• Letter to the Editor •

Surgical treatment for proliferative diabetic retinopathy with ocular albinism

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Dear Editor,

We are writing to present an uncommon case of a 45-year-old woman with bilateral proliferative diabetic retinopathy (PDR) and ocular albinism (OA). Albinism is a group of inherited disorders of tyrosinase activities and melanin biosynthesis resulting in little or no production of the pigment melanin, which affects the color of eyes (OA) as well as the skin and hair (oculocutaneous albinism, OCA)[1-2]. OA is characterized by hypopigmentation of the uvea and retinal pigment epithelium (RPE) resulting in multiple ocular abnormalities, such as severe impairment of visual acuity and stereoscopic vision, nystagmus, strabismus, photophobia and glare, and occasionally optic nerve dysplasia. OA can be further divided into OA type 1 (OA1), which is X linked recessive and affects males almost exclusively, and autosomal recessive ocular albinism (AROA), which is autosomal recessive, with females affected as severely as males[3]. Due to the lack of pigment in the uvea and RPE, the diagnosis and surgical interventions of vitreoretinal disorders are challenging, such as the difficulty in visualization of retinal breaks in retinal detachment and the inability for laser photocoagulation or cryopexy to achieve a good chorioretinal adhesion[4]. We herein report a case of PDR in a patient with OA, which is rarely reported in the literatures, and discuss the therapeutic strategies and challenges involved.

Ethical Approval This study complied with the tenets of the Declaration of Helsinki. The research was conducted in compliance with a suitable accredited Institutional Review Board from the Ethics Committee of Joint Shantou International Eye Center of Shantou University and the Chinese University of Hong Kong. Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

CASE REPORT

A 45-year-old woman with a history of bilateral poor vision since childhood reported progressive visual loss in her left eye for one month. She had a past medical history of OA, type 2 diabetes, hypertension, and was notable for bilateral retinal laser treatment. On examination, her best corrected visual acuity (BCVA) was 20/200 OD and finger count OS and the intraocular pressure in both eyes was normal. Bilateral concomitant exotropia of 45 degrees was present whereas nystagmus was not observed. Slit-lamp microscopy showed bilateral iris transillumination (Figure 1A, 1B) and cortical lens opacities. The posterior pole of the fundus in both eyes showed a turbid "orange-red" appearance indicating hypopigmentation of RPE and choroid (Figure 1C, 1D). Neovascularization of the disc associated with vessel distortion was noted in her left eye (Figure 1D). Scanning laser ophthalmoscopy revealed fundus-obscuring vitreous hemorrhage with a few laser scars on the peripheral area of the right eye (Figure 1E) and tractional retinal detachment in the left eye (Figure 1F). Bilateral vitreous hemorrhage and proliferative vitreoretinopathy of the left eye were demonstrated by B-scan (Figure 1G, 1H). Low-quality optical coherence tomography images due to media opacity and vitreous hemorrhage revealed bilateral macular edema with unidentified retinal layers (Figure 1I, 1J). Laboratory examination at first presentation showed fairly controlled diabetes (fasting blood glucose 5.8 mmol/L, HbA1c 5.8%) and mild renal dysfunction (BUN 6.3 mmol/L, creatinine 145 μmol/L). There is no white skin or white scalp hair. Diagnosis of bilateral high-risk PDR and OA was made.

The patient underwent combined phacoemulsification and 23-gauge pars plana vitrectomy (PPV) in her left eye. Intravitreal injection of anti-vascular endothelial growth factor (VEGF) agents (conbercept; KH902; Kanghong Biotech Co.,

