Case Report

A Case of Congenital Corneal Anesthesia with Hypopyon in China

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Abstract

Purpose: To report a case of congenital corneal anesthesia (CCA) associated with hypopyon and tactile hypoesthesia.

Methods: Case report.

Results: A 3-year-old girl presented with redness and mild photophobia in the left eye accompanied by corneal ulcer, hypopyon, and corneal neovascularization. Corneal sensation was reduced bilaterally. She exhibited an absence of normal response to painful stimuli. She also had extensive ulceration of the lateral borders and tip of the tongue. The left corneal biopsy revealed negative cultures for bacteria and fungi. She was treated with human amniotic membrane transplantation in the left eye. The response to treatment was good.

Conclusion: This is the first case of CCA reported in China. CCA is often misdiagnosed as infectious keratitis. Amniotic membrane transplantation is effective in repairing the severe corneal ulcer which may be associated with CCA. (***Eye Science 2012; 27:106-108***)

Keywords: congenital corneal anesthesia; corneal sensation; corneal ulcer; corneal sensitivity

Introduction

Congenital corneal anesthesia (CCA) is a rare condition that may occur in isolation but has also been associated with Riley-Day syndrome, Goldenhar syndrome, multiple somatic abnormalities, and MURCS. CCA is often misdiagnosed and mismanaged[5]. Poor vision, photophobia, conjunctival injection, and corneal ulceration in the absence of pain and distress in a child should alert the clinician of the possibility of CCA; however, we are now reporting a case of bilateral CCA that was initially misdiagnosed.

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Case report

A 3-year-old girl presented to the Ophthalmology Department of Eye & ENT Hospital with red eye for two months. Her mother mentioned that she had a white lesion in the left eye, which had caused mild photophobia but no other discomfort. She was recently diagnosed with suppurative keratitis on her left eye with unknown origin, and she received topical treatment of gatifloxacin, tobramycin, and mydrin in the local ophthalmology department.

In the visual acuity test, the patient lacked cooperation. Slit lamp examination showed the conjunctivae of the left eye were moderately and diffusely injected. There was an oval-shaped corneal ulcer with hypopyon and neovascularization (Figure 1A). Cornea stained with fluorescein showed multiple epithelial defects in the right eye, accompanying a patch of superficial stromal opacity. The patient was given antibiotic eye drops and lubrication; however, her condition failed to respond to the treatments. Corneal thinning and descemetocele developed. Until that time, the patient was suspected to have neurotrophic keratitis. Corneal sensation checked with the cotton tip was reduced in both eyes (Figure 2A-B). Further history inquiry revealed she had a history of repeated traumatic injuries, including bruise and empyrosis. Extensive ulceration of the lateral borders and tip of the tongue were noticed. Neurological and pediatric examination ruled out other systemic disorders. The patient was finally diagnosed with congenital corneal anesthesia localized to the eyes. Amniotic membrane transplantation was performed. Antibiotic eye drops and preservative-free artificial tears were given after surgery. Corneal scraping for bacterial and fungal smear and culture during the operation did not show

any pathogens. The corneal lesion in the left eye of the patient showed a promising response to this therapeutic regimen, with the corneal ulcer healed within 10 days (Figure 1B).

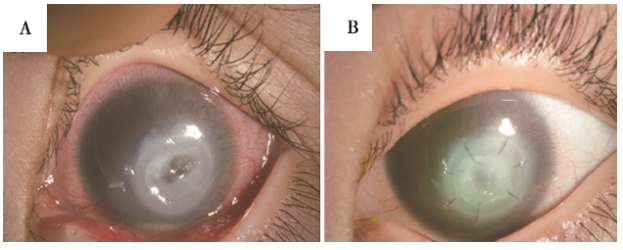

Figure 1 A.Slit lamp photograph of the left eye showing a superficial stromal ulcer, and then followed by hypopyon and corneal neovascularization. B.Photograph of the same eye after surgery (note the human amniotic membrane, the hypopyon was absorbed completely and the corneal ulceration healed gradually).

