Ophthalmic Manifestation; Corneal Perforation and its Management in a Saudi Child with Dandy-Walker Syndrome (DWS) -A Case Report

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ABSTRACT

Dandy-Walker syndrome (DWS) has vermis agenesis or hypoplasia and cystic expansion of the fourth ventricle. Ophthalmic manifestations include nystagmus, cataracts, choroidal or iris coloboma, microphthalmia, palpebral ptosis, hypertelorism, strabismus, and hydrocephalus-related optic neuropathy. However, DWS with corneal perforation due to exposure keratopathy has not been reported. The author presents corneal perforation (CP) in the right eye of a child with DWS managed by cyanoacrylate glue and a multilayer amniotic membrane. The left eye had corneal thinning, and severe dry eyes were treated with tarsorrhaphy and punctual plugs. During a seizure, tarsorrhaphy opened and corneal perforation recurred, which was treated with bandage contact lenses and lubricating eye drops.

Keywords: Dandy-Walker syndrome, Corneal perforation, Cyanoacrylate glue, Amniotic membrane graft, Bandage contact lens.

INTRODUCTION

Dandy-Walker syndrome (DWS) is a rare disorder with an incidence of 1 in 25,000–35,000 live births in the United States. It is characterized by malformation of the posterior cranial fossa, anomaly vermis agenesis or hypoplasia, and cystic expansion of the fourth ventricle.[1,2] With the help of radio imaging, changes in the brain can be described in detail.[1] Ophthalmic manifestations of DWS include nystagmus, cataracts, choroidal or iris coloboma, microphthalmia, palpebral ptosis, hypertelorism, strabismus, and hydrocephalus-related optic neuropathy.[3,4] DWS is linked with partial trisomy of the 13q location on the chromosome.[5] DWS has been reported in Saudi children, but this may be the first case with ocular manifestations of corneal perforation due to exposure to keratopathy.

Case report

A 15-month-old Saudi girl suffering from DWS had developmental delayed milestones, spastic quadriplegia, and a medical history of hydrocephalus and seizure managed at the intensive care unit. She was referred for ophthalmic consultation with complaints of right eye redness and watery eye discharge. She had a broad forehead, sparse eyebrows, epicanthal folds, down-slanted palpebral fissures, deeply set eyes, incomplete eyelid closure, low-set ears, tongue bifurcation, and teeth malformation.

The ocular assessment revealed a 2 mm scalarization of the cornea, lagophthalmos, bilateral conjunctival redness, and watery discharge from the right eye. A portable slit lamp (Keeler-PSL-Portable-Slit-Lamp-Kit, UK) was used to examine the anterior segment of the eyes. The right eye showed corneal perforation 1.5 mm below the center of the cornea with a shallow anterior chamber. The left eye had a clear cornea with a low tear meniscus and punctate erosions on the cornea and conjunctiva. The visual acuity and intraocular pressure could not be assessed due to the child's poor cooperation. The pupil was normal in size and reacting to the light. The posterior segment evaluation by binocular indirect ophthalmoscopy revealed no abnormality in the retina, optic disc, or vitreous.

Under a strict aseptic environment, the closure of corneal perforation was performed using cyanoacrylate glue and a multilayer amniotic membrane. After one month, the child was reevaluated, and authors noted a watery discharge from the right eye. (**Figure: 1**) There was corneal perforation 6 mm paracentral with incarcerated iris. The patient was admitted and underwent a corneal tectonic graft operation. Three weeks after the corneal transplant, the tectonic graft was clear. The left eye had corneal opacity inferolateral with 20% corneal thinning and severe dryness of the cornea and conjunctiva of both eyes. Bilateral lateral one-third permanent tarsorrhaphy was carried out, and the lower lids puncta were closed with fibrin plugs.

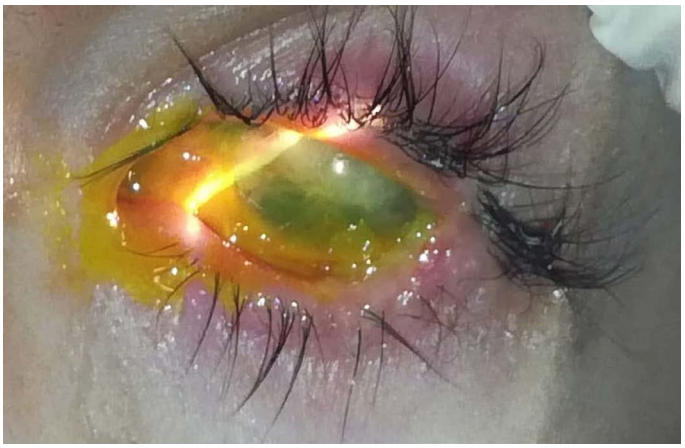

Figure 1. Negative Fluorescein staining of the cornea in the right eye of a child with Dandy-Walker Syndrome (DWS) after closure of the corneal perforation with cyanoacrylate glue and preserved amniotic membrane.

After one month of tarsorrhaphy, the child had a seizure at home. The tarsorrhaphy opened partially in both eyes. On examination, the left eye showed a 4 mm paracentral corneal perforation. It was closed with tissue-adherent glue, multilayered amniotic membrane, and

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sutures. Bandage contact lenses were applied to both eyes. The child's eyes were treated with lubrication, topical moxifloxacin, and topical prednisolone 1%, which was tapered after two weeks.

DISCUSSION

This is a child with ocular complications of DWS. The slanting palpebral fissures and lagophthalmos caused severe dry eye and exposure keratitis. The corneal thinning and perforation in both eyes needed urgent interventions. The initial treatment of sealing the corneal perforation and using an amniotic membrane enabled us to stop the leaking wound. However, seizures caused perforation at the thinned cornea in the fellow eye and opened the tarsorrhaphy; as a result, a therapeutic graft had to be performed. This shows how ophthalmologists, neurologists, pediatricians, and parents needed to work together to manage DWS complications that already had prolonged care in the intensive care unit.

Synthetic glue, like cyanoacrylate adhesives and polyethylene-glycol-based hydrogel sealants, are useful in acute perforations but have toxicities to corneal endothelium and are often replaced by biological adhesives such as fibrin glue. Their benefits, mode of application, and limitations are described in detail.[6] Managing CP in eyes with dry eye diseases is challenging, as seen in the present case. The preferred practice pattern and guidelines are worth following while managing such cases.[7] Long-term care could utilize prosthetic replacement of the ocular surface ecosystem treatment.[8]

Eighty percent of DWS cases are detected in the first year of life. However, the mortality rate ranges from 10% to 66% and reduces with increasing age and care in hospitals.[9] Therefore, it is recommended that patients follow up with a team of health experts periodically for the rest of their lives.

Some DWS cases have been identified in adults accidentally and without symptoms and confirmed with radio-imaging investigations and genetic evaluation.[10] Intrauterine detection of DWS during the first trimester of pregnancy by sonography and genetic evaluation is recommended for early intervention.[11] In the Saudi population, the most frequent birth defects include cardiac, genitourinary, craniofacial, and nervous system defects, including DWS.[12,13] Health-care professionals should be vigilant for such malformations and request an ophthalmic opinion once such a case is diagnosed.

CONCLUSION

This case of DWS had down-slanted palpebral fissures and lagophthalmos, which caused loss of tears, dry eyes, redness, discharge, chronic corneal abrasion, corneal thinning, and corneal perforation. The management was challenging and required multiple surgeries and lifelong use of lubricating eye drops and monitoring.

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Competing Interest: None

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