

Eales' Disease in Inuit: A Short Report and Clinical Update

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Purpose: We report a case of Eales disease in Inuit and reflect on advances in telemedicine and treatment of retinal disease since the first report of Eales' disease in Greenlandic Inuit was published.

Patients and Methods: A 41-year-old Inuit female complaining of blurred vision was referred to our eye department. There had been no sign of diabetic retinopathy during diabetic eye screening and the patient had been treated for tuberculosis in 2010. Telemedical assessment was suspicious for vitritis or vitreous hemorrhage in the right eye, and the patient was flown from Greenland to Denmark, where examination revealed mild vitritis in the right eye, vitreous hemorrhage in the left eye and retinal neovascularization in both eyes. Fundus fluorescein angiography showed vessel leakage and areas of retinal non-perfusion. There was left epiretinal membrane with retinal thickening on macular optical coherence tomography.

Results: Based on the patient's clinical findings and history of tuberculosis infection, a diagnosis of Eales' disease was made. The left eye was treated with pars plana vitrectomy with epiretinal membrane peeling, endodiathermy and endolaser. The right eye was treated in outpatients with sectoral laser photocoagulation. At seven weeks' follow-up, the visual acuity had improved from 6/12 to 6/6 (right eye) and from 6/36 to 6/7.5 (left eye).

Conclusion: The prevalence of tuberculosis in Greenland is very high and it is recommended that clinicians remain alert to the possibility of Eales' disease, as beneficial visual outcomes are associated with prompt diagnosis and treatment. Telemedicine allows more frequent follow-up of Greenlandic patients.

Keywords: Eales' disease, Greenlandic Inuit, telemedicine, tuberculosis

Introduction

Since the first report of Eales' disease in Greenlandic Inuit was published, significant advances have been made in the fields of telemedicine and treatment of retinal disease. In 2002, equipment used for telemedicine existed in few places in Greenland.¹ Nowadays, nine out of 17 towns in Greenland are equipped with Optos[®] ultra wide-field fundus cameras, from which images are uploaded through a server and assessed for diabetic retinopathy by specialist ophthalmic nurses at Steno Diabetes Center Copenhagen.² Optos[®] ultra wide-field fundus imaging can enhance outcomes for patients with diabetic retinopathy and other retinal disorders by providing a view of 200 degrees of the retina in a single capture.³ Furthermore, patients with diabetic maculopathy can now receive intravitreal anti-vascular endothelial growth factor (anti-VEGF), which has been available in Denmark for the treatment of retinal disease since 2006.⁴ An update regarding the diagnosis and management of Eales' disease in Inuit is therefore warranted.

The first report of Eales' disease in Inuit included four patients, concluding that the occurrence of Eales' disease in Greenland is no longer negligible and reaffirming its association with tuberculosis (TB).⁵ Furthermore, it was suggested that there may be undiagnosed patients and clinicians should be alert to possible new cases.⁵ Promoting greater awareness of Eales' disease in Greenlandic Inuit is especially important because the prevalence of TB in Greenland is still very high.⁶ Greenland is geographically remote and there are problems accessing healthcare which may further delay diagnosis and treatment and lead to negative visual outcomes. For example, there are very few connecting roads in

Greenland; travel between towns is either by air, or by boat and by dog sled. Similarly, there are no resident ophthalmologists in Greenland, although telemedicine-assisted consultations with clinicians in Denmark help to exclude a range of eye diseases and telemedicine has been used successfully in diabetic retinopathy screening.² When considering healthcare inequalities affecting people in Greenland, it is worth noting that people born in Greenland have a shorter life expectancy compared with people born in Denmark by approximately 10 years: 69.6 years for males and 73.5 years for females, compared with 79.6 years for males and 83.4 years for females,^{7,8} which is primarily due to the high mortality rate caused by accidents and suicide.⁷

Patients and Methods

A 41-year-old Inuit female complaining of blurred vision in the left eye was referred to our eye department. The patient's past medical history included diabetes mellitus type 2, arterial hypertension and hypercholesterolemia. The patient had no past ocular history and there was no sign of diabetic retinopathy during diabetic eye screening six months prior, although a possible area of neovascularization was found in the right eye that could not be attributed to diabetes. In retrospect, this area of possible neovascularization may have been an early sign of Eales' disease: the patient had received antibiotic treatment for TB in 2010 following a positive QuantiFERON[®] test. Telemedical assessment including ultra wide-field fundus imaging could neither confirm nor deny a specific diagnosis, but images were suspicious for vitritis or vitreous hemorrhage in the right eye. Eales' disease could not be ruled out, and the patient was flown from Greenland to Denmark for further evaluation, including investigation of the cause of her suspected vitreoretinopathy. Arrangements were made for the patient to stay at the Greenland patient hotel in Copenhagen for the duration of her treatment.

