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Letter to the Editor

Terminologies regarding sickle cell retinopathy

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Dear Editor,

We read the article by Beral and associates with great interest. The study showed that that retinopathy and maculopathy are common in sickle cell disease (SCD) and found no association with hematological parameters, blood rheology or genotype [1].

We have some considerations that might help understanding the study.

Retina is a complex intraocular anatomic structure responsible for initiating the visual process [2]. It can be anatomically divided in posterior pole and peripheral retina and, Macula is a 4.5–6 mm area present in the posterior pole [2]. The terms Retinopathy and Maculopathy are general and refer respectively to disease of the retina and disease of the macula.

Since Goldberg's classification of sickle cell retinopathy (SCR) in 1971, it's the gold standard for funduscopic diagnosing and classification of the disease [3, 4]. The first description of the macular ischemic involvement in SCD was in 1972 but traction maculopathy was already described related to proliferative SCR [5]. In other words, maculopathy is part of SCR and a correlation between the peripheral vascular nonperfusion and the macular vessel density was already proven [6].

In their article, Beral and colleagues used the terminology "Retinopathy" and "Maculopathy" as two different diseases and did not describe the diagnostic criteria used neither the classification of both. We understood that they applied "Retinopathy" as the peripheral vascular alterations and "Maculopathy" as the macular ischemic impairment but we are not sure what stage of peripheral retinopathy nor what OCT finding were consider for the diagnosis.

It's also known that fluorescein angiography (FA) can detect a higher stage of peripheral SCR when compared to clinical evaluation [7] and that Optical Coherence Tomography (OCT) is more sensitive than fundus examination in detecting sickle cell maculopathy [8]. Beral and colleagues used a high-sensitive method for detecting macular involvement and a less sensitive method to detect the peripheral disease and, there is no further information about the details of this tests in their article. We would be pleased to read more about the study methodology regarding ophthalmological evaluation.

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We also would like to add that Dell'Arti et al. performed a similar study using OCT technology aiming to identify systemic risk factors for sickle cell maculopathy [9]. They conclude that chronic chelation therapy and, potentially, high levels of HbF were described as possible protective factors for the presence of sickle cell maculopathy [9].

We believe that the considerations highlighted above might enrich the valuable scientific information assembled by Beral and colleagues and hope for reading more from their study.

We celebrate Beral and colleagues for the presentation and offer our respects.

Conflict of interest

The authors have no conflict of interest to declare.

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