CASE REPORT

AWARENESS TO PROLAPSE OF THE EYE IN BABY

Athi Purnasari¹, Ratna Doemilah¹, Ismi Zuhria¹

¹ Ophthalmology Department Airlangga University School of Medicine - Dr. Soetomo General Hospital
Jl. Mayjen. Prof. Dr. Moestopo No.47 Surabaya, Indonesia
Corresponding Author : Athi’ Purnasari (athipurnasari@yahoo.co.id)

ABSTRACT

Corneal perforation may be associated with prolapse of ocular tissue. We present a rare case report an 11 months-old female presented prolapsed of right eye. She was earlier diagnosed with right corneal ulcer and descemetocele. Physical examination revealed perforated cornea. On ultrasound examination showed vitreous haziness and membranous opacity assumed due to inflammation. Based on examinations we suspected malignancy process and performed enucleation with autogenous Dermis Fat Graft (DFG) which harvested on thigh region. Histopathology examination revealed chronic inflammation process. Hence, determining the causative factors of the prolapsed eye is the key for better management and prognosis.

Keywords: corneal perforation, enucleation, dermis fat graft

INTRODUCTION

Corneal perforation cause ocular morbidity and visual loss. It is the end result of various infectious and noninfectious disorder. Corneal perforation needs immediate treatment in order to preserve the anatomic integrity of the cornea and prevent further complications such as secondary glaucoma and endophthalmitis. Corneal melting and subsequent perforation is a classic feature of corneal ulcers which do not respond to medical therapy. Thus, a surgical approach is required for most corneal perforations. (Jhanji, et al, 2011).

Descemet’s membrane is an essential barrier to microorganisms. When stromal layer melts away, Descemet’s membrane bulges forward and forms a descemetocele. Corneal ulceration leading to descemetocele and corneal perforation requires prompt treatment to prevent significant visual morbidity. Failure to diagnose this condition can result in further corneal damage, loss of anterior chamber integrity, endophthalmitis, glaucoma, profound visual loss and ultimately loss of the eye (Jhanji, et al, 2011).

In case of severe keratitis, enucleation can be the treatment necessary to prevent further spread of infection. The indication of enucleation are nonhealing microbial keratitis in a blind eye, endophthalmitis, panophthalmitis, and corneal perforation that cannot be treated with gluing or keratoplasty (Hongyok & Leelaprute, 2016).

We report, 11 months-old female with earlier diagnosed with right eye corneal ulcer and descemetocele and presented in emergency ward with prolapsed of right eye. Enucleation and using autogenous Dermis Fat Graft (DFG), an orbital implant for volume replacement post
enucleation from thigh region of patient, had been done to the patient.

CASE REPORT

We report the case of an 11-month-old female who presented in emergency ward Dr. Soetomo General Hospital Surabaya with prolapsed of right eye, and we have got consent to present this case. Heteroanamnesis suggested a reccurent reddish on right eye and whitening of cornea since 3 months ago. Previously, she was diagnosed with right eye corneal ulcer and descemetocele. Family history didn’t indicate same condition in his family.

On general examination we noticed all vital signs were within normal limit. On ocular we found visual acuity of both eyes were difficult to be evaluated. Anterior segment examination revealed perforated cornea (Figure 1). Laboratory and Chest X-Ray results were within normal limit. Ultrasound examination showed vitreous haze and membranous opacity assumed due to inflammation (Figure 2).

Based on all examination we suspected malignancy process and performed enucleation and using autogenous Dermis Fat Graft (DFG) an orbital implant for volume replacement post enucleation from thigh region of patient. The procedure began with identification of the rectus muscles, and then sutured it with vicryl 6.0 (Figure 3). After identifying the rectus muscles, enucleation procedure was done (Figure 4). For the implant, we prepared dermal fat graft from the left thigh region (Figure 5). Next step would be attaching the graft to the rectus muscles. The implantation process ended by closing tenon’s capsule conjunctiva with vicryl 6.0 and placing conformer (Figure 6). Histopathology examination was done to determine definitive diagnosis.

No operative or early complications were observed. There were no complications involving the graft donor site. Histopathology examination showed chronic inflammation process. A month after the procedure, the patient was evaluated (Figure 7). Eight weeks after the procedure ocular prosthesis fitted and resulted in a good cosmetic (Figure 8).

