**Title:** “Arcus retinalis”–A novel clinical marker of sub-internal limiting membrane haemorrhage.

**ABSTRACT**

**Introduction**: Preretinal hemorrhages can be located in subhyaloid or sub-internal limiting membrane (ILM) space. Though optical coherence tomography (OCT) may help to distinguish the exact location of haemorrhage in selected cases, it is often difficult to discern clinically. The purpose is to describe a novel clinical sign named “arcus retinalis” as a clinical marker of sub-ILM haemorrhage.

**Methods:** Retrospective observational case series.Ophthalmic records of 31 eyes with sub-ILM haemorrhage were analyzed. The sub-ILM nature of the haemorrhage was confirmed either by the presence of two distinct layers (ILM and posterior hyaloid) on OCT or on intraoperative sub-ILM localization of the haemorrhage during vitrectomy.

**Results:** 15 out of 31 eyes with sub-ILM haemorrhage demonstrated a retinal yellowish-white arc corresponding to the outer margin of the sub-ILM haemorrhage. A complete circle was not seen in most of the cases. This arc corresponded to a vertical peg-like structure in the outer retina on OCT and had a tendency to fade away over months. In 10 eyes, it was visible at presentation and in 5 eyes it could only be seen after clearing of the haemorrhage.

**Conclusion.** Retinal yellowish-white arc (“arcus retinalis”) is a useful clinical marker of sub-ILM haemorrhage with distinct OCT features that tends to disappear with time.

**Key-words:** Arcus retinalis; Internal limiting membrane; Haemorrhage; optical coherence tomography; Fundus photographs

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**Introduction**: A preretinal haemorrhage is a common occurrence in a variety of systemic and ocular conditions including valsalva retinopathy,1 Terson syndrome,2 anaemic or leukemic retinopathy,3 proliferative retinal disorders,4 retinal artery macroaneurysms and trauma.5. It has a characteristic boat shaped appearance and is sharply demarcated. When premacular, this condition is often alarming to the patient as the visual decline is severe and sudden. The exact location of preretinal haemorrhage has been debated in literature and it may be located either in the subhyaloid space or in the sub internal limiting (ILM) membrane space or both.6

Clinically it is difficult to distinguish subhyaloid haemorrhage from sub-ILM haemorrhage.7 The presence of a glistening light reflex may point towards a sub-ILM haemorrhage.7 Optical coherence tomography (OCT) is a useful tool to confirm sub-ILM haemorrhage if two layers (posterior hyaloid and ILM) can be demonstrated in front of the haemorrhage.8 No other indicators exist till date to distinguish the location of these hemorrhages.

While most preretinal hemorrhages resolve spontaneously over a period of time, long standing haemorrhage may lead to toxic damage to the underlying retina due to hemoglobin and its catabolites.2,9 The ILM provides an important barrier to these and has a protective effect towards the retina.10 It may thus be prudent to intervene earlier in cases of premacular sub-ILM haemorrhage, to prevent this toxic damage that may result in irreversible visual loss. This makes it important to decipher the location of these hemorrhages. We describe a retinal yellowish white arc, named herein as “arcus retinalis” as a novel clinical marker of sub-ILM haemorrhage.

**Methods:** This was a retrospective observational case series of consecutive patients with sub-ILM haemorrhage encountered from the period 2008 to 2017. The study adhered to the tenets of declarations of Helsinki and to the institutional research guidelines. Informed consent was obtained from all the patients.

The medical records of all patients with preretinal haemorrhage were analyzed and the patients diagnosed as sub-ILM haemorrhage irrespective of the etiology were included in the study. The diagnosis of sub-ILM haemorrhage was confirmed by the presence of two distinct membranes in front of the premacular haemorrhage on OCT or intraoperative localization of haemorrhage in the sub-ILM space (in eyes which underwent vitrectomy for clearing the haemorrhage). The records of the patients were reviewed for demographic data, history to elucidate the cause and duration of the haemorrhage, best corrected (Snellen) visual acuity (BCVA), colour fundus photographs, OCT and management details. The OCT scans had been performed either on spectral domain (RTvue, Optovue Inc or Cirrus, Zeiss Inc) or swept source platform (Triton, Topcon Inc).

**Results:** A total of 31 eyes of 28 patients with sub-ILM hemorrhage were identified. Three patients had bilateral sub-ILM haemorrhage at presentation. Mean age of the patients was 27.4 years. There were 16 male and 12 female patients. The cause of the haemorrhage was Valsalva retinopathy in 21 eyes and anemic retinopathy (due to various causes) in 10 eyes. BCVA ranged from light perception to 20/30. 8 eyes were managed conservatively, 20 eyes underwent Nd:YAG laser membranotomy and 3 eyes needed pars plana vitrectomy (PPV) to clear the hemorrhage.

