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Case Report

Ligneous Conjunctivitis: A Diagnostic Dilemma - Case Report

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Abstract

Ligneous conjunctivitis is a rare form of conjunctivitis characterized by chronic, recurrent conjunctivitis associated with pseudomembrane formation, which may involve other mucous membranes such as gingiva, ear, respiratory tract, female genital tract and kidneys. Following is the case report of a 19-year-old girl with complaints of chronic redness and discharge, not associated with pain, itching or blurred vision. Ocular examination showed bilateral severe conjunctival congestion with mucoid discharge, membrane like lesions involving palpebral conjunctiva with no papillary or follicular reaction. Patient was initially started on topical mast cell stabilizers and steroids; topical cyclosporine added subsequently following inadequate response. Excision of the membranous material was done which showed acute inflammatory exudates and amorphous pink purple material with focal areas of calcification in between. She has had two episodes of recurrence of membrane during her 3-year follow-up but otherwise maintaining well on topical cyclosporine with steroids being added during recurrence.

Ligneous conjunctivitis should be considered a differential diagnosis of allergic and vernal keratoconjunctivitis. Inspite of lack of woody hardness of the conjunctiva, clinical findings led to the diagnosis of ligneous conjunctivitis. Definite histological diagnosis is tricky, because of lack of common histological characteristics among previously reported cases of ligneous conjunctivitis.

Keywords: *Ligneous Conjunctivitis; Pseudomembrane; Cyclosporine*

Background

Ligneous conjunctivitis was first reported in a 46-year-old man with bilateral pseudomembraneous conjunctivitis as early as 1847 [1]. Verhoeff coined the term "recurrent pseudomembraneous granuloma of the palpebral conjunctiva" [2] while Mingers., *et al*. proposed the term "pseudomembraneous disease" to describe systemic character of this clinical entity [3]. In 2001, Lecame., *et al*. estimated a total of 119 reported cases of ligneous conjunctivitis in last 50 years [4]. However, many sporadic cases of ligneous conjunctivitis may not have been published, and true incidence is not known. Familial occurrence has been reported, mainly inherited in autosomal recessive pattern [5].

Ligneous conjunctivitis is a rare form of chronic "idiopathic membraneous" conjunctivitis characterized by initial chronic tearing and redness of conjunctiva with subsequent formation of pseudomembranes mostly on palpebral surface of upper tarsal conjunctiva that progress to white, yellow-white, or red thick masses with a wood-like consistency that replaces normal mucosa; lower eyelid and bulbar conjunctiva are less frequently involved [6]. It is characterized by multiple recurrences after local excision. Corneal involvement occurs in 26 - 30% of cases and may lead to blindness as a result of scarring, vascularization, keratomalacia or corneal perforation. Infants and children are usually affected; however, disease may manifest at any age [6]. Bilateral in almost half (51%) the cases, duration of disease has been reported to range from a few months to 44 years [2,6]. It may be triggered by local injuries [7], local and systemic infections, lime burns [8], and different surgical interventions [8].

During acute phase, it may be associated with fever, infections of upper respiratory, urinary, or female genital tract that may either precede or concomitantly occur with ocular disease and may act as a trigger for development of pseudomembranes [6]. In addition, the ocular manifestations may be associated with involvement of other mucosal sites such as gingiva [6], ear [9], respiratory tract [6], female genital tract [10] and kidneys.

Histopathology shows a thinned or eroded epithelium with superficial or subepithelial deposits of amorphous hyaline-like eosinophilic material and foci of persisting granulation tissue with accompanying inflammatory cells, mainly lymphocytes, plasma cells, and granulocytes; surface ulceration may cover large areas with overlying exudate [6]. Rest of ephithelium may show hyperplastic changes that may extend irregularly into substantia propria in the form of cysts, cords, and gland like structures. In patients with ligneous conjunctivitis a serofibrineous transudate, possibly caused by an increased permeability of blood vessels [6], undergoes subsequent coagulation, with formation of granulation tissue and fibrin-rich material. This material becomes hard resulting in formation of ligneous pseudomembrane.

Cardinal feature of ligneous conjunctivitis is an impaired wound-healing capacity with arrest at the stage of granulation tissue formation and excessive fibrin deposition. Systemic lys-plasminogen application has found to restore wound-healing capacity and resolve ligneous lesions; thus, severe type I plasminogen deficiency may explain all pathologies found in ligneous conjunctivitis and associated lesions in other mucous membranes.

