

CASE REPORT

Long Term Outcome in Unilaterally Treated Retinopathy of Prematurity: A Case Report

MUSHAWIAHTI M¹, ROKIAH O², UMI KMN¹, LEOW SN¹

¹Department of Ophthalmology, Faculty of Medicine, Universiti Kebangsaan Malaysia Medical Centre, Jalan Yaacob Latif, Bandar Tun Razak, 56000 Cheras, Kuala Lumpur, Malaysia.

²Department of Optometry, Faculty of Allied Sciences, Universiti Kebangsaan Malaysia, Jalan Raja Muda Abd Aziz, 50300, Kuala Lumpur, Malaysia.

ABSTRAK

Retinopati pramatang (ROP) adalah penyakit berkaitan dengan pertumbuhan saluran darah tidak matang pada bayi berat lahir rendah disebabkan oleh kekurangan oksigen pada retina yang sedang dalam pertumbuhan. Keadaan ini boleh membawa kepada risiko kebutaan. ROP selalunya terjadi pada kedua-dua mata, namun ada juga kes yang menunjukkan kelainan tahap keterukan penyakit di antara kedua mata. Kami melaporkan contoh kes yang mempunyai presentasi keterukan penyakit yang berbeza di antara kedua matanya dan rawatan ablasi retina hanya diberikan pada mata yang teruk sahaja. Kes ini memberi contoh salah satu kesan jangka panjang kepada pesakit yang mengalami ROP yang lebih teruk di dalam satu mata.

Kata kunci: retinopati pramatang, ambliopia, anisometropia, miopia, fotokoagulasi cahaya

ABSTRACT

Retinopathy of prematurity (ROP) is a disorder describing an immature vascularisation of a developing retina in low birth weight preterm infants. This condition potentially leads to blindness. ROP developed as a response of hypoxia of the eye due to incomplete development of the retinal vessels. ROP is commonly reported as bilateral disease, a small percentage of infants have asymmetrical changes. We report a case of long-term outcome of an asymmetry ROP changes with peripheral retinal ablation in a single eye. This particular case demonstrates the possible long-term outcome of unilaterally treated ROP which could either be due to the severity of the disease itself or the treatment she received. It is important to highlight the possibility of unequal development of the eye in asymmetrical presentation of ROP.

Address for correspondence and reprint requests: Dr. Mushawiahti Mustapha, Department of Ophthalmology, Faculty of Medicine, Universiti Kebangsaan Malaysia Medical Centre, Jalan Yaacob Latif, Bandar Tun Razak, 56000 Cheras, Kuala Lumpur, Malaysia. Tel: +60391455981 Fax: +60391737856 Email: drmusha@yahoo.com/ drmusha@gmail.com

Keywords: retinopathy of prematurity, amblyopia, anisometropia, myopia, light photocoagulation

INTRODUCTION

Although, the management of premature infants has improved over the years, the long-term morbidity of these infants, especially due to conditions such as retinopathy of prematurity (ROP), is still a great challenge. The risk factors for developing ROP are birth weight less than 1500gm and prematurity of less than 32 weeks (Palmer et al. 1991). ROP accounts for 3% of all cases of childhood blindness (Rahi & Cable 2003). ROP occurs as a result of hypoxia of the eye due to incomplete development of the retinal vessels. The severity of the disease is determined on the basis of the size of the avascular area, amount of abnormal vessels, and the presence of the features of plus disease that increase the risk of further disease progression. Treatment was recommended only if threshold criteria were met (Palmer et al. 1991).

The incidence of blindness from untreated ROP has been significantly reduced by the availability of treatment (Hardy et al. 1997). Management of peripheral retinal ablation either with laser treatment or cryotherapy induces rapid regression of abnormal vessels and allows for vascularization to continue normally towards the ora serata. According to the findings of the Cryotherapy for Retinopathy of Prematurity (CRYO-ROP) study, treatment is necessary only for eyes with "threshold" disease. Even if successful regression of the disease is

achieved immediately after treatment, other ocular morbidities ensue with the growth of the infant. Strabismus and amblyopia are still being reported as the long-term morbidities observed in such cases (VanderVeen et al. 2011).