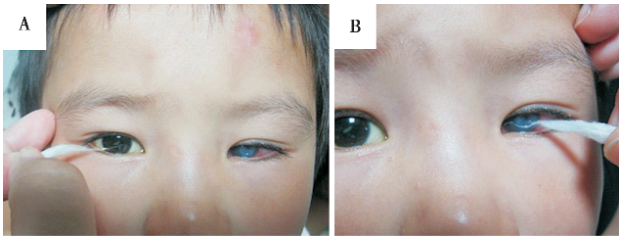

Figure 2 A and B. Photograph of eyes showing corneal sensitivity were reduced bilaterally, a reduction was noted on stimulation with a wisp of cotton.

Discussion

To The present authors' knowledge, this is the first case of CCA reported in China. CCA is a rare disorder and commonly misdiagnosed. Due to a lack of understanding of the disease, CCA is often misdiagnosed as infectious keratitis. In the present case, the patient was initially misdiagnosed with infectious keratitis because of the hypopyon. However, the corneal tissue culture was negative for both bacteria and fungus. Decreased corneal sensitivity was the symptom directing us to the diagnosis of CCA.

According to Rosenberg, CCA can be classified into three groups[1]:

Group Ⅰ: Isolated CCA without systemic anomalies.

Group Ⅱ: CCA Associated with embryological dysplasia, such as Goldenhar syndrome, Mübius syndrome, congenital insensitivity to pain, Riley-Day syndrome (familial dysautonomia, methacholine-induced miosis, alacrimia, corneal hypesthesia, and/or exodeviation), VACTERL association (cardiovascular, renal, anal, tracheoesophageal, vertebral, and limb defects), MUCUS association (Müllerian duct aplasia, cervical somite dysplasia, and renal aplasia).

Group Ⅲ: CCA associated with focal brainstem symptoms without mesenchymal and somatic anomalies.

Simple CCA is normally presented without systemic anomalies. In the present case, the patient was characterized by insensibility of harmful stimuli such as pricking, patting, or heat, although the sense of acute pain remained. However, the patient was ruled out of systemic anomalies by pediatric and neurologic examinations in part of the decreased sensitivity to pain manifested as repeated traumatic injuries. Thus, the case was classified as Group I CCA.

Proper treatment of CCA has been proposed by clinicians. These include lubrication, artificial tear, therapeutic contact lens, amniotic membrane graft, and tarsorrhapy[2]. Amniotic membrane graft was considered in this case because of the patient's severe stromal thinning and descemetocele. Post-operative effect of human amniotic membrane transplantation was remarkable in the present case, as seen by the dramatic reduction in ocular inflammation through corneal epithelialization. Human amniotic membrane can promote migration and adhesion of epithelial cells, preventing corneal epithelial apoptosis and neovascularization[3]. Furthermore, amniotic membrane can secrete various nerve growth factors, including acetylcholine and substance P, which nourish corneal nerves and facilitate corneal epithelialization[4]. Although the corneal ulcer was healed rapidly, a corneal scar formed, which made the patient's vision rehabilitation another challenge due to the poor prognosis of corneal grafts. Recurrence is often recorded with CCA, so we advised the patient to undergo long-time follow up.

Mild CCA is characterized by punctuate, persistent, and non-healing corneal epithelial defects. In severe disease progression, patients may develop into non-bacterial corneal ulceration, even perforation in our present case[5]. False diagnosis of CCA will

lead to inadequate treatment such as penetrating keratopathy, resulting in serious complications to the patient[6]. Therefore, it is crucial to provide accurate diagnosis and proper treatment, which can alleviate decreased visual acuity and prevent serious complications.

References

1 Rosenberg ML. Congenital trigeminal anaesthesia.A review and classification. Brain, 1984; 107 (Pt 4): 1073-1082.

Hewson GE. Congenital trigeminal anesthesia. Br J Ophthalmol,1963;47:308- 311.

2 Tsai RJ, Li LM, Chen JK. Reconstruction of damaged corneas by transplantation of autologous limbal epithelial cells.N Engl J Med,2000;343(2):86-93.

3 Dua HS, Azuara-Blanco A. Amniotic membrane transplantation.Br J Ophthalmol, 1999;83:748-752.

4 Ramaesh K,Stokes J,Henry E,et al.Congenital corneal anesthesia.Surv Ophthalmol, 2007; 52(1):50-60.

5 Reed JW, Joyner SJ, Knauer WJ. Penetrating keratoplasty for herpes zoster keratopathy. Am J Ophthalmol, 1989;107(3):257–261.