

Successful treatment of a rhegmatogenous retinal detachment in a patient of African descent with oculocutaneous albinism

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Background

Albinism relates to a congenital disorder characterised by partial or complete absence of melanin pigment. ¹ Retinal detachment repair poses a surgical challenge in this context.

The visualisation of retinal tears in such cases is challenging as a result of poor contrast between the choroid and retina. The lack of melanin leads to an inability to attain effective chorioretinal adhesion with laser or cryo retinopexy. The effect of nystagmus creates vitreous currents that increase the risk of retinal break reopening.

We present a case of successful rhegmatogenous retinal detachment in a patient of African descent with oculocutaneous albinism.

Results

A 44 year old lady of African ethnicity with oculocutaneous albinism presented with a one week history of a shadow to her right eye on a relevant background significant for high myopia, foveal hypoplasia and nystagmus.

Her presenting visual acuity was hand movements in the right eye. Dilated fundal exam revealed a posterior vitreous detachment and an inferior, shallow macula off rhegmatogenous retinal detachment. There was a grade B proliferative vitreoretinopathy and the causative break was an inferotemporal large horse-shoe tear in lattice with an additional horse-shoe tear in the superonasal quadrant in flat retina.

Our patient underwent a 25G pars plana vitrectomy in September 2023 under local anaesthesia. A complete vitrectomy with indented vitreous base shave, cryotherapy to retinal tears and silicone oil 5700 cs tamponade was performed. The retina was successfully reattached and the silicone oil was removed with 23 G under local anaesthesia 5 months later. Her retina remains flat and her VA post-op remains stable at 6/45.

Conclusion

Identifying retinal breaks poses a challenge both in the preoperative and post-operative phases due to the pale fundus. Pre-operatively the nystagmus movements add to the challenge of identifying the breaks in an albinotic fundus. A pre-op ultra-widefield Optos image helps with the identification of retinal tears.

Chorioretinal scar production is suboptimal owing to decreased melanin synthesis. Cryotherapy retinopexy is a better option in hypopigmented fundi and a high viscosity silicone oil tamponade is in general required due to the known low success rate in such cases and risk of oil emulsification that is increased by the nystagmus movements.

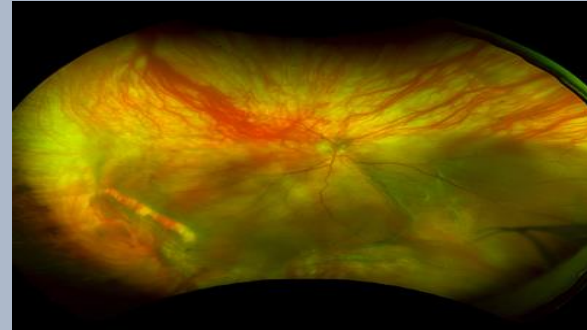
Purpose

This case reveals the added complexities and intricacies associated with albinism.

The augmented challenge of retinal detachment in albinism gives this case a unique edge. A good chorioretinal scar ablates the effects of intraocular currents. In an albinotic fundus the chorioretinal adhesion is suboptimal and the nystagmus increases the intraocular currents. ²

Long-term silicone oil tamponade is seen as a solution as the redetachment rate is high. In this particular case as the retina remained flat for 5 months a decision was made to remove the silicone oil.

1A.



1B.

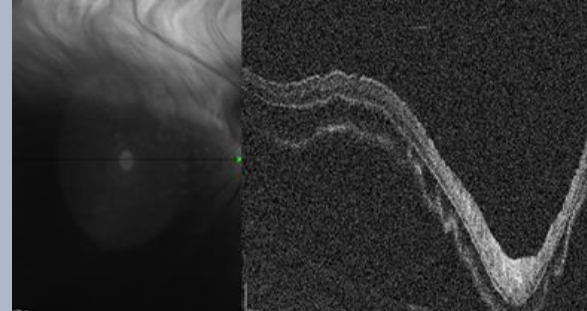


Fig 1 A. Optos photo revealing an inferior retinal detachment in a hypopigmented fundus

Fig 1 B. OCT macula scan showing a macula-off retinal detachment.

Methods

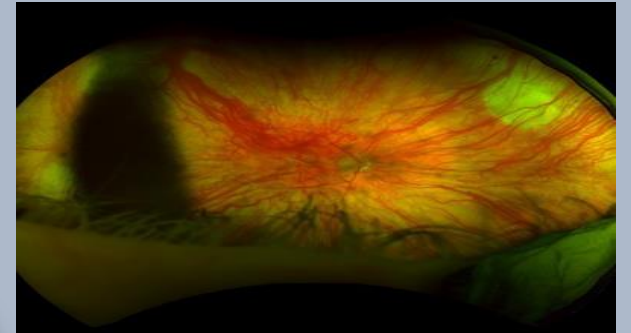
A retrospective chart review was conducted with attention drawn towards niche retinal detachment cases.

Informed consent was obtained from the patient.

References

1. Levin AV, Stroh E. Albinism for the busy clinician. J AAPOS. 2011 Feb;15(1):59-66.
2. Sinha MK, Chhablani J, Shah BS, Narayanan R, Jalali S. Surgical challenges and outcomes of rhegmatogenous retinal detachment in albinism. Eye. 2016;30:422–425.

2A.



2B.

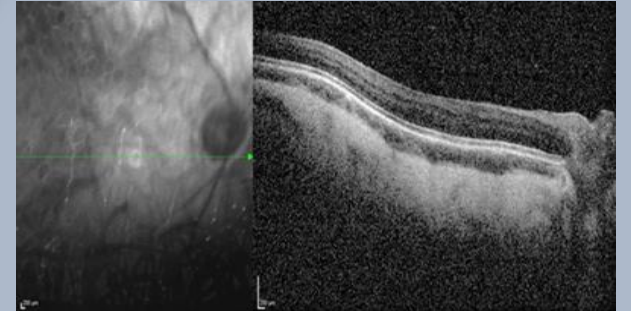


Fig 2 A. Optos photo of the right eye 5 months post RD repair and post removal of silicone oil

Fig 2 B. OCT macula demonstrating attached retina