Sutureless amniotic membrane transplantation KID syndrome

SOUTHERN COLLEGE OF OPTOMETRY

Breanne B. McGhee, O.D, MEd

Bond Wroten Eye Clinic, Denham Springs, LA

Background

Keratitis ichthyosis deafness (KID) syndrome is a rare inherited autosomal dominant genetic disorder that results from mutations in the GJB2 gene coding for connexin 26 [1,2,3]. The syndrome is characterized by keratotic ectodermal thickening, sensorineural hearing loss, and vascularizing keratitis. This syndrome predisposes patients to squamous cell carcinoma and increased incidence of bacterial, viral, and fungal infections [2,4]. Individuals with KID may become symptomatic for ocular pain, increased photophobia, and abnormal corneal neovascularization and scarring which may ultimately result in decreased vision or blindness [3].

Fig 1

This photo depicts the dermatological manifestations of a KID syndrome including thickened "leather-like" plaques of skin (erythrokeratoderma) that are characteristically located on the forehead, cheeks, elbows, knees, face and scalp. [1]

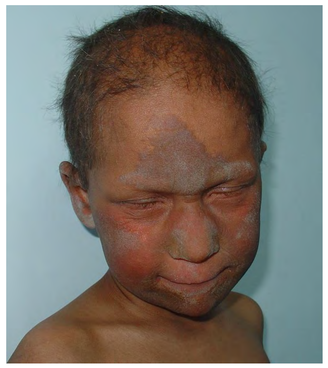
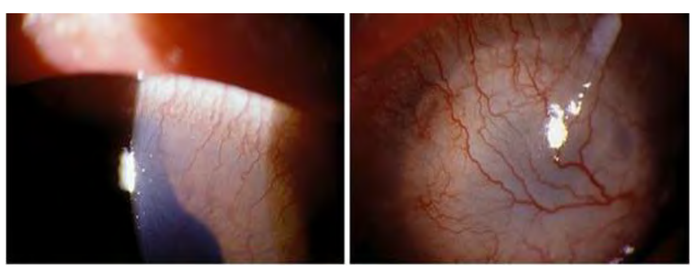


Fig 2



Possible corneal signs secondary to KID syndrome may include vascularization keratitis and opacification. [1]

Case Report

An 18 year old Hispanic female presented to clinic with her parent for a vision exam with concerns of ocular pain, photophobia and stable reduced vision. Medical history revealed a prior diagnosis of keratitis ichthyosis deafness (KID) syndrome that was confirmed by prior positive genetic testing for the disorder. Dermatological observation in office revealed epidermal thickening with multiple irregular erythematous rashes encasing her entire body, which are clinical findings consistent with the syndrome. Mild hearing loss with support of hearing aids was reported. Unaided acuities were worse than 20/400 OD, OS (distance) and 20/50 OU (near) at a short working distance. Slit lamp biomicroscopy revealed bilateral vascularized keratitis with moderate stromal haze impacting the visual axis.

Sutureless amniotic membrane transplantation via ProKera was inserted in the office under topical anesthesia and replaced twice in both eyes over for a course of 2 weeks with concomitant use of topical antimicrobials and antibiotics. Multiple follow ups were warranted to ensure absorption of the membrane, safety and efficiency, and no occurrence of ocular pathological insults. Sodium fluorescein with cobalt blue filter during slit lamp assessment was used to evaluate the progression of epithelial healing. A high oxygen permeability therapeutic bandage contact lenses replaced the ProKera lens after the amniotic tissue dissolved to continue to promote corneal healing and ocular relief.

Discussion

Amniotic membrane therapy consists of utilizing inner avascular connective tissue of the placenta, which is composed of bioactive materials and components including fibronectin, laminin, keratocyte growth factors, cytokines to all collaborately work toward reepithelialization, reduce inflammation and inhibit neovascularization [6]. There are two types amniotic membrane methods available including a cryopreserved version (ProKera/BioTissue) and a dehydrated preparation (AmbioDisk/IOP Ophthalmics), both of which are available in various thicknesses, both products are available in a range of thicknesses [6,8].

As a result of sutureless amniotic membrane transplantation therapy via ProKera, the corneal surface defects reepithelized successfully and stromal inflammation reduced ultimately improving visual acuities to 20/60+2 OD, 20/50-2 OS. Corneal integrity remained stable 3 months following treatment. ProKera lenses are easy to use in office and effective in aiding good anterior ocular health in various surface disorders.

References

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