Ligneous conjunctivitis in a Dandy–Walker syndrome: A rare case report

Arjun Srirampur, Muralidhar Ramappa, Sunita Chaurasia, Geeta Vemuganti[1]

Ligneous conjunctivitis (LC) is a rare form of pseudomembranous conjunctivitis seen in children, perhaps due to plasminogen deficiency, which manifest as a chronic refractory pseudomembranous conjunctivitis. LC cases are incapable in maintaining their fibrinolytic activity due to plasminogen deficiency; consequently, transudates of plasma assume as a thick, gelatinous, woody membranes over the mucosal surfaces. This is a short case report on a child with a LC, who presented with recurrent pseudomembranous conjunctivitis in conjunction with progressive congenital hydrocephalus due to aqueductal stenosis (Dandy–Walker syndrome). This rare association was clinically confirmed and prompt corrective surgical measures were instituted.

Key words: Dandy–Walker syndrome, hydrocephalus, ligneous conjunctivitis

Ligneous conjunctivitis is a rare form of chronic pseudomembranous conjunctivitis characterized by the development of fibrin-rich, woody-like pseudomembranous lesions mainly on the tarsal conjunctiva.[1] Less frequently, similar lesions may occur on other mucosal surfaces signifying underlying systemic disease, perhaps due to plasminogen deficiency.[2] Therefore, these eyes are prone to develop pseudomembranes over the mucosal surfaces, either spontaneously or following a trivial trauma. Basic pathogenesis of LC is thought to be due to impaired wound healing of bruised mucosal surfaces due to ineffective extracellular fibrinolysis. Histopathology of such peeled of membranes does suggestive of residual by-products of clotted fibrin.[3] Herein, we describe a child who presented with a rare clinical association of LC with a congenital hydrocephalus, conceivably due to aqueductal stenosis.

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Department of Cornea and Anterior Segment, Kallam Anji Reddy Campus, Tej Kohli Cornea Institute, LV Prasad Eye Institute, [1] School of Medicine, Hyderabad University, Hyderabad, Telangana, India

Correspondence to: Dr. Muralidhar Ramappa, Kallam Anji Reddy Campus, Tej Kohli Cornea Institute, LV Prasad Eye Institute, Banjara Hills, Hyderabad - 560 034, Telangana, India. E-mail: muralidhar@lvpei.org

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Case Report

A 10-month-old infant was referred to us by a local ophthalmologist for further evaluation with the suspected clinical diagnosis of chronic conjunctivitis. The child first presented to us with parents describing swollen lids, mucus discharge from both eyes, and gradual enlargement of head since birth.

Prenatal USG at 34th week of gestation showed dilated 3rd and 4th ventricle with a posterior fossa cyst, communicating with 4th ventricle indicative of Dandy–Walker syndrome [Fig. 1a and b].

During newborn physical assessment, the primary care pediatrician had noted an abnormal head size and referred the child for a thorough neurological examination. Neuroimaging did confirm the diagnosis of Dandy–Walker syndrome for which child then had undergone a right-sided ventriculoperitoneal shunt. Family history was negative for known genetic disorders. Family history revealed a 2nd degree parental consanguinity and his four elder male siblings and two still born diagnosed to have hydrocephalus on prenatal USG scans had a similar manifestation of swollen lids and hydrocephalus at birth; eventually all of them expired due to the neurological complications at the age of 1–2 years.

General physical examination revealed a healthy appearing baby with an enlarged head [Fig. 2a] with a palpable subcutaneous tube from head to abdomen. The anterior fontanel was soft and appeared enlarged in size for his age. Rest of the systemic examination was within normal limits.

Anterior segment eye examination under general anesthesia revealed markedly swollen lids. On eversion of the lids, the conjunctiva showed grayish white, thick, gelatinous, pseudomembranes with woody consistency adherent to the tarsal conjunctiva not attached to the bulbar conjunctiva or cornea, more in right eye then left [Fig. 2b and c]. Both eyes cornea showed few superficial punctuate keratitis. Rest of the anterior segment was within normal limits. The intraocular pressure and fundus examination were within normal limits in both the eyes.

The child was diagnosed with LC with congenital hydrocephalus with a ventriculoperitoneal shunt in place. This prompted us to get a serum plasminogen levels and a referral to a clinical hematologist for evaluation. The hematological examination was consistent with a clinical diagnosis of plasminogen deficiency.

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Surgical details

Diagnostic conjunctival swabs were obtained and sent for microbiological workup prior to membrane removal. Then, pseudomembranes were gently peeled off from the mucosal surfaces meticulously and minimal bleeding was noted after the removal, which was controlled with pressure mops. The underlying tarsal conjunctiva was healthy with no evidence of necrosis. The peeled off membranes were sent for histopathological examination. The above clinical findings and ease of membranes peeling with minimal bleeding further confirmed our clinical diagnosis of pseudomembranous conjunctivitis.

Postoperatively, the child was put on topical tobramycin 0.3% eye drops every 4 hourly, prednisolone acetate 1% eye drops 4 times a day, and hydroxyl propyl methyl cellulose eye ointment 2 times a day. Our plan was to start topical heparin and cyclosporine combinations after the confirmation of diagnostic microbiology and histopathology reports. Conjunctival diagnostic swabs were remained sterile at the end 1 week. Histopathological findings showed a thick, amorphous, hyaline-like eosinophilic material suggestive of by-products of fibrin and inflammatory membranes of the conjunctiva compatible with LC. Child was started on combination of topical steroids; heparin and cyclosporine eye drops were effective in minimizing the relapses. Parents were advised to continue the protective goggles and the importance of long-term follow-up. When child was last seen, he was doing well and thereafter he was lost to follow-up.