Results

On examination, visual acuity was 6/12 in the right eye and 6/36 in the left eye (Snellen). Fundoscopy revealed mild vitritis in the right eye, a vitreous hemorrhage in the left eye with neovascularization of the disc in the left eye (Figure 1a, Figure 1b) and retinal neovascularization elsewhere in both eyes (Figure 1c, Figure 1d). Fundus fluorescein angiography showed vessel leakage and areas of non-perfusion in the retinal mid-peripheries (Figure 1e, Figure 1f). There was left epiretinal membrane with retinal thickening on macular optical coherence tomography.

Eales' disease is an occlusive vasculitis involving the retinal mid-periphery that commonly presents with vitreous hemorrhage. It is further characterized by periphlebitis, vascular occlusion and subsequent retinal neovascularization. Based on the patient findings and history of TB infection in this case, a diagnosis of Eales' disease was made. The patient's epiretinal membrane was thought to be visually significant, leading to treatment of the left eye with pars plana vitrectomy with epiretinal membrane peeling, endodiathermy and endolaser. The right eye was treated in outpatients with sectoral laser photocoagulation. At seven weeks' follow-up, the visual acuity had improved to 6/6 (right eye) and 6/7.5 (left eye). The patient had met with the visual acuity requirement for driving-licence vision in either eye and both eyes were deemed to be sufficiently treated.

Discussion

Eales' disease is a diagnosis of exclusion, and the differential diagnosis includes causes of vitreous hemorrhage such as a hemoglobinopathy; causes of periphlebitis such as sarcoidosis; causes of vascular occlusion including various collagen diseases and retinal vein occlusion and conditions that predispose to neovascularization, such as diabetic retinopathy.⁵ These vitreoretinal findings may overlap, adding to the complexity of making a diagnosis of Eales' disease. Although the pathophysiology remains unknown, Eales' disease may be a hypersensitive reaction caused by a tuberculin antigen.^{9,10} Studies have implicated *Mycoplasma tuberculosis* and *Mycoplasma fortuitum* in the development of the disease,^{11,12} which is likely to be multifactorial.¹³ The QuantiFERON[®] test is a more recent aid to diagnosis than the Mantoux test; hence the patients described in the first report of Eales' disease in Inuit were given the Mantoux test, according to which all were positive for TB. Unlike the QuantiFERON[®] test, however, the Mantoux test may be positive after BCG vaccination.¹⁴ To combat the increase in incidence of TB in Greenland in the 1990s, national TB interventions including neonate BCG vaccination were initiated in 2000 and strengthened in 2007,¹⁵ which may mean that the Mantoux test is becoming a less reliable method of diagnosis of Eales' disease in Greenland. Fewer reliable diagnostic methods may

