7 days, the vessels beyond the areas of hemorrhages showed signs of getting sclerosed and developed typical silver wired appearance over a course of 10 days [Fig. 4]. The central vision continued to improve and was 6/6 on the last follow-up. The IOP remained normal. Silver wiring of the vessels was present all around in the peripheral retina, more marked and more posteriorly placed in the temporal retina. Fluorescein angiography repeated 3 months after the injury revealed a typical sea fan pattern of neovascularization, at the junction of perfused and nonperfused retina, temporally [Fig. 5], for which laser photocoagulation was advised. Hemoglobin (Hb) electrophoresis revealed the presence of sickle cell trait (HbAS).

#### Case 2

Twenty-six-year-old male presented with diminished vision in RE since 15 days, following trauma with stone. Visual acuity on the presentation was Hand movements (HM) close to the face. Examination of RE showed circumferential congestion and corneal edema. Anterior chamber showed total hyphema. The IOP was 41 mmHg. Following anterior chamber wash on the subsequent day, IOP returned to normal, and vision was Finger Counting (CF) close to face with a relative afferent pupillary defect. Fundus examination showed a normal disc, attenuation of arteries with areas of localized obstruction circumferentially around 2 Disc Diameters (2DD) around the disc, along with retinal hemorrhages. Beyond the obstruction, the blood vessels were slightly tortuous and filled with dark colored blood. Fluorescein angiography showed abrupt cessation of fluorescein entry in both arteries and veins in all quadrants, starting from around 2DD from the optic disc. Intravenous methylprednisolone was given for 3 days followed by oral prednisolone in tapering doses along with topical steroids. On subsequent follow-ups, IOP remained normal, and the vessels became sclerosed. Hb electrophoresis revealed the presence of sickle cell trait (HbAS).

On follow-up, 2 months after injury, the patient had developed partial optic atrophy, though the vision was

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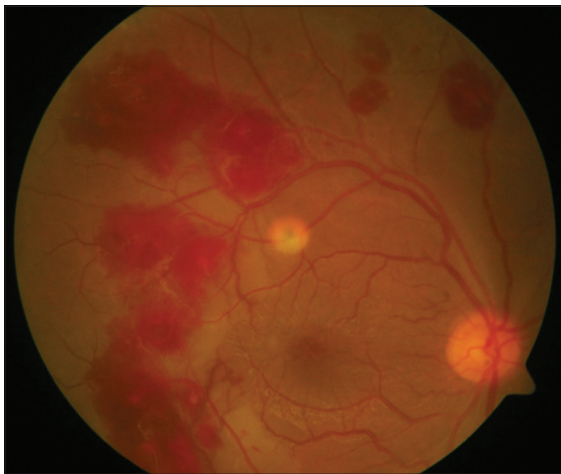
improved to 6/60. There were localized areas of vitreous hemorrhage and areas of arteriovenous communications and Neovascularisation elsewhere (NVEs) [Figs. 6 and 7]. A panretinal photocoagulation was carried out.

## Discussion

Prevalence of sickle cell disease is seen in decreasing order in blacks of Tropical African Ancestry, Mediterranean Basin, Saudi Arabia, Kuwait, Iran, and India. In India, the sickle cell gene is distributed mainly in Madhya Pradesh, Chhattisgarh, Maharashtra, Orissa, Jharkhand, parts of Andhra Pradesh, Kerala, Karnataka, and Tamil Nadu. In some communities, the prevalence of the sickle cell trait is as high as 30%.<sup>[3]</sup> Adult Hb is a globular protein molecule with two pairs of polypeptide chains, globin ' $\alpha$ ' and globin ' $\beta$ ', each folded around a heme molecule. Substitution of glutamic acid with valine at position six of the beta chain, results in sickle Hb, while its substitution with lysine results in HbC. Inheritance of two  $\beta$  S genes results in sickle cell anemia, whereas