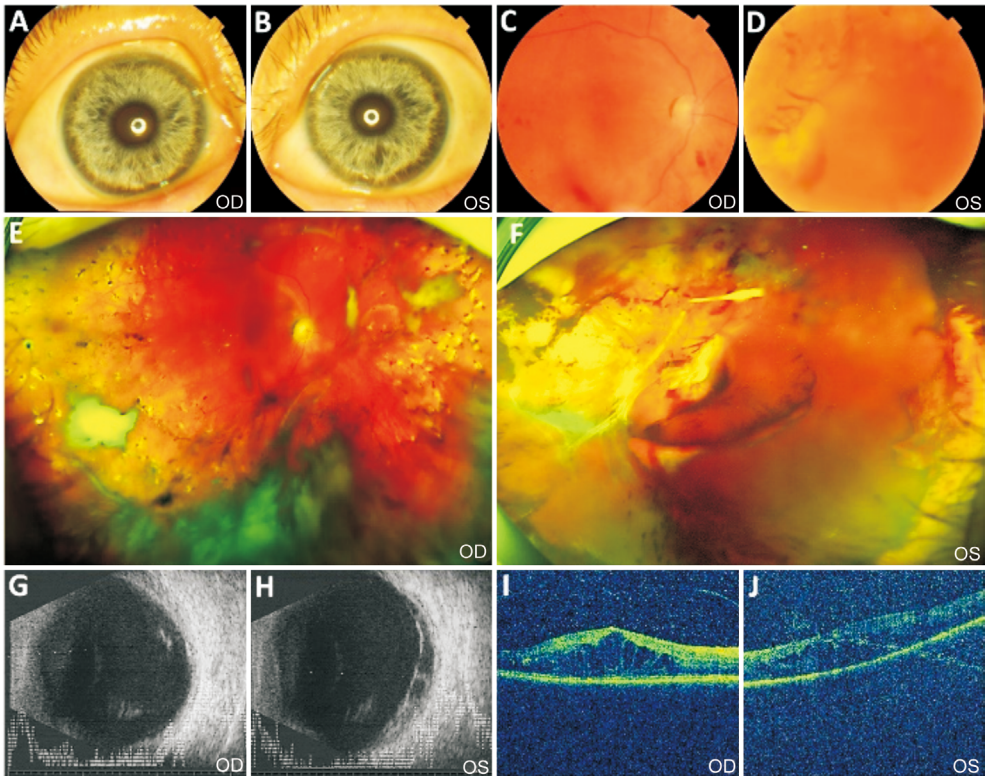

Figure 1 Clinical findings in a patient with proliferative diabetic retinopathy and ocular albinism at first presentation A, B: Slit-lamp examination showed bilateral hypopigmentation of the iris. C, D: Fundus photography showed a "red-orange" appearance with turbid media. Disc neovascularization and distorted vessels were noted in the left eye (D). E, F: Scanning laser ophthalmoscopy revealed bilateral vitreous hemorrhage. Left traction retinal detachment can be observed (F). G, H: B-scan indicated vitreous hemorrhage in both eyes and proliferative vitreoretinopathy in the left eye. I, J: Optical coherence tomography showed macular edema in both eyes. OD: Right eye, OS: Left eye.

Ltd., China) was performed 1wk before the PPV surgery as pretreatment. After removal of the hemorrhagic vitreous, the proliferative membrane was carefully dissected and removed without inducing severe retinal hemorrhage and iatrogenic breaks. Hypopigmentation of the RPE and choroid with scattered retinal hemorrhage were visible. Supplemental retinal photocoagulation was administered in the mid-peripheral and peripheral areas of the left eye (532 nm green laser, PurePoint, Alcon Inc, USA, 200-240 mW power and 200ms duration). Filtered air was used as an internal tamponade. At one-month follow-up, her left BCVA improved to 60/200. Operation of the right eye was advised but the patient declined any intervention. One year later, her left eye had stable visual acuity whereas the right eye developed more severe vitreous hemorrhage and the BCVA declined to finger count so that the same surgical procedures including anti-VEGF intravitreal injection and PPV were carried out. The BCVA of her right eye improved to 40/200 one month postoperatively. There were visible laser scarring and clear choroidal vessels in the posterior pole with no recurrence of vitreous hemorrhage (Figure 2A, 2B). Optical coherence tomography confirmed the improvement of macular edema in both eyes (Figure 2C, 2D). At the 2-year follow-up, her BCVA maintained stable at 40/200 OD and 60/200 OS without any diabetes-associated neovascular complications (Figure 2E, 2F). She reported having well-controlled diabetes and hypertension but developed uremia requiring hemodialysis.

DISCUSSION

OA is an X-linked recessive or autosomal recessive disorder characterized by hypopigmentation of the RPE and uvea, often associated with severely reduced visual acuity, photophobia, strabismus, nystagmus, and foveal hypoplasia[3]. The treatment of retinal diseases, such as retinal detachment and PDR, in patients with OA remains challenging as the lack of melanin in RPE and choroid makes it theoretically and technically difficult to perform laser photocoagulation[5].