On careful examination of colour fundus photographs, it was seen that 15 eyes of 13 patients demonstrated the presence of a retinal yellowish white arc (termed “arcus retinalis”) at the outer edge of sub-ILM haemorrhage. The mean age of this subgroup of patients was 27.6 years. Six out of these 13 patients were females. 4 eyes had sub-ILM hemorrhage due to anemia (two due to Dengue, one due to malaria and one due to aplastic anemia) while the remaining eleven had Valsalva retinopathy. The duration of the sub-ILM haemorrhage ranged from 2-6 weeks. 13 out of 15 eyes were confirmed to have sub-ILM hemorrhage on OCT while in two the location was determined intra-operatively during PPV. Nine eyes underwent Nd:YAG laser membranotomy; two eyes underwent vitrectomy while four eyes were kept on observation.

The arcus retinalis was present at the outer margin of sub-ILM haemorrhage for a variable degree (Figure 1a, b). It was incomplete in most of the cases with no case having continuous ring for 360 degrees. On OCT this line corresponded to a vertical peg-like structure at the level of outer retina (Figure 1c,d). This point corresponded well to the insertion of ILM at the outer edge of haemorrhage. With the resolution of haemorrhage and subsequently the premacular cavity, arcus retinalis faded away (Figure 1e). The peg-like structure on OCT also became smaller (Figure 1f). However these changes persisted till haemorrhage resolved (Figure 1f, h). With the passage of time, complete resolution of the arcus retinalis as well as the peg-like structure on OCT could be seen (Figure 2). Similar changes were observed in extra foveal hemorrhages as well (Figure 3).

Ten eyes were identified to have arcus retinalis at presentation, while in five eyes it could be picked up only after clearing of the haemorrhage (Figure 4, 5). Out of these five eyes, two had undergone PPV (one for non resolving vitreous haemorrhage and one for persistent premacular haemorrhage) while three eyes had been treated with Nd:YAG laser membranotomy for clearing the sub-ILM hemorrhage. **In one out of these three eyes, the arcus retinalis was very faintly visible at the superior border before laser membranotomy (Figure 5).** In eyes kept on observation, arcus retinalis faded away with time as the haemorrhage resolved (Figure 6). The rest of eyes with arcus retinalis are presented in supplemental figure 1.

**Discussion: The** clinical picture may be remarkably similar in patients with sub-ILM and sub-hyaloid hemorrhages. Since the ILM may have a protective effect on the retina from blood and its degradation products,10,11 it may be prudent to intervene earlier in cases with sub-ILM haemorrhage. Therefore it becomes important to discern the exact location of these preretinal hemorrhages. A glistening light reflex on the surface of the haemorrhage may be the only clinical indicator of sub-ILM haemorrhage. OCT may be useful, especially if two separate layers depicting the ILM and the posterior hyaloid can be deciphered in front of the haemorrhage.

We describe a novel clinical sign pointing towards a sub-ILM haemorrhage and its OCT characteristics. In 50% of the eyes with sub-ILM haemorrhage in our study we could find a yellowish white retinal arc (“arcus retinalis”). The arc was almost never a complete ring and extended for a variable distance around the haemorrhage. On OCT, a vertical peg like structure was seen corresponding to the arcus retinalis and coincided well to the point, where the ILM was lifted off the retinal surface. We did not encounter findings similar to arcus retinalis in cases of sub-hyaloid haemorrhage.

It is our contention that these peg-like structures on OCT, which are seen clinically as arcus retinalis, are the result of localized traction on the retina by the stretched ILM. With the passage of time as the pre-retinal cavity collapses and traction by the ILM decreases, the arcus retinalis tends to disappear (Figure 1,2). Exudates or hemorrhage in the outer retina derived from the sub-ILM hemorrhage may be another possibility in the causation of arcus retinalis as traction due to ILM was not visible in all the cases (Figure 3,4). The presence of ellipsoid zone (EZ) disruption near the arcus retinalis indicates its possible composition by photoreceptor components. It is important to distinguish arcus retinalis from a ragged whitish yellowish arc formed by residual altered blood that is seen frequently at the inferior edge of the sub-ILM cavity. On OCT this is seen as hyperreflective material just beneath the ILM, where it is attached to the retina (Figure 7).

