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PURPOSE

To report the multimodal evaluation of the retina in an uncommon association of angioid streaks with homozygous sickle cell.

INTRODUCTION

Angioid streaks (ASs) constitute radial peripapillary breaks in a calcified, thickened, and brittle Bruch's membrane (BM). ASs can be idiopathic but more than 50% of patients have a concurrent disease, mainly pseudoxanthoma elasticum. A less common association is sickle cell hemoglobinopathies, which incidence varies between 0.9% and 6%. (1) Considering that we highlight the importance of ophthalmologic examination in patients with hemoglobinopathies.

CASE REPORT

A 51-year-old man diagnosed with sickle cell disease in childhood was referred to the ophthalmology department after experiencing progressive loss of vision in the left eye (OS). He had a history of retinal detachment in the OS, leading to neovascular glaucoma. The examination revealed a best correct visual acuity of 20/40 in the right eye (OD) and no light perception in the OS. Anterior segment biomicroscopy was unremarkable in the OD and revealed a collapsed anterior chamber, absence of pupillary light reflex, and total cataract in the OS.

Funduscopy in the OD showed irregular radiating lines extending from the optic nerve to the retina and a macular scar. A multimodal evaluation of the retina was performed (Fig.1). Fundus evaluation in the OS was impossible due to the opacity of the media.

DISCUSSION

Assumptions based on histological findings explain the causality of ASs in sickle cell disease. The studies suggest two different mechanisms leading to BM rupture: first, secondary to widespread elastic tissue degeneration. Second, a deposition of iron in BM due to chronic hemolysis. As a result, both mechanisms render a BM easier to crack. (2) The loss of BM's integrity can result in choroidal neovascularization (CNV) formation. Macular neovascularization is one of the main causes of visual loss in ASs. At the final stages, fibrosis and macular atrophy are the leading causes of irreversible visual loss. (1) In this scenario, multimodal evaluation is essential to closely monitor these patients and avoid poor outcomes.

Bibliography

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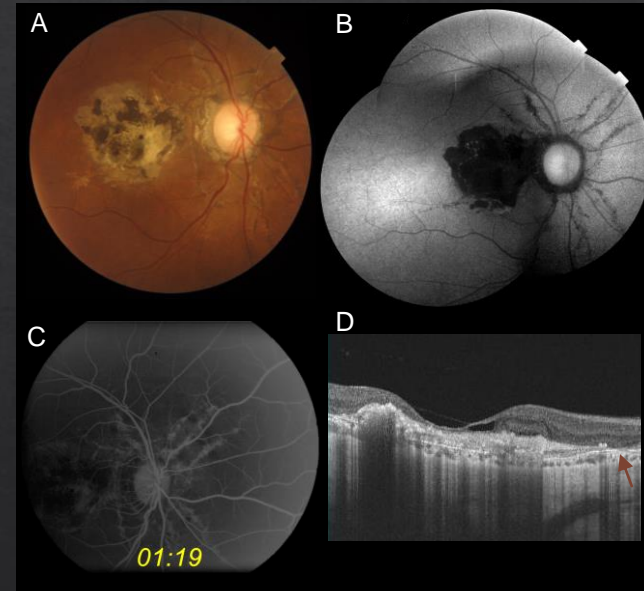


Figure 1. (A) Color retinography, (B) Autofluorescence, and (C) Fluorescein angiography showing the typical pattern of angioid streaks and a macular scar. (D) SD-OCT presenting images of hyper-reflective material above and through interruptions of Bruch's membrane (arrow).