inheritance of one S and one normal gene results in sickle cell trait (HbAS).<sup>[4]</sup> Under such conditions of hypoxia, acidosis, oxidative stress, infection, and dehydration the abnormal Hb undergoes a conformational change, changing the erythrocytes into sickle-shaped cells. These cells are rigid and more prone to hemolysis than normal red blood cells. Under such conditions, there is an aggregation of sickled cells, which form small plugs. The available Hb is deoxygenated, and the oxygen demand of the tissues is not fulfilled resulting in tissue acidosis. This causes greater sickling, increased blood viscosity, and decreased blood flow leading to the "vicious cycle of erythrosthesis." While the SS type results in the most severe clinical symptoms, SC genotype is associated with the most severe retinal manifestations.<sup>[5,6]</sup> The AS type is uncommonly associated with retinal changes.<sup>[2]</sup>

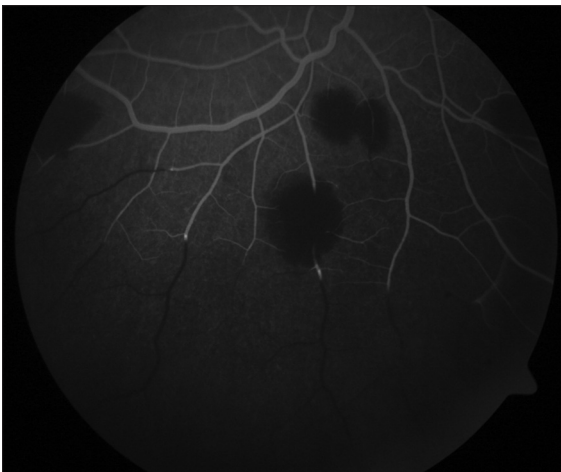
However, patients with sickle cell trait are at a risk of retinopathy if coincident ocular or systemic disease is present.<sup>[7]</sup> Retinal vascular occlusion has been previously reported in patients with sickle cell trait as a complication



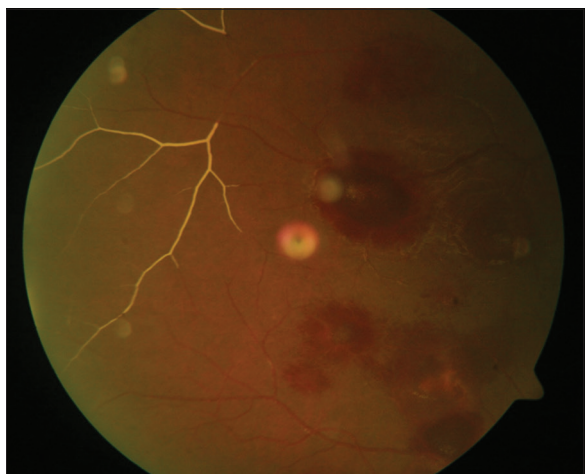
**Figure 1:** Fundus photograph of Case 1, right eye, 15 days after sustaining trauma showing multiple retinal hemorrhages and underlying retinal edema



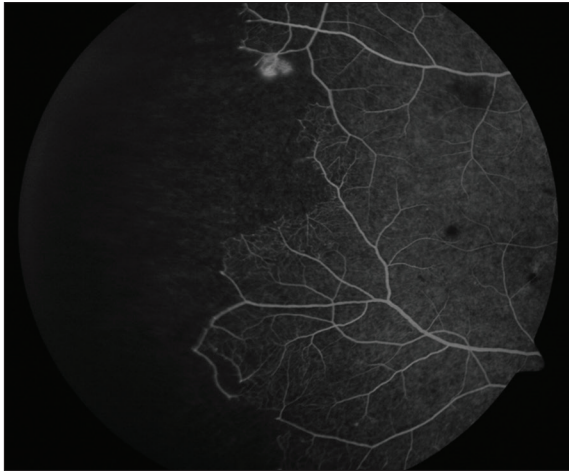
**Figure 2:** Fluorescein angiogram of Case 1, right eye, 15 days after injury showing abrupt cessation of fluorescein entry in vessels beyond the area of hemorrhages



