Background: Osteo-odonto-keratoprosthesis (OOKP) is a radical procedure that was developed 50 years ago by Strampelli, working uniquely for a hostile dry keratinised ocular surface. It is currently the keratoprosthesis of choice for patients with inflammatory corneal blindness, using the patient’s own tooth root and alveolar bone to support a central optical cylinder. The cylinder replaces the optical elements of the eye, providing significantly improved image resolution and quality.

Case description: A 54-year-old Caucasian female presented with Stevens-Johnson syndrome secondary to a prolonged course of diclofenac. Initially the patient had symptoms of dryness, however this developed into left corneal perforation and required tectonic lamellar grafts. The grafts however failed, and the disease further progressed causing bilateral keratinisation, vascularised, scarred and thinning of her peripheral cornea. Her visual acuity was perception of light in her right eye and hand movements in her left. Due to the extent of her disease, she was suitable for OOKP in the left eye. After the procedure performed over 2 years, her final corrected visual acuity was 6/4 in her left eye.

Conclusions: This case report demonstrates the primary indication for OOKP and aims to increase awareness of the treatment option. It has the potential to dramatically improve the visual acuity in patients resulting in a better quality of life.

1 Introduction

Stevens-Johnson Syndrome (SJS) is a rare life threatening adverse reaction known to be induced by infection and drugs. Non-steroidal anti-inflammatory drugs have been implicated, however only a few cases relating to diclofenac have been reported in the literature (Shetty, Chatra, Shenai, & Rao, 2010). SJS can cause ocular surface inflammation and ulceration and destruction of the glands that secrete and maintain the tear film. This culminates in severe dry eyes that can be difficult to manage and is not indicated for keratoplasty due to the extensive corneal scarring (Jain et al., 2016). Osteo-odonto-keratoprosthesis (OOKP) is therefore an option in these patients.

OOKP is a radical procedure that was developed 50 years ago (Strampelli & Marchi, 1970), working uniquely for a hostile dry keratinised ocular surface. Keratoprosthesis in-
Restoring vision in keratinised corneas involves the surgical replacement of a diseased cornea with an artificial cornea. The OOKP aims to use the patient's own tooth root and alveolar bone to support a central optical cylinder. The cylinder replaces the optical elements of the eye, providing excellent image resolution and quality. It is currently the keratoprosthesis of choice for patients with inflammatory corneal blindness. Patients with corneal damage may initially report symptoms and signs including pain in the eye, light sensitivity, reduced and/or blurry vision and redness or inflammation of the eye. Severe corneal damage can be treated with penetrating keratoplasty (PK), a full-thickness transplant of the cornea from a donor corneal graft. However, PK is contraindicated for patients with complete loss of vision. OOKP is therefore an option in whom PK is not amenable in the treatment of damaged corneas. Indications for OOKP include patients with bilateral end-stage corneal blindness resulting from severe Stevens-Johnson syndrome, ocular cicatricial pemphigoid, chemical burns, trachoma, severe dry eyes and multiple corneal graft failure. OOKP is only considered in end stage corneal disease when no other options are available to the patient.

2 Purpose

The aim of this case report is to increase awareness of Osteo-odonto-keratoprosthesis as a treatment option for end-stage corneal blindness.

3 Case description

A 54-year-old Caucasian female, presented with Stevens-Johnson syndrome secondary to a prolonged course of diclofenac. Initial symptoms included itchy eyes with discharge, progressing to severe dryness and resulting in perforation of her left cornea, for which she required tectonic lamellar grafts (reconstructive grafts to preserve the corneal anatomy). Her grafts failed, due to a vascularised and scarred cornea. The right eye, previously amblyopic, showed reduced and delayed responses to light. Lid operations were performed to correct cicatrical entropions (outward eyelid rotation due to shortening of the skin and orbicularis muscle — anterior lamella). The result of the disease was bilateral keratinisation, vascularisation and thinning of the peripheral cornea. Ultrasound B scan revealed a normal vitreoretinal interface. Visual acuity was defined as perception of light in her right eye and perception of hand movements in the left eye. The patient was suitable for keratoprosthesis in the left eye. The two-stage procedure was subsequently performed over a period of almost 2 years, with an initial delay due to graft membrane thinning from infection. Her final corrected visual acuity was 6/4 in her left eye.

