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Spontaneous Corneal Perforation in Premature Infants

J S Wong, V Balakrishnan, K L Tan

ABSTRACT

Spontaneous corneal perforation in premature infants not due to birth trauma is a rare event, with only 8 cases reported in the literature. We recently encountered a case of spontaneous corneal perforation in an extremely premature infant born at 26 weeks of gestation. The mechanism leading to this event was corneal exposure resulting in corneal epithelial defect. Clinical evidence of secondary infection ensued and this led to cornea thinning, descemetocoele formation and subsequent perforation with extrusion of intraocular contents. Although other mechanisms were responsible in the previously reported cases, we feel that exposure keratopathy leading to this complication poses a potential hazard to all premature infants. An awareness among care givers of this potential complication together with avoidance of exposure keratopathy in this group of patients is paramount in preventing this visually catastrophic event.

Keywords: premature infants, exposure keratopathy, spontaneous corneal perforation

INTRODUCTION

Spontaneous corneal perforation in neonates not related to birth trauma is an uncommon event, and is associated with a very grim visual prognosis. To date, only 8 cases of spontaneous corneal perforation in premature infants have been reported in the literature[1-7]. Although the exact pathophysiology is unknown, various processes had been described as possible mechanisms leading to this visually catastrophic event. We present a case encountered recently.

Department of Ophthalmology
National University Hospital
5 Lower Kent Ridge Road
Singapore 119074

J S Wong, MBBS, M Med (Ophth), FRCSEd
Registrar

V Balakrishnan, M Med (Ophth), FRCSEd, FRCOphth
Senior Lecturer

Department of Paediatrics
National University Hospital

K L Tan, FRCPE, FRACP, DCH
Professor

Correspondence to:
Dr J S Wong

CASE REPORT

An extremely premature pair of male twins were delivered vaginally at 26 weeks of gestation on 22 April 1995. Twin I weighed only 505 gramme at birth and Twin II was a stillborn. The Apgar scores were 3 and 5 at 1 and 5 minutes respectively and the baby was put on ventilatory support. The baby had a patent ductus arteriosus which did not close with 3 doses of indomethacin.

An ophthalmological consult was obtained on 7 days post-delivery because of bilateral haziness of the corneas despite measures to keep the cornea moist. Portable slitlamp examination revealed bilateral epithelial defect involving 80%-90% of corneal surfaces with purulent discharge in the left side. A corneal swab for culture was obtained. Gentamicin 8 mg/mL and cephazolin 50 mg/mL eyedrops every 2 hourly were administered to the left eye. The right eye was treated with liberal bland ointment lubricants (Duratears) application and artificial tears every half hourly. The cultures were negative.

Eleven days later (20 days of age), spontaneous extrusion of crystalline lens and vitreous prolapse from the left eye occurred. The retina showed folds and there was haemorrhage from the attachment of hyaloid vessels at the optic disc. A small corneal perforation in the centre of cornea was identified and was sealed with cyanoacrylate glue the following morning. Gentamicin eyedrops was then discontinued in view of possible corneal epithelial toxicity; cephazolin eyedrops every 6 hourly and artificial tears ointment were continued.

The perforation remained sealed with a repeat cyanoacrylate glue application 2 days later (the initial glue was dislodged). A repeat culture of corneal surface swab and vitreous content remained negative. On subsequent follow-up the right cornea epithelial defect healed and cornea became clearer. The left cornea developed total vascularisation and the eye became phthisical when reviewed at 27 days of age. Antibiotic eyedrops were stopped a week later.

Unfortunately, the baby developed methicillin resistant *Staphylococcus aureus* septicaemia at about 4 weeks of age and several days later, he developed evidence of necrotising enterocolitis. Despite aggressive treatment, he finally succumbed to the septicaemia and passed away at 6 weeks of age. Postmortem examination of the globes showed a phthisical left eye with scarring response surrounding the old corneal perforation. The right cornea did not reveal any abnormal structural abnormalities.

DISCUSSION

There are several proposed mechanisms leading to spontaneous corneal perforation in these newborns. A review of the literature showed that underlying structural abnormality such as Peter's anomaly predisposing to a weakened cornea together with increased intraocular pressure were responsible in 5 premature infants[2,5,6,7]. Epithelial defect and infectious processes leading to a thinned cornea is thought to be responsible in at least 2 cases[3,4], and

one premature baby had necrotising keratitis[1]. The timing of perforations ranged from at birth to 39 days of age.

In our case, cornea exposure leading to epithelial defect was the postulated initiating event. Normal lid disjunction occurs at about fifth to sixth months of gestation age[8], and since most premature infants are born after this period, the potential for exposure keratopathy is present in these infants. However, many of such premature infants have adequate lid closure, though one must be on the watch out for those infants who have inadequate lid closure leading to cornea exposure. With the breach of intact epithelium, the natural passive resistance to infection is lost and infective keratitis may supervene. If this is detected early, treatment with antibiotics together with adequate corneal protection may abort the process and the corneal integrity may be retained, although stromal haze and scarring may be the consequence of such an event.

However in some cases, relentless deterioration and corneal thinning leading to a descemetocoele formation may follow. This eventually leads to spontaneous perforation as seen in our case and in previously reported cases. It is possible that due to an immature immune system among the premature infants, natural immune mechanisms in the tear film such as the IgA level, the tear composition, or the tear production be deficient or altered. This may result in the susceptibility of this special group of infants to severe ulcerative keratitis. In our case, the diagnosis of infective keratitis was made clinically, as no positive organisms were identified. Postmortem histological examination did not reveal any underlying structural abnormalities in the adjacent cornea tissue that may weaken the cornea and hence predispose it to spontaneous perforation.

We feel that neonatologists and paediatric ophthalmologists must be aware of the possibility of this potentially blinding condition in premature infants with exposure keratopathy. Avoidance of corneal exposure is paramount in preventing this condition and all neonates with poor lid closure should be administered liberal ocular lubricants round the clock. Once an epithelial defect has been recognised, aggressive treatment and close follow-up by the paediatric ophthalmologist is imperative.

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