Association of Ligneous Conjunctivitis with Congenital Hydrocephaly: A Case Report

Mohammad Zare, MD¹ • Mohammadreza Fallah Tafti, MD² • Fateme Zare³
Bahram Eshraghi, MD² • Fahimeh Asadi Amoli, MD³ • Mohammad Yaser Kiarudi, MD⁵

Abstract

Purpose: To report an association of ligneous conjunctivitis (LC) and congenital hydrocephalus

Case report: The patient was a 3.5-year-old boy with a history of long standing conjunctivitis with copious ocular discharge and photophobia, waxing and waning for some time. He also had suffered from occlusive congenital hydrocephalus that required placement of a ventriculoperitoneal shunt. Conjunctivitis did not respond to topical medications and recurred after several excisions. Finally an intraläsional methylprednisolone injection was performed. Significant resolution of the lesions was observed after one week and after one year, LC was relatively controlled and there was no need for more excisions.

Conclusion: In patients with recurrent recalcitrant pseudomembrane, this treatment shortens the treatment period, evokes rapid visual rehabilitation and obviates the need for future excisions. Also, this report reemphasizes the association of LC and congenital hydrocephalus, which maybe ignored.

Keywords: Ligneous Conjunctivitis, Plasminogen Deficiency, Congenital Hydrocephalus

Introduction

Ligneous conjunctivitis (LC) is a rare disease of conjunctival epithelium with development of pseudo-membranes on the palpebral surfaces and then producing thick nodular masses that replace the normal mucosa.¹ The presentation time of the symptoms are during the first weeks or months of life² and the diagnosis is made by the wood-like appearance of the membranes on the palpebral conjunctival surface. Although ophthalmic involvement is the most common manifestation of the disease, the less frequent involvement of other mucosal membranes such as larynx, vocal cords, gingiva, trachea, nasopharynx, tympanic membranes, vagina and cervix and an association with congenital hydrocephalus have been reported.³ Plasminogen deficiency has been introduced as the most common cause of LC, and the formation of membranes is the result of an impaired wound healing process⁴; LC seems to be the most common symptom of this systemic disorder.

1. Associate Professor of Ophthalmology, Eye Research Center, Farabi Eye Hospital, Tehran University of Medical Sciences
2. Assistant Professor of Ophthalmology, Eye Research Center, Farabi Eye Hospital, Tehran University of Medical Sciences
3. Resident in Pediatrics, Ali Asghar Hospital, Iran University of Medical Sciences
4. Associate Professor of Pathology, Department of Pathology, Eye Research Center, Farabi Eye Hospital, Tehran University of Medical Sciences
5. Resident in Ophthalmology, Eye Research Center, Farabi Eye Hospital, Tehran University of Medical Sciences

Received: May 8, 2010
Accepted: October 7, 2010

Correspondence to: Mohammad Yaser Kiarudi, MD
Eye Research Center, Farabi Eye Hospital, Tehran University of Medical Sciences, Tehran, Iran, Tel:+98 21 55414941-6,
Email: kiarudi@razi.tums.ac.ir, y_kia76@yahoo.com

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Simple excision of the woody membranes leads to recurrence, so different methods are recommended in several case reports in the literature\textsuperscript{5-7} although in general it is believed that there is no treatment regimen for such conjunctivitis, in this study we report a young boy with LC associated with occlusive hydrocephalus and its medical management.

**Case report**

The patient was a 3.5-year-old boy with a history of long standing conjunctivitis (Since he was 5-month-old) with copious ocular discharge and photophobia, waxing and waning for some times. He was the only child of an Iranian parents with first degree consanguinity (They were cousins). He had suffered from occlusive congenital hydrocephalus (Hydrocephaly was detected by prenatal ultrasonography) that required placement of a ventriculoperitoneal shunt at 12 months of age (Subsequently revised five times and the last time was a year ago). His hydrocephaly was controlled. On examination, there were bilateral conjunctival hyperemia, sticky discharge with a woody-like consistency and also some degrees of acquired blepharophymosis. Ocular examination under general anesthesia was performed and revealed clarity of cornea, media and no pathologic finding in posterior segment. He had frontal bossing (Figure 1). There were no otolaryngological manifestations and no genital tract involvement. The gingiva was hyperemic but not hyperplastic. Unfortunately, plasminogen functional assay and plasminogen antigen were not measurable because of our laboratory limitations. The patient had two unsuccessful excisions before referring to our clinic. Pathological analysis confirmed pseudomembranous lesions consistent with LC with each excision.

