



CFEH Facebook Case #1

A 27 year old African female presented to CFEH for further examination of peripheral retinal changes noted at her recent eye test. Optomap central pseudo-colour and autofluorescence images as well as Spectralis OCTs scans are shown below. (The resolution of the OCT images is reduced due to the peripheral nature of these lesions).

What is your diagnosis?



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ANSWER

Sickle-Cell Retinopathy

The area scanned in the right eye and the area marked 2 in the left show peripheral neovascularization with fibrosis. There is a sunburst lesion in the left eye (lesion marked 1). The vitreous opacities seen inferiorly on the right Optomap images are consistent with old vitreous haemorrhages. This patient was subsequently referred to a retinal specialist for management.

Sickle cell retinopathy results from a mutant gene that codes for the amino acids contained in haemoglobin. This causes the red blood cells to deform into a "sickle" shape when oxygen pressure is low. Sickle cell retinopathy is most prevalent in the African American population.

Sunburst lesions are localized areas of retinal pigment hypertrophy, hyperplasia and pigment migration as seen in the OCT. The lesions often appear speculated and are typically found in the peripheral retina in a perivascular location. The "sunburst" lesions usually form secondary to reabsorbed sub-retinal haemorrhages and appear on fundus autofluorescence as an area of hypo-autofluorescence with a hyper-autofluorescent border. They are characteristic of sickle-cell retinopathy.

Decreased visual acuity occurs due to the occlusion of parafoveal capillaries and arterioles, as well as spontaneous occlusion of the central retinal artery in some patients.

This patient also has a second characteristic feature of sickle cell retinopathy – pre-retinal "sea fan" neovascularization. This typically occurs in sickle-cell patients at the posterior border of an area of retinal non-perfusion, and can lead to vitreous haemorrhage and eventually tractional retinal detachment.

Other possible clinical findings typical of sickle cell retinopathy include the segmentation of blood in the blood vessels of the conjunctiva or the small surface vessels of the optic disc.

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