Histopathology

The peeled pseudomembranes sent for histopathological examination with eosin and hemotoxylin stains showed thinned and eroded epithelium with subepithelial deposits of hyaline-like eosinophilic material containing clotted fibrinogen, immunoglobulins, and albumin [Fig. 3a]. In addition, foci of granulation tissues with lymphocytes, plasma cells, and granulocytes were seen [Fig. 3b].

Discussion

The plasminogen deficiency is a common cause of LC,[4] and it is inherited as an autosomal recessive disorder, with low plasminogen levels reported in both homozygous and compound heterozygous individuals.[5] Minger *et al*. had elegantly reported a case of severe LC in association with congenital hydrocephalus in a homozygous plasminogen deficiency.

LC is seen frequently in children, typically characterized by presence of pseudomembranes that are woody in consistency on the palpebral conjunctival surface. In addition, patients may have dense mucoid discharge from affected eyes. These symptoms may be accompanied by fever, upper respiratory tract infection, urinary tract infection, or genital tract infection in the acute period. LC appears to be the ocular manifestation of a systemic disease, which might be accompanied by formation of pseudomembranes on the mucosa of the mouth,[6] ligneous gingivitis or peridontitis, the tracheobronchial tree,[7] the kidneys,[8] and the female genital tract[9] (ligneous vulvovaginitis or cervicitis). Furthermore, in sun-exposed skin areas, juvenile colloid milium may be present.[10] Some patients additionally suffer from congenital occlusive hydrocephalus.

The management of LC is quite challenging. Simple pseudomembrane removal leads to recurrence unless necessary measures are taken. The most promising approach seems to be the initial application of a topical fibrinolytic agent followed by surgical removal of the pseudomembranes with subsequent intense and long-term topical application of heparin. Heparin appears to reduce the otherwise high risk of local recurrence after surgery alone.

In this context, the key challenge in the management of LC is the prevention of ocular trauma, which relies on early diagnosis of systemic underlying blood dyscrasia for targeted measures aimed at preventing systemic complications.

Although LC with congenital hydrocephalus has been documented, its association with Dandy–Walker Syndrome has not been reported in published literature.

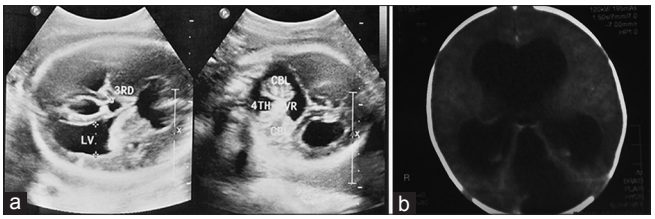

Figure 1: (a) Prenatal USG at 34th week of gestation showed dilated 3rd and 4th ventricle with a posterior fossa cyst, communicating with 4th ventricle and (b) computed tomography scan of the brain showing enlarged 3rd and 4th ventricle with a posterior fossa cyst

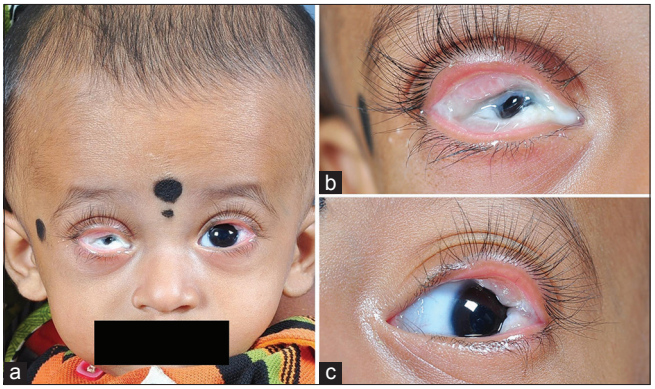

Figure 2: (a) Clinical photograph of the index patient showing enlarged head, swollen lids, and discharge and (b and c) grayish white, thick, gelatinous, pseudomembranes adherent to the tarsal conjunctiva more in the right eye than left

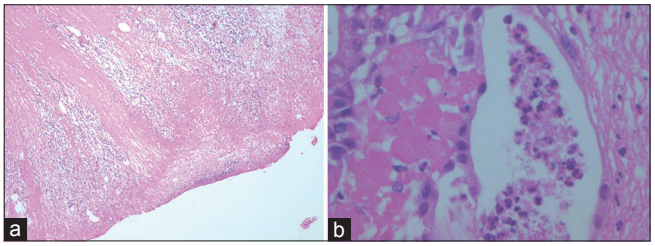

Figure 3: (a) Histopathological examination with eosin and hemotoxylin stains of the pseudomembranes showing thinned and eroded epithelium with subepithelial deposits of hyaline-like eosinophilic material containing clotted fibrinogen, immunoglobulins and albumin and (b) foci of granulation

Conclusion

We believe that LC remains an underdiagnosed rare genetic condition. Timely recognition of this clinical condition is crucial in minimizing the life-threatening sequelae. A multidisciplinary approach is necessary, proper training about these and other lifestyle measures for affected individuals, their parents, other caregivers, and school staff. Definitive molecular diagnosis allows for appropriate preventive management, including advice and aids to preclude future major consequences for quality of life.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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