Previous studies hold different opinions regarding the use of laser photocoagulation when treating retinal diseases in OA. Some researchers found that laser reaction could not be achieved during surgery while retinal cryoablation was needed for the treatment[6-7]. Some others successfully achieved visible laser burns using either Argon green laser (532 nm) with 200-400 mW power and of 150-300ms duration[8], Krypton red laser[9], or Argon yellow laser with a longer duration (300-500ms)[10], in the treatment of rhegmatogenous retinal detachment. Due to the lack of melanin within the RPE and choroid, a laser wavelength should be selected, when possible, with strong absorption to hemoglobin in choriocapillaris in

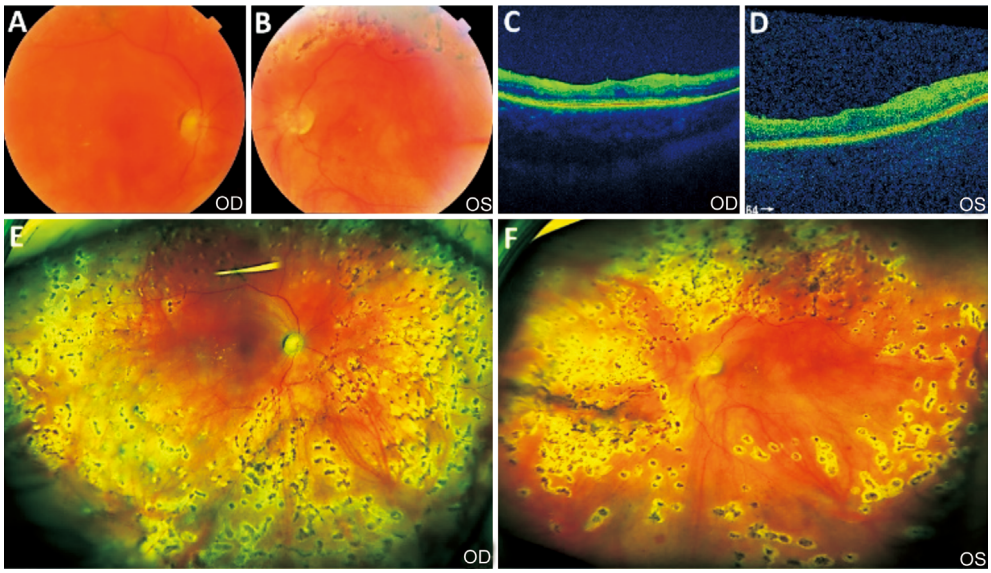

Figure 2 Imaging of the patient after bilateral surgical interventions A, B: Fundus photography showed a "red-orange" appearance and visible laser scarring. C, D: Optical coherence tomography showed regression of macular edema in both eyes. E, F: Scanning laser ophthalmoscopy revealed a clear fundus with diffuse laser scars and large choroidal vessels. OD: Right eye; OS: Left eye.

order to reach a thermal and damaging effect involving the RPE and outer retina. In our case, we used 532 nm green wavelength laser to maximize absorption of the laser energy by hemoglobin. Attention should also be given to the power and duration of laser. In the presence of OA, increased power and long duration may be necessary to obtain the desired destruction of the RPE-photoreceptor complex. On the other hand, overuse of laser photocoagulation should be avoided as PDR can be controlled using relatively light and subclinical burns, as compared with a stronger laser to induce a firm chorioretinal adhesion in retinal detachment[11]. Surgeons can make appropriate adjustments during the procedures to achieve an optimal laser reaction. In our case, we used 200-240 mW power and 200ms duration and obtained visible laser burns in the mid-peripheral and peripheral retinal area, suggesting that laser photocoagulation can be successfully achieved in OA patient at suitable laser processing parameters.

We agree with previous ideas[12] that cryotherapy should not be prioritized due to its association with intraocular inflammation and pain, unless laser strategy could not achieve visible burns. For severe neovascular complications including tractional retinal detachment and nonclearing vitreous hemorrhage, surgical intervention such as PPV or phaco-PPV can be considered.

The anti-VEGF pretreatment has been established in the management of complicated PDR, which is believed to reduce the incidence of intraoperative bleeding and early recurrent vitreous hemorrhage, to avoid the use of silicone oil tamponade, to facilitate easier surgery and shorten its duration, and to have better postoperative visual acuity[13-14]. We performed intravitreal injection of anti-VEGF agents one week before PPV in our patient and achieved satisfactory outcomes. Notably, a recent study suggested minimal laser photocoagulation whereas more aggressive anti-VEGF treatment for patients with diabetic retinopathy in the presence of OA, since that the amount of laser administered to these patients would significantly reduce the peripheral vision which in the case of a low vision patient due to central vision issues from the albinism[5]. We endorse such ideas. However, in case of a high-risk PDR with OA, insufficient retinal photocoagulation will result in neovascular complications so that pan retinal photocoagulation is still necessary.

In summary, we reported a case of OA with bilateral PDR successfully treated with phaco-PPV following laser photocoagulation. The treatment of PDR with OA is difficult and challenging owing to lack of melanin in the RPE and choroid. Nevertheless, laser photocoagulation can still work and should be considered to avoid worsening of high-risk diabetic retinopathy.

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Conflicts of Interest: Huang ZJ, None; **Huang DG,** None; **Chen WQ,** None.

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