We started treatment with topical acetylcysteine 10% every 6 hours, fluorometholone every 8 hours and frequent use of preservative free artificial tears. Conjunctival hyperemia and discharges were reduced within first 2 weeks of treatment but conjunctival thickening persisted. After one month fluorometholone tapered to every 8 hours and patient was followed for 6 months. During the follow-up time patient had 2 episodes of relapses and worsening of his condition following tapering of topical medication that subsided immediately by restarting topical therapy. Finally an intralesional methylprednisolone injection (0.1 ml in each pseudomembrane) was performed. Significant resolution of the lesions was observed. The child could open his eyes and about 9 months after the injections, there was no need for more excisions (Figure 2).

![Figure 1. Thick and woody pseudomembranes, macrocephaly and frontal bossing at 6 months of age](image1.jpg)

![Figure 2. Shows resolving of pseudomembranes with favorable result after intralional injection of methylprednisolone](image2.jpg)

**Discussion**

Review of literature shows that there are a few cases of association of LC and hydrocephaly. Aslan et al stated that of total 16 patients with...
LC and hydrocephaly in literature, seven of them had plasminogen deficiency.  

Severe hypoplasminogenemia is associated with impaired extracellular fibrin clearance during wound healing which causes formation of pseudomembranous (Ligneous) lesions on affected mucous membranes. In affected patients, the level of plasminogen antigen and functional plasminogen activity are reduced. Unfortunately, we could not measure these parameters because of our laboratory limitations. Schuster et al reviewed the clinical manifestation of 74 patients with severe hypoplasminogenemia. The most common findings were LC (81%) and a minority of them had congenital occlusive hydrocephalus. The fact that pseudomembranous lesions mainly form in conjunctival mucosa rather than mucous membranes, is explained by more exposition of conjunctiva to variety of external irritants like dust, local infections, small foreign bodies, frequent scratching and minor traumas, that trigger local inflammation and pseudomembranous formation. Regarding to this hypothesis, Fuentes-Páez et al reported a 27-year-old female patient with unilateral LC and Crohn’s disease. The symptoms of patient appeared one month after a resolved conjunctival chemical burn with 17% hydrochloric acid, so they pointed out chemical injury as the triggering LC. The initial choice for treatment of the conjunctival lesion is topical therapy with plasminogen-containing eye drops with concentration of approximately 1 mg/ml. After the resolution of pseudomembranes, treatment follows with intensive topical standard heparin (1000-5000 U/ml) in combination with topical corticosteroids or alpha-chymotrypsin. Other reported treatment regimens are combination of heparin and fresh frozen plasma (FFP), topical and systemic FFP together with conjunctival membrane excision, topical proteolytic enzymes (hyaluronidase and chymotrypsin); fibrinolysin drops and recombinant tissue plasminogen activator; topical anti-inflammatory agents such as corticosteroids and cyclosporine. Plasminogen concentrate is expensive and not widely available and treatment regimens such as injection of FFP require hospitalization. We used a single depot injection of steroid (0.1 ml methylprednisolone) in each thick, woody pseudomembrane lesion. Significant resolution of membranes occurred in one week and no new membrane formation was observed after 9 months. We suggest in patients with severe recurrent recalcitrant pseudomembrane, when other treatment modalities are not available, this treatment shortens the treatment period, has rapid visual rehabilitation and provides sufficient time for clinicians to consider definite treatment. One should keep in mind that mentioned treatments are topical therapies in cases affecting only the eyes (LC). However in patients with severe hypoplasminogenemia only systemic therapies such as hormonal treatment approaches could increase functional plasminogen activity.

**Conclusion**

In this study we introduced another association of LC and hydrocephalus. Since hypoplasminogenemia, a systemic hematologic disorder, is a strong etiological factor, care must be taken to other comorbid like hydrocephalus.

References