Immaturity of the retina is described on the basis of the area of the avascular zone; presence of changes in zone 1 is associated with the most severe form of immature retina, while those in zone 3 were the least severe form of the incomplete development of the retina (Cryotherapy for Retinopathy of Prematurity Cooperative Group 1988). Plus disease is defined by the presence of arterial tortuosity and venous dilatation, which are the critical factors for evaluating treatment. While the threshold for treatment is at least five contiguous or eight cumulative sectors (clock hours) showing stage 3 ROP in zone 1 or II in the presence of plus disease (Cryotherapy for Retinopathy of Prematurity Cooperative Group 1988).

Although ROP is commonly reported as a bilateral disease, a small percentage of infants have asymmetrical changes. Hence, the indication of treatment may differ and one eye might remain untreated. To the best of our knowledge, no detailed report has been published on the long-term outcome of unilateral ROP treatment. We present a case demonstrating one of the possible long-term outcome in a patient with peripheral retinal ablation in a single eye for threshold ROP. At the time of

treatment constitution, the CRYO-ROP study was the main treatment guideline.

CASE REPORT

The patient was a baby girl born at 29 weeks of gestation in 2004 with a birth weight of 1500 gm. At birth, the patient had respiratory distress syndrome and received respiratory support for more than three weeks. Her general condition improved with the treatment. During routine ophthalmologic screening, bilateral changes were consistent with ROP detected at 34 weeks following conception. There were asymmetrical changes in both eyes. The patient's left eye fundus revealed changes of "threshold" ROP (stage 3 ROP with changes for more than 5 clock hours and preretinal hemorrhage in zone 2 with plus disease). However, the right eye showed less severe changes (stage 2 ROP changes less than 4 clock hours in zone 2) with no evidence of plus disease. As per the CRYO-ROP criteria, treatment was indicated only in left eye that had threshold disease. Therefore, indirect argon laser treatment was administered only for the left eye. Meanwhile, no treatment was administered to the contralateral eye with less severe disease and not meeting the threshold criteria.

The infant responded well to the treatment, and both eyes showed rapid regression of the abnormal vessels. The patient was examined every six months and evaluated for refractive errors every six months. At six months of age, the spherical equivalent was found to be +0.75 D in both eyes with no evidence of squint at the beginning. The patient's

myopic changes progressed more rapidly in the left eye than in the right eye. Myopic changes at 18 months of age showed significant differences between the two eyes. The spherical equivalent of the treated eye was -2.25 D, while that of the untreated eye was only -0.30D. Significant difference in refractive error between the two eyes putting her at risk to develop amblyopia in the future. Patching of the good eye which is one of the treatment modalities to prevent amblyopia was instituted early. However, the patient did not comply with this treatment. Anisometropia (significant difference of the refractive error between the two eyes) progressed with her age. At eight years of age, the myopia in her left eye worsened to -6.25D with best-corrected visual acuity of 6/36, while her right eye remained emmetropic (no significant refractive error) with 6/6 vision. Ocular biometry at the age of 8 years revealed steeper corneal curvature in the left eye, with a longer axial length of 24.17mm in the affected eye and 21.37mm in the other eye. With the resultant anisometropia and poor compliance to treatment, the patient experienced dense amblyopia with subsequent squint of the left eye.

DISCUSSION

In premature infants, prolonged exposure to assisted ventilation, which may be necessary for survival, is often detrimental to the eye, leading to conditions such as ROP (Todd et al. 1990). ROP is a disease affecting the development of the immature vasculature of the eyes of premature

babies. It presents in various grades of severity, ranging from very mild disease without any significant visual effect or very aggressive disease with abnormal vessel proliferation resulting in retinal detachment and blindness. Asymmetrical presentation of ROP is not uncommon; it has been reported that the differences in the severity of the eyes of the same patient with only one eye requiring treatment occurs in up to 20% of cases (Azad et al. 2010).

The infant in this case developed threshold changes with plus disease in the left eye and less severe form of ROP without the presence of threshold in the fellow eye. Hence, treatment was constituted only in the eye meeting the threshold criteria in accordance with the standard guideline of CRYO-ROP, which was the main treatment guideline at the time of presentation. Peripheral retina laser ablation is the main stay of treatment.

Despite reducing the risk of complete blindness, significant refractive error particularly myopia (short sighted) which may lead to amblyopia is still frequently reported in a high proportion of treated infants (Al-Gamdhi et al. 2004). Amblyopia (also known as lazy eye) is a condition which involves reduced vision in an eye that otherwise appears normal, or out of proportion to associated anatomical problems of the eye. Detecting the condition in early childhood increases the chance of successful treatment.