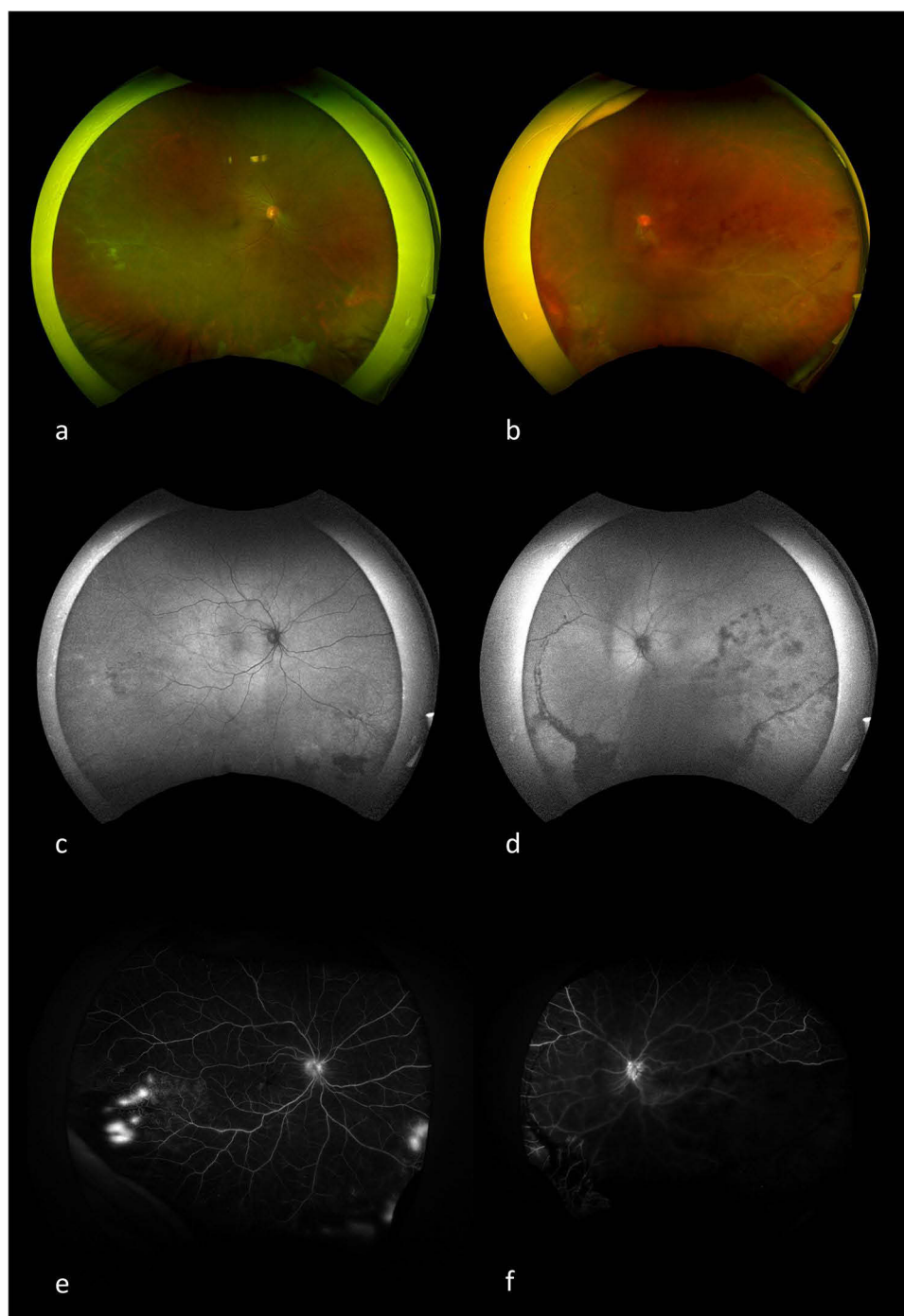


Figure 1 Composite showing images from the right and left eye of an Inuit patient with Eales' disease: (a) and (b) wide-field fundus photography with retinal ghost vessels and neovascularization in both eyes and vitreous hemorrhage and neovascularization of the disc in the left eye; (c) and (d) wide-field fundus autofluorescence photography showing evidence of retinal neovascularization at 4, 5, 8 and 9 o'clock in the right eye and at 8 o'clock in the left eye; (e) and (f) wide-field fundus fluorescein angiography showing areas of fluorescein leakage and ischemia of the inferior peripheral retina.

contribute to Eales' disease being underdiagnosed and a lack of reported cases of Eales' disease in Greenland may explain why estimating disease prevalence in the Inuit population of Greenland is so difficult.⁵

Eales' disease in the proliferative stage is treated by photocoagulation and is characterized by neovascularization occurring at the junction between perfused and non-perfused retina. Contraction and rupture of these vessels leads to recurrent vitreous hemorrhage with or without tractional retinal detachment, which may necessitate vitreoretinal surgery.

In cases where there are significant areas of retinal non-perfusion, periocular steroid may help with local inflammation, oral steroid may be used to treat inflammation systemically, and intravitreal anti-VEGF can help prevent further neovascularization. In the first report of Eales' disease in Inuit, all patients had best corrected visual acuity (BCVA) in the best eye of 6/24 or better at final follow-up.⁵ However, no single eye had BCVA that was the same or better than that of our patient and one vitrectomized eye had BCVA below 6/60.⁵ In addition, all patients were treated with bilateral panretinal photocoagulation as follow-up of Greenlanders was then possible only on a once-a-year basis.⁵ Better access to telemedicine now enables more frequent follow-up of patients in Greenland, and in this case the patient's right eye was treated with sectoral laser photocoagulation in accordance with local treatment protocols.

Conclusion

The prevalence of TB in Greenland is still very high and it is therefore recommended that clinicians remain alert to the possibility of Eales' disease, as beneficial visual outcomes are associated with prompt diagnosis and treatment. Furthermore, recent advances in telemedicine allow better and more frequent follow-up of Greenland patients.

Ethical/Copyright Concerns

Institutional approval was not required to publish the case details.

Patient Consent

Consent to publication of personal information in print or online, including the use of images, has been obtained prior to publication.

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Disclosure

The authors report no conflicts of interest in this work.

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