Sir,

Long-term outcome of bilateral penetrating keratoplasty in a child with xeroderma pigmentosum: case report and literature review

Xeroderma pigmentosum (XP) is a rare autosomal recessive disorder due to defect in DNA repair mechanism. Corneal involvement is seen in 17% of cases.[1,2] Outcomes of penetrating keratoplasty (PKP) are usually poor.[1,3] We report the youngest child with XP to have PKP with the longest follow-up till date.

Case report

A 6-year-old male child of consanguineous parents was seen with a clinical diagnosis of XP. His visual acuity in both eyes (OU) was hand movements close to face. The lid position, congruity, and tear meniscus height were normal in OU. Examination under anaesthesia revealed full-thickness corneal scarring (Figure 1a) with calcific degeneration and normal anterior chamber in OU.

The patient underwent an uneventful PKP in his left eye (OS) in June 2004 and right eye (OD) in April 2005. Subsequent postoperative follow-ups were uneventful, and dexamethasone eye drops were tapered to once daily over 6 weeks. Suture removal was done by 12 weeks after surgery. He developed posterior sub-capsular cataract in OU for which he underwent lens aspiration in September 2007 (OD) and January 2008 (OS). His both eyes were highly myopic and hence intraocular lens was not implanted.

At the most recent visit, May 2010, on crowded logMAR, the patient could manage 0.8 at 33 cm using $+0.50\text{DSph}/-2.50\text{DCyl}\times 135$ correction with OD and 0.8 at 70 cm using $-2.5\text{DSph}/-0.50\text{DCyl}\times 135$ correction with OS. With OU, he could read N36 at 33 cms. The grafts in both eyes of the patient were clear (Figures 1b and c) and intraocular pressures were within normal limits.

Comment

The median age of onset of ocular manifestations has been reported to be 4 years.[1] Literature shows PKP done in only two cases in children (Table 1).[4] These two cases had an early graft failure and required a repeat PKP for visual rehabilitation.[4]

Amblyopia due to stimulus deprivation in this case has resulted in low vision. Nevertheless this child can now read enlarged print, navigate in unfamiliar surroundings, and attend a normal school with educational support. This child's grafts may have done well due to healthy lid and tear film, which in part may have been due to a shorter duration of the disease process prior to surgery. Hence, retaining healthy eyelids and in turn ocular surface will increase the longevity of corneal grafts even in chronic diseases such as XP.

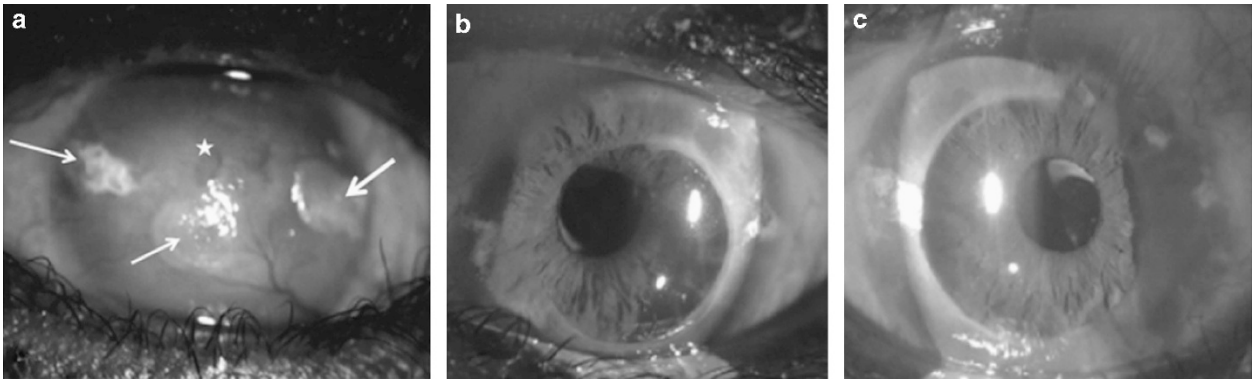

Figure 1 Clinical pictures showing (a) right corneal scarring with calcific plaque-like degeneration (white arrows) and generalised corneal opacification (white star) before PKP, (b) showing clear graft in left eye and (c) right eye at the most recent follow-up.

Table 1 Literature review of children with xeroderma pigmentosum who had penetrating keratoplasty

Author	*Age at surgery*	*Diagnosis*	*Preoperative VA*	*Postoperative VA*	*Complications*	*Regraft*	*FFU (m)*	*Final outcome*
Freed man[4]	16 years	Opacity	PL +	20/30	Rejection-graft failure	3 months	6	Clear graft
	13 years	Opacity	PL +	20/80	Rejection-graft failure	3 months	6	Clear graft
Present study 2012	7 years (OS)	Opacity	HM	0.8 at 33 cm on crowded logMAR	Cataract, amblyopia	—	71	Clear graft
	8 years (OD)	Opacity	HM	0.8 at 70 cm on crowded logMAR	Cataract, amblyopia	—	61	Clear graft

Abbreviations: cm, centimeters; FFU, final follow-up; m, months; HM, hand movements; OD, oculus dexter; OS, oculus sinister; PL, perception of light; VA, visual acuity.