Our study is limited by its retrospective and non-comparative nature and small sample size. Further, arcus retinalis was not visible in all the cases of sub-ILM hemorrhage. **This may be due to insufficient forces created by a small or a recent sub-ILM hemorrhage though we did not correlate its presence with the size/extent or duration of hemorrhage.**

To conclude we describe a previously unreported clinical indicator of sub-ILM haemorrhage named herein as arcus retinalis, the corresponding OCT features of which include a vertical peg-like structure. This arcus retinalis tends to disappear with time. Together with OCT, this clinical marker may help to segregate the cases of sub-ILM hemorrhage from subhyaloid hemorrhage that may need earlier intervention. It may also contribute to further understanding the pathophysiology of sub-ILM haemorrhages.

**The Author(s) declare(s) that there is no conflict of interest.**

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**Figure Legends:**

**Figure 1:** A 20-year-old female with decreased vision both eyes since three weeks following dengue fever. BCVA was 20/40 and 20/60 in the right and left eye respectively. Colour fundus photographs of right (a) and left eye (b) showed preretinal hemorrhages with yellowish white arcs at the outer margin of haemorrhage (arcus retinalis). Swept source OCT showed preretinal haemorrhage in a premacular cavity with peg shaped structures in the outer retina (white arrows) corresponding to the arcus retinalis in both the eyes (c, d). One month later, the haemorrhage resolved in the right eye with BCVA returning to 20/20 (e), the premacular cavity collapsed (g) and the peg in the outer retina became smaller (white arrow, g). The left eye improved to 20/40 with the premacular haemorrhage (f) and cavity (h) decreased in size. The vertical pegs persisted (white arrows, h)

**Figure 2: At** six months follow up, a very faint arcus retinalis was visible in the right eye (a) though the OCT was unremarkable (c). The left eye showed resolution of the haemorrhage and premacular cavity (b, d), fading arcus retinalis (b) and a barely discernible small peg on OCT (white arrow, d). BCVA returned to 20/300. At ten months follow up, the preretinal haemorrhage, cavity and arcus retinalis completely resolved with no residual changes on OCT (e-h). BCVA remained stable at 20/20 in the right eye and 20/30 in the left eye both eyes.

**Figure 3:** A 24-year-old male with decreased vision and scotoma since two weeks following Valsalva retinopathy. BCVA was 20/30. The right eye showed a sub-ILM haemorrhage in the supero-temporal aspect of macula along with arcus retinalis that was more prominent temporally (a). Swept source OCT (e) confirmed a sub-ILM haemorrhage, separate posterior hyaloid and outer retinal peg (black arrow). The haemorrhage and arcus retinalis resolved gradually over 3 months (b), 6 months (c) and 12 months (d).

**Figure 4:** Prominent arcus retinalis is seen on colour (a) and red free photographs (b) of the left eye of a 40-year-old female who underwent vitrectomy for non-resolving sub-ILM haemorrhage **and scattered vitreous hemorrhage** following Valsalva retinopathy. (c) Arcus retinalis in the form of a large ring is also seen in the right eye of a 30-year-old female, who underwent vitrectomy for non-resolving vitreous haemorrhage during which a large sub-ILM haemorrhage was found supero-nasally. (d) Swept source OCT showed vertical peg in the outer retina corresponding to the arcus retinalis.

**Figure 5:** (a) A 22 year old female with premacular haemorrhage due to Valsalva retinopathy underwent Nd:YAG laser hyaloidotomy. **Arcus retinalis is faintly visible at the superior border of the hemorrhage** (b). Two weeks later, when the haemorrhage cleared, arcus retinalis was evident on colour photograph (c). Spectral domain OCT showed a premacular cavity confirming that it was a sub-ILM hemorrhage.

**Figure 6:** A 30-year-old male presented with blurring of vision since 3 weeks following heavy weight lifting. A premacular haemorrhage not obscuring the fovea was seen (a). Arcus retinalis could be made out at the temporal aspect of the haemorrhage. The brownish linear deposits seen nasal to arcus retinalis were that of altered blood and should not be confused with the latter.At 6 weeks (b) and 12 weeks (c) follow up, the haemorrhage decreased in size and the arcus retinalis became smaller as well.

**Figure 7:** Arcus retinalis (blue arrowheads) seen on the temporal aspect of a resolved sub-ILM haemorrhage (a). The blue arrow indicates residual altered blood beneath the ILM while the yellow arrow shows the direction of the OCT scan. (b) Spectral domain OCT scan demonstrates a premacular cavity with hyperreflective collection beneath the ILM (blue arrow) corresponding to the altered blood.

Supplemental digital content 1.tiff: Fundus images of the remaining seven eyes with arcus retinalis