Figure 1. Perforated right cornea
Figure 2. USG examination showed vitreous haze and membranous opacity assumed due to inflammation

Figure 3. Rectus muscles identification, suture with vicryl 6.0

Figure 4. Enucleation Procedure

Figure 5. DFG From Left Thigh

Figure 6. Attach rectus muscle to DFG, close tenon’s capsule conjunctiva with vicryl 6.0, place conformer and pressure patch
DISCUSSION

Eye redness in children can be due to number of different underlying causes, such as Viral conjunctivitis, bacterial conjunctivitis, corneal abraction / trauma, subconjunctival haemorrhage, blepharitis, periorbital cellulitis, allergic eye disease, corneal ulcer, uveitis, and psocleritis. Careful history taking and examination with direct opthalmoscope can possibly distinguish either the diseases are benign or a early sign of a serious cases. Occasionally, a more worrying condition may present and red flags such as staining of the cornea, corneal haze / opacity, abnormal red reflex, photophobia / significant pain, lid swelling / blepharospasm, associated new-onset squint, reduced visual acuity, failure to improve by 2 weeks, contact lens use (older children), abnormal pupillary reaction, difficulty in examination, history of high-velocity; can guide clinicians as to whether further urgent referral to secondary care (Rainsbury et al.2016). One should not be forgotten is assessing red reflex in children. Abnormal red reflex represents some pathologies including scaring or infection of the cornea, cataract or opacities within vitreous, or retinal pathology such as tumours or neoplasm (Wong & Anniger, 2014).

The most frequent neoplasm of the eye is retinoblastoma, and it accounts 3% of all childhood malignancies. It is a cancer of very young; two-thirds are diagnosed before 2 years of age, and 95% before 5 years (WHO, 2014). Symptoms usually begin with leucoria (white pupil). When parents report a strange reflection in the child’s eye (red reflex abnormality) retinoblastoma should be the top differential diagnosis. The second most common sign is strabismus (misaligned eye), when central vision is lost. Advanced disease stage might present with iris colour change, recurrent eye redness, enlarged cornea and eye due to increase in intraocular pressure, or non infective orbital inflamation. In very late stage, the eye may bulge or even prolapse from the orbit. An
early approach to the disease is the key and cancer control initiative. It also has the potential great impact, both improving cure rates and minimizing the intensive treatment. (Dimaras, et al., 2015)

The treatment of retinoblastoma is multidisciplinary, aims at saving life and preserving vision, based on the extent of the disease. Early intraocular stage is curable and it is the candidate for eye preservation; in circumstances, treatment includes systemic or intraarterial chemotherapy for cryoreduction, coupled with aggressive focal therapies such as thermotherapy, brachytherapy, and cryotherapy, and external beam radiotherapy. Advanced intraocular disease requires enucleation, adjuvant chemotherapy and radiation therapy may be indicated in a subset of patients with high risk pathology. When the disease extend to extraocular, the outcome will be worsen (Dimaras, et al., 2015)

Another condition to cause eye redness is corneal ulcer. Corneal ulcers are among the leading causes of corneal blindness, which is responsible for 1.5 – 2.0 million annual new cases of monocular blindness globally. In some cases, corneal ulcers may result in the loss of eye (Hongyok & Leelaprute, 2016). Corneal ulcer that does not respond to the medical therapy can lead to the corneal perforation.

The major causes of corneal ulceration leading to corneal perforation can be boardly grouped as infectious, noninfectious (ocular surface related and autoimmune), and traumatic. Severe and recalcitrant infectious keratitis is a common cause of corneal perforation. Whereas bacterial and fungal corneal infections are frequent in the developing world, recurrent herpetic keratitis causing stromal necrosis is the major cause of corneal perforation in developed countries (Moorthy et. al., 2010). Dry eye syndrome is a major contributor to chronic epithelial defect and when it is combined with poor healing may lead to sight threatening corneal ulceration and perforation (Cohen, 1982). Collagen vascular disease such as rheumatoid arthritis, systemic lupus erythematosus, temporal arthritis, wagner granulomatosis, sarcoidosis, and inflammatory bowel disease may be associated with corneal melting (Siracuse-Lee and Saffra, 2006). Corneal trauma can result from a penetrating or perforating eye injury. Eye with previous cataract or refractive surgery are prone to corneal damage and melting following blunt trauma. Corneal melting may also occur with chemical injury. Persistent inflammation prevents epithelization and accelerates ulceration. Increase in the activity of collagenase along the ischemia leads to corneal melting and often associated with a poor prognosis (Hongyok & Leelaprute, 2016).

Corneal perforation requires prompt diagnosis and management. Most patients with corneal perforation experience a sudden drop in visual acuity. Relevant ophthalmic history includes ocular trauma, ocular surgery, contact lens use, herpetic eye disease, dry eyes, or use of topical corticosteroids. All patients should be asked about history of autoimmune disease. Patients should be instructed not to squeeze their lids. Iris prolapse is diagnostic of corneal perforation. A positive Seidel test with 2% fluorescein is also conclusive (Jhanji, et al, 2011).