In this particular case, at the age of eight years, the infant was found to have a longer axial length in the treated eye than in the other eye which may explain the significant refractive error

condition. The mechanisms of myopia in infants with ROP are debatable. Most experts agree that the myopic effect is the result of poor anterior segment development in ROP, particularly of the lens and cornea (Garcia-Valenzuela & Kaufman 2005). At the same time, the incidence of myopia in treated infants was reported to correlate with increases in the stage of ROP, clock hours of involvement, number of laser spots, and period of regression (Katoch et al. 2011).

However, others have also reported that the possible effect of treatment particularly peripheral retinal ablation may interfere with the normal ocular development (Ng et al. 2002). Therefore, the exact mechanism of myopia in ROP with or without treatment is still poorly understood. It is always presumed that regression of ROP and resumption of the normal development of retinal vessels will help the patient achieve good visual acuity in the future. However, obviously regression of the disease is not the only determinant factor for a good visual outcome.

It is important to highlight the possibility of unequal development of the eye in asymmetrical presentation of ROP. The unresolved issues regarding the long-term outcome of ocular development in ROP patients warrants consideration during the planning of treatment in asymmetrical cases. Strict monitoring of amblyopia treatment would help to prevent poor visual outcome in such cases.

CONCLUSION

Asymmetrical presentation of ROP and the resultant differences in the

treatment regimens for both eyes may have different effects on the long-term ocular development of the eyes of the patient. Although regression of ROP after treatment increases the chances of achieving a useful level of vision in the patient, regular monitoring and strict intervention are equally important for the management of significant refractive error to avoid poor vision in the future.

REFERENCES

- Al-Ghamdi, A., Albiani, D.A., Hodge, W.G., Clarke, W.N. 2004. Myopia and astigmatism in retinopathy of prematurity after treatment with cryotherapy or laser photocoagulation. *Can J Ophthalmol* **39**(5): 521-5.
- Azad, R., Chandra, P., Patwardhan, S.D., Gupta, A. 2010. Profile of asymmetrical retinopathy of prematurity in twins. *Indian J Ophthalmol* **58**(3): 209-11.
- Cryotherapy for Retinopathy of Prematurity Cooperative Group. 1988. Multicenter trial of cryotherapy for retinopathy of prematurity: preliminary results. *Arch Ophthalmol* **106**(4): 471-9.
- Garcia-Valenzuela, E., Kaufman, L.M. 2005. High myopia associated with retinopathy of prematurity is primarily lenticular. *J AAPOS* **9**(2): 121-8.
- Hardy, R.J., Palmer, E.A., Schaffer, D.B., Phelps, D.L., Davis, B.R., Cooper, C.J. 1997. Outcome-based management of retinopathy of prematurity. Multicenter Trial of Cryotherapy for Retinopathy of Prematurity Cooperative Group. *J AAPOS* **1**(1): 46-54.
- Katoch, D., Sanghi, G., Dogra, M.R., Beke, N., Gupta, A. 2011. Structural sequelae and refractive outcome 1 year after laser treatment for type 1 prethreshold retinopathy of prematurity in Asian Indian eyes. *Indian J Ophthalmol* **59**(6): 423-6.
- Ng, E.Y., Connolly, B.P., McNamara, J.A., Regillo, C.D., Vander, J.F., Tasman, W. 2002. A comparison of laser photocoagulation with cryotherapy for threshold retinopathy of prematurity at 10 years: part 1. Visual function and structural outcome. *Ophthalmology* **109**(5): 928-34.
- Palmer, E.A., Flynn, J.T., Hardy, R.J., Phelps, D.L., Phillips, C.L., Schaffer, D.B., Tung, B. 1991. Incidence and early course of retinopathy of prematurity. The Cryotherapy for Retinopathy of Prematurity Cooperative Group. *Ophthalmology* **98**(11): 1628-40.
- Rahi, J.S., Cable, N. 2003. Severe visual impairment and blindness in children in the UK. *Lancet* **362**(9393): 1359-65.
- Todd, D., Kennedy, J., Roberts, V., John, E. 1990. Risk factors in progression beyond stage 2 retinopathy of prematurity. *Aust N Z J Ophthalmol* **18**(1): 57-60.
- VanderVeen, D.K., Bremer, D.L., Fellows, R.R., Hardy, R.J., Neely, D.E., Palmer, E.A., Rogers, D.L., Tung, B., Good, W.V. 2011. Prevalence and course of strabismus through age 6 years in participants of the Early Treatment for Retinopathy of Prematurity randomized trial. *J AAPOS* **15**(6): 536-40.