4 Procedure

Patient is assessed for previous ocular history. A and B scans are performed to ascertain an intact, functioning retina and determine axial length of eye. A psychological assessment of the patient is performed. Oral assessment for buccal mucosal membrane graft and selection of appropriate tooth to form lamina is then required. OOKP surgery is performed usually in two stages spaced two to four months apart.

Stage 1 involves ocular surface reconstruction and the fashioning of an osteo-odontol lamina and its optical cylinder. Buccal mucous graft is used to create a new ocular surface. Tooth root and surrounding bone form lamina. Lamina contains dentine on one side and bone on the other, with optical cylinder cemented in. Placed into sub-muscular pocket under orbicularis oculi muscle to acquire soft tissue covering.

Stage 2 involves retrieval of the lamina and insertion of device into the eye. Centre of cornea is marked and small hole trephined. Iridodialysis, lens extraction and anterior vitrectomy is then performed. Posterior lamina is inserted through central corneal hole and sutured onto the cornea and sclera. Mucosal flap is replaced with a hole for optical cylinder protrusion (Figure 1).

Following the OOKP, patients are monitored life-long to ensure the viability of the mucosa and lamina is maintained and to treat any complications.
5 Discussion

Despite being developed over 50 years ago, the OOKP enables patients to achieve vision of 6/12 or better (Liu et al., 2008) and can last up to 20 years (Falcinelli, Falsini, Taloni, & Colliardo, 2005). The durability of the OOKP is due to the preservation of lamina (Falcinelli et al., 2005). Lamina volume is important as it provides structure for the OOKP. Loss of lamina volume by resorption can cause changes in visual acuity, aqueous leak and endophthalmitis (Zarei-Ghanavati, Avadhanam, Vasquez Perez, & Liu, 2017). Lamina resorption rates can be up to 28% (Falcinelli et al., 2005; Tan, Tan, & Mehta, 2012). Methods to reduce resorption include bone augmentation using mandibular bone grafts (Iyer et al., 2015).

Other complications associated with the OOKP include basement membrane graft problems such as mucosal overgrowth, glaucoma (Liu et al., 2005) and retinal detachment (Hughes et al., 2008). The prompt treatment of these complications relies on close follow-up and patient recognition of signs and symptoms to preserve vision.

Nonetheless, synthetic laminae for the OOKP is currently being investigated. Preliminary reports have demonstrated promotion of cell adhesion and growth (Avadhanam, 2016).

6 Conclusion

This case report demonstrates the primary indication for OOKP and aims to increase awareness of the treatment option. It has the potential to dramatically improve the visual acuity in patients resulting in a better quality of life. However, these patients will experience a limited visual field, and complications of the procedure include glaucoma, retinal detachment and lamina resorption. Therefore, the procedure is only amenable in patients that have no other options of treatment. Life-long, meticulous, follow-up is required to detect these early and offer potential treatments.

7 Learning points

- Common medications can cause significant adverse events.
- Severe dry eyes can, if progressive and left untreated, lead to corneal perforation and blindness.
- OOKP is a viable treatment option for severe end-stage corneal blindness when other options have failed or are unlikely to be successful.

Author statements

Conflicts of interest statement

No conflicts of interest have been declared by any authors.

Authorship statement

All authors fulfill ICMJE authorship criteria, which can be accessed at http://www.icmje.org/recommendations/browse/roles-and-responsibilities/defining-the-role-of-authors-and-contributors.html. All authors have read and approved the final version, and accept responsibility for information published.

Ethics statement

Authors declare that no ethical approval was required for this article.

Editorial and peer review statement

The review process for this manuscript was double blind, where authors and peer reviewers were blinded to each others identity and institution.

Open access and distribution statement

Authors agree to open access and distribution by CC BY Attribution 4.0, which can be accessed at https://creativecommons.org/licenses/by/4.0/deed.ast

References