The size and location of the perforation as well as the extent of stromal involvement are important parameters in determining management. Small corneal perforations may be amenable to conservative treatment with bandage contact lens or corneal gluing, whereas large perforations may require a primary repair or corneal transplantation in the form of patch graft or tectonic keratoplasty. Impending perforations may be heralded by folds in Descemet’s membrane (Jhanji, et al, 2011).

Management of corneal perforation consist of surgical and nonsurgical matters. Those nonsurgical management are: antibiotics, antivirals, antiglaucoma drug, anticollagenase,
antiinflammatory agent, and artificial tear to optimize epithelial healing. Surgical management consist of corneal gluing, conjunctival flap, amniotic membrane transplantation, and corneal transplantation (Jhanji, et al, 2011). When all of these methods are failed, the last method to prevent further damage and infection is enucleation. Enucleation procedure results in an anophthalmic socket that needs to be managed. One of the method to manage the anophthalmic socket is dermis fat graft. Dermis-fat graft is a viable option to treat volume deficiency following enucleation or evisceration both in the adults and the children, and primary grafting is generally thought to be more effective than secondary procedures. Secondary procedure usually performed on patients with fibrotic recipient beds and without direct apposition of the rectus muscles to the graft. Some disadvantages arise when using an autologous material, such as inflammation, scarring, and the potential risk of infection associated to a second surgical site. However, with dermis-fat grafts adverse effects are often predictable, and complications, such as central tissue breakdown, conjunctival inclusion cysts, or granuloma formation are rare. Excessive growth of the implant may be observed but it can be easily managed by debulking, and scarring at donor site usually gets unnoticed and tends to improve with time. Additional surgery may be performed at the same time of the graft to get better cosmetic results and manage the excessive growth. Motility of the prosthesis is found to be good. (Quantara-Leoni, et al, 2016).

CONCLUSION
Prolapsed of the eye in baby could be caused by many factors such as infection and malignancy. Consultation with other division like pediatric and histopathology division can be help to determine definitive diagnosis. Management of prolapsed eye could be performed enucleation surgery and Dermis Fat Graft (DFG) orbital implant for better cosmetic result.

REFERENCES
Cohen, K.L. 1982. Sterile corneal perforation after cataract surgery in Sjogren’s syndrome. Br J Ophthalmol;66:279-82
Dimaras, H. Corson, T.W. Cobrinik, D. White, A. Zhao, J. Munier, F.L. Abramson, D.H. Shield, C.L. Chantada, G.L. Njuguna, F. Gallie, B.L. 2015. Retinoblastoma. Nature Reviews: Disease Primer Vol 1: 6-11. Macmillian Publishers Limited.
Hongyok, T., and Leelaprute, W. 2016. Corneal Ulcer Leading to Eviceration or Enucleation in a Tertiary Care Center in Thailand: Clinical and Microbiological Characteristic. J Med Assoc Thai 2016; 99 (Suppl.2): S116-S122
Jhanji, V. Young, A.L. Mehta, J.S. Sharma, N. Agarwal, T. Vajpayee, B.R. 2011. Management of Corneal Perforation.

Survey of Ophthalmology 56(6): 533-538. Elsevier Inc.
Moorthy, S. Jhanji, V. Constantino, M. Beltz, J. Grave-Hernandez, E.O. Vajpayee, R.B. 2010. Clinical experience with n-butyl cyanoacrylate tissue adhesive in corneal perforations secondary to herpetic keratitis. Cornea; Volume 29, Number 9, September 2010
Quaranta-Leoni, F.M. Sposato, S. Ragione, P. Mastromarino, A. 2016. Dermis-Fat Graft in Children as Primary and Secondary Orbital Implant. Ophthal Plast Reconstr Surg; 32:214-219. The American Society of Ophthalmic Plastic and Reconstructive Surgery Inc.
Rainsbury, P.G. Cambridge, K. Selby, S. Lochhead, J. Red eye in children : red flags
and a case to learn from. *British Journal of General Practice*, December 2016 633
Siracuse-Lee, D and Saffra, N. 2006. Peripheral ulcerative keratitis in sarcoidosis: a case report. *Cornea*;25:618-620

Wong, M.M. and Anniger, W. 2014. The pediatric red eye. *Pediatr Clin North Am* 2014; 61(3): 591-606

WHO. 2014. Union for International Cancer Control: Retinoblasto