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Sickle Cell Retinopathy: Case Report and a Literature Review in Japan

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Abstract **Case Report**

A 48-year-old Peruvian woman had lived in Japan presented with myodesopsia in the right eye. On ophthalmic examination, best-corrected visual acuity was 1.2 in the right eye and 0.5 in the left eye. Funduscopy of both eyes showed avascular area with neovascularization simulating "sea-fan" in the peripheral retina. Erythroblast, target cell, and Howell Jolly body were detected in her peripheral blood smear. We strongly suspected sickle cell retinopathy in this patient. Although sickle cell retinopathy is not seen among Japanese, it is important to consider the possibility of sickle cell retinopathy when a foreign patient presents with myodesopsia and avascular area with neovascularization in the peripheral retina.
Key words: Sickle cell retinopathy.

INTRODUCTION

Sickle cell disease (SCD) is characterized by the production of abnormal hemoglobins that deform and stiffen red blood cells, causing increased blood viscosity and microcirculation occlusion to varying degrees [1-12]. Ocular manifestations of SCD include orbital, conjunctival, uveal, papillary, and especially retinal changes (sickle cell retinopathy; SCR) [3-12]. Vaso-occlusions occur primarily in younger people, and are first observed at the periphery of the retina, resulting in non-perfused and presumably ischemic areas [3-12]. The prevalence of SCD is especially high among African-Americans, while it is not seen among Japanese. Here, we report a Peruvian woman with SCR.

CASE REPORT

A 48-year-old Peruvian woman had lived in Japan presented with myodesopsia in the right eye. On ophthalmic examination, best-corrected visual acuity was 1.2 in the right eye and 0.5 in the left eye. The anterior segment of both eyes was unremarkable. Funduscopy of both eyes showed avascular area with neovascularization simulating "sea-fan" in the peripheral retina (Figure 1).

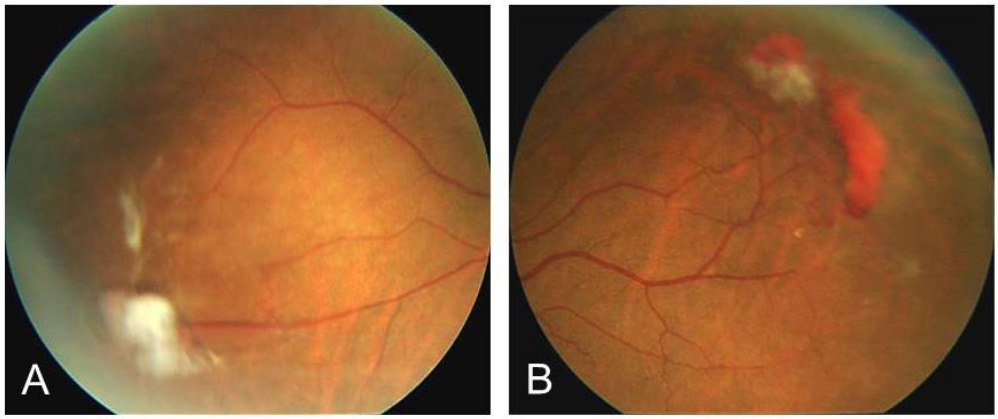

Fig-1: Fundus photographs of the (A) right and (B) left eyes
Note avascular area with neovascularization simulating "sea-fan"

Fluorescein angiography confirmed avascular area and neovascularization (Figure 2).

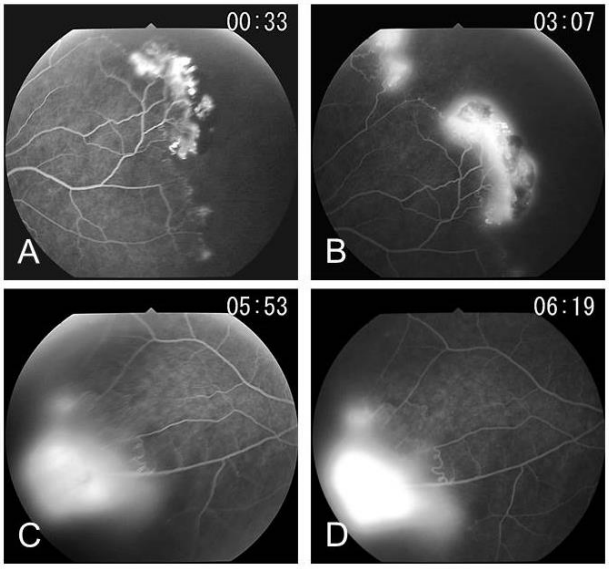

Fig-2: Fluorescein angiography of the (A, B) right and (C, D) left eyes

Note avascular area and neovascularization

In her peripheral blood smear, erythroblast, target cell, and Howell Jolly body were detected. Thus, we strongly suspected SCR in this patient. We requested further examination including hemoglobin electrophoresis but she did not visit. A part of this case report had been previously described in Japanese [13].

DISCUSSION

Whilst SCD was originally a disease that was prevalent in tropical and subtropical regions, forced migration and ongoing population movement has now made it a global problem. According to the review article described by Amissah-Arthur *et al*. [11], there are more than 400,000 children born each year with SCD with 84% of cases originating from sub-Saharan Africa making it one of the most ubiquitous genetic disorders. Estimated birth rates reach 10.68 per 1000 in Africa, 0.68 per 1000 in South and Southeast Asia, 0.49 per 1000 in the Americas and 0.07 per 1000 in Europe. Within continental regions, there is a large variability in the prevalence of sickle cell trait. Although 7% of the world population is carriers of the abnormal sickle gene, it is reported in 1–38% in some parts of Africa, 0–29% in Eastern Mediterranean, 7–30% in India and affects 8% of African descents in the United States of America and the Caribbean conferring an immense disease burden. Thus, the prevalence of SCD is especially high among African-Americans, while it is not seen among Japanese.

SCR was divided by Goldberg [3] in five stages: stage I is characterized by definitive arteriolar occlusion. In stage II, the budding of new vessels begins, with possible dilatation, aiming to join the vascular and avascular retina. In stage III, under the action of angiogenic events, pre-retinal neovascularization occurs, forming the so-called retinal "sea-fan". This facilitates the occurrence of vitreous hemorrhage and characterizes stage IV of proliferative retinopathy in SCD. The repetition of these hemorrhagic phenomena leads to rupture, retinal detachment (stage V) and vision loss, the final stage of SCR.

There are only three case reports [13-15] in Japan including this patient [13]. Shirakami *et al*. [14] reported a 27-year-old Ghanaian woman. Her funduscopic findings corresponded to stage III in the right eye and stage IV in the left eye. Maruhashi *et al*. [15] reported a 45-year-old Ghanaian man. His funduscopic findings corresponded to stage III in the right eye and stage V in the left eye. In this present case, SCR was presented stage III in both eyes.

CONCLUSIONS

Although SCR is not seen among Japanese, it is important to consider the possibility of SCR when a foreign patient presents with myodesopsia and avascular area with neovascularization in the peripheral retina.

Disclosure

The authors have no conflicts of interest to disclose for this paper.

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