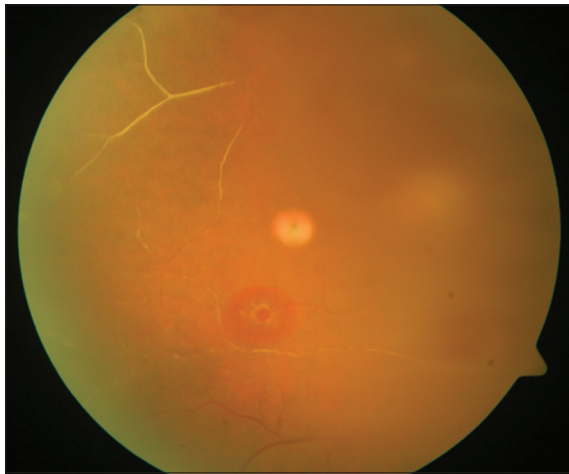
**Figure 3:** Fluorescein angiogram of Case 1, right eye, 15 days after injury showing hyperfluorescent columns at the junction of perfused and nonperfused retina



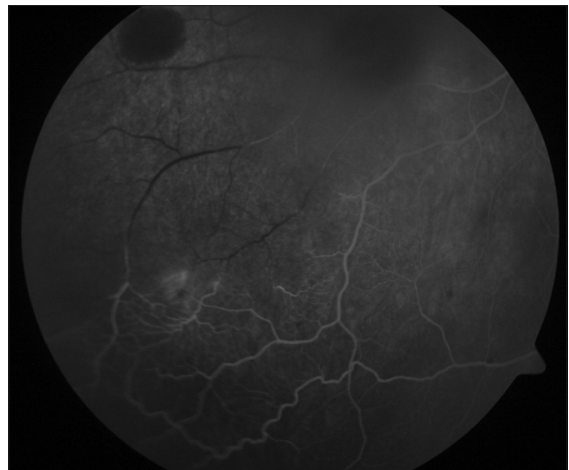
**Figure 4:** Fundus photograph of Case 1, right eye showing silver wiring of vessels



**Figure 5:** Fluorescein angiogram of Case 1, right eye showing sea fan pattern of neovascularization at the junction of perfused and nonperfused retina



**Figure 6:** Fundus picture of Case 2, right eye, 2 months after injury showing vitreous hemorrhage and silver wiring of vessels



**Figure 7:** Fluorescein angiogram of Case 2, right eye, 2 months after injury showing vitreous hemorrhage, arteriovenous communications, and NVE

of traumatic hyphema and glaucoma, following persistently raised IOP secondary to hyphema.<sup>[7,8]</sup> The clinical appearance of our cases suggests a localized retinal pathology, which led to sickling. A localized area of retinal hypoxia due to the impact of trauma may have been the precipitating event. The impact probably led to the transient rapid rise of IOP in a circumferential area around the disc leading to localized retinal edema, and sickling of erythrocytes in that area with resultant stagnation. A sustained rise in IOP is not required to precipitate the vaso-occlusive event, as it is manifested in Case 1 described above, where it occurred in spite of a normal IOP on presentation. The clinical picture in both cases is suggestive of a sickle cell crisis occurring in the retinal vessels. The sickle cell crisis in patients having sickle cell trait may be precipitated by any hypoxic condition either local or systemic. The vascular occlusions following trauma in patients of sickle cell trait may, therefore, be independent of a persistently raised IOP as was previously thought.<sup>[7,8]</sup>

## Conclusion

Sickle cell trait, though considered as a benign condition may lead to sight-threatening complications in the presence of precipitating factors. Impact of blunt trauma may lead to localized hypoxia, promoting sickling of erythrocytes leading to vaso-occlusion and resultant stagnation of blood. This may eventually lead to rapid development of proliferative sickle cell retinopathy.

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## Conflicts of interest

There are no conflicts of interest.

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