Conflict of interest

The authors declare no conflict of interest.

References

1 Kraemer KH, Lee MM, Scotto J. Xeroderma pigmentosum. Cutaneous, ocular and neurologic abnormalities in 830 published cases. *Arch Dermatol* 1987; **123**: 241–250.
2 Gaasterland DE, Rodrigues MM, Moshell AN. Ocular involvement in xeroderma pigmentosum. *Ophthalmology* 1982; **89**: 980–986.
3 Jalali S, Boghani S, Vemuganti GK, Ratnakar KS, Rao GN. Penetrating keratoplasy in xeroderma pigmentosum case reports and review of the literature. *Cornea* 1994; **13**: 527–533.
4 Freedman J. Corneal transplantation with associated histopathologic description in xeroderma pigmentosum occurring in a black family. *Ann Ophthalmol* 1979; **11**: 445–448.

A Narang[1], JC Reddy[2,3], Z Idrees[1], AM Injarie[1] and KK Nischal[1,3,4]

[1]The Clinical and Academic Depatment of Ophthalmology, Great Ormond Street Hospital for Children NHS Trust, London, UK
[2]Cornea and Anterior Segment Service, L V Prasad Eye Institute, Hyderabad, India
[3]UPMC Eye Center and UPMC Childrens Hospital of Pittsburgh, Pittsburgh, PA, USA
[4]Developmental Biology Unit, University College London Institute of Child Health, London, UK
E-mail: kkn@btinternet.com

Eye (2013) **27,** 775–776; doi:10.1038/eye.2013.43; published online 5 April 2013

Sir,

Vitreomacular traction syndrome: the role of intravitreal plasmin injection is still not clear

We read the article by Codenotti *et al*[1] with interest; however, we believe that the results may be partly biased because of the study design, and some important limitations should be discussed. Firstly, only 8 out of the 13 recruited patients underwent intravitreal autologous plasmin enzyme (APE). It seems clear that a sample size that is too small may produce inconclusive results. Likewise, despite the small sample of patients and without applying any normality test, the authors used the one-way analysis of variance instead of using a non-parametric method such as Kruskal–Wallis test in order to compare the means of BCVA, macular thickness, and macular sensitivity during follow-up. A further limitation is the lack of a control group, which in this case might have a crucial role in the interpretation of the results. Moreover, the unmasked, too subjective evaluation of posterior hyaloid peeling, judged as 'difficult', 'easy' or 'very easy', may not be accurate. Although it is true that this is the first study investigating intravitreal injection of APE in eyes affected by focal vitreomacular traction (VMT), the authors could have compared their results with the MIVI-IIT trial,[2] in which a nonsurgical resolution of VMT was obtained in up to 44% of included patients, and with other studies that used intravitreal APE, for example, in diabetic macular edema associated with VMT,[3] and macular epiretinal membranes and VMT syndrome.[4]

Finally, the study deals with an important contradiction. In the conclusion, it is suggested that 'a single intravitreal APE injection seems to be insufficient to induce a complete PVD in patients affected by focal VMT syndrome', whereas in the summary and according to the authors, 'what the study adds is that intraocular APE appears to be a useful tool in vitreoretinal surgery by obtaining an easier-to-peel posterior hyaloids—a high rate of spontaneous resolution of VMT may occur'. This point should also be clarified.

Conflict of interest

The authors declare no conflict of interest.

References

1 Codenotti M, Maestranzi G, De Benedetto U, Querques G, Della Valle P, Iuliano L *et al*. Vitreomacular traction syndrome: a comparison of treatment with intravitreal plasmin enzyme *vs* spontaneous vitreous separation without treatment. *Eye* 2013; **27**: 22–27.
2 Stalmans P, Delaey C, de Smet MD, van Dijkman E, Pakola S. Intravitreal injection of microplasmin for treatment of vitreomacular adhesion: results of a prospective, randomized, sham-controlled phase II trial (the MIVI-IIT trial). *Retina* 2010; **30**: 1122–1127.
3 Elbendary AM, Elwan MM, Azzam HA, Eldeeb DR. Predictability of vitreous detachment following intravitreal plasmin injection in diabetic macular edema associated with vitreomacular traction. *Curr Eye Res* 2011; **36**: 534–539.
4 Díaz-Llopis M, Udaondo P, Cervera E, García-Delpech S, Salom D, Quijada A *et al*. Vitrectomía enzimática por inyección intravítrea de plasmina autóloga como tratamiento inicial de las membranas epirretinianas maculares y el síndrome de tracción vitreomacular. *Arch Soc Esp Oftalmol* 2009; **84**: 91–100.

A Grzybowski[1,2] and FJ Ascaso[3,4]

[1]Department of Ophthalmology, Poznan City Hospital, Poznan, Poland
[2]Medical Faculty, University of Warmia and Mazury, Olsztyn, Poland
[3]Department of Ophthalmology, 'Lozano Blesa' University Clinic Hospital, Zaragoza, Spain
[4]Aragon Health Sciences Institute, Zaragoza, Spain
E-mail: ae.grzybowski@gmail.com

Eye (2013) **27,** 776; doi:10.1038/eye.2013.61; published online 12 April 2013