Case History

A 19-year-old female presented with complaints of redness OU for the past 2 years, associated with mild whitish discharge which was non-sticky and serous in nature. It was not associated with any pain or blurring of vision. There was no history of fever, joint pains or oral ulcerations, chemical or thermal injury to eyes, use of contact lens or glasses, any allergy to dust particle, or any medications used in the past.

On examination, best corrected visual acuity (BCVA) of the patient was 6/6 (Snellen) OU. There was no systemic abnormality found on physical examination. Mucoid discharge and conjunctival congestion was present in both eyes with dilated, tortuous vessels with early obliteration of inferior fornix in right eye (Figure 1,2). There was no circumciliary congestion, no papillae or follicles. A diagnosis of allergic conjunctivitis was made and patient started on mast cell stabilizer (olopatidine 0.2%) and low potency topical steroid (loteprednol 0.5%). The persistence of membrane and inadequate response prompted us to add immunomodulator (2% cyclosporine) to her treatment regimen. Initial response to this regimen was encouraging, which prompted us to taper the dosing gradually. On decreasing the frequency of medication, however patient reported with recurrence of conjunctival congestion.

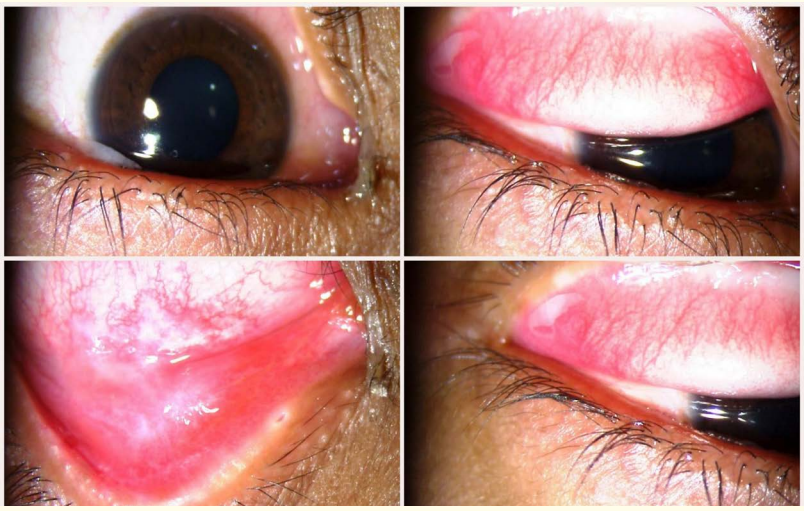

Figure 1: *Slit-lamp photograph of right eye showing conjunctival congestion and symblepharon formation in inferior fornix.*

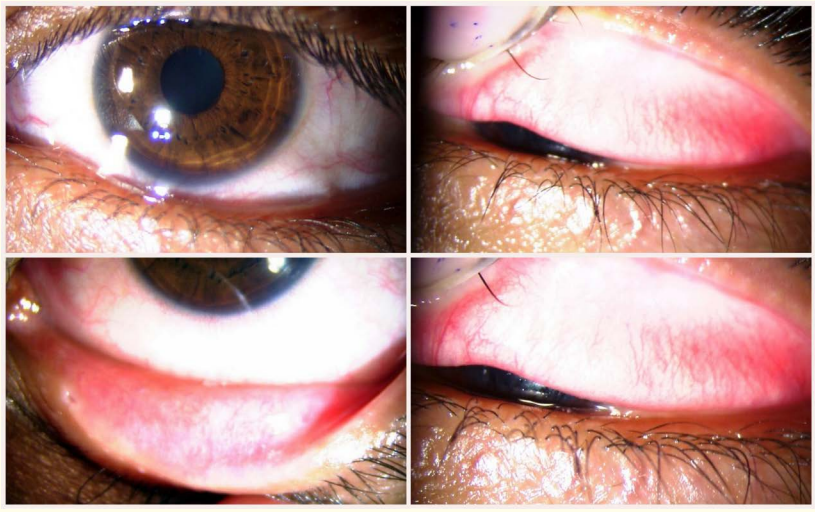

Figure 2: *Slit-lamp photograph of left eye showing severe conjunctival congestion.*

Subsequently, excision of the membranous material was done which on histopathological examination showed acute inflammatory exudates, leucocytic infiltration, amorphous pink purple material (fibrin exudation) with focal area of calcification in between (Figure 3). Thereafter, patient was put on low dose steroids and cyclosporine eye drops. Patient has had two episodes of recurrence during her 3 year follow up with dose of steroids stepped up during recurrence and conjunctivitis controlled without resorting to excision. At present, she has stable course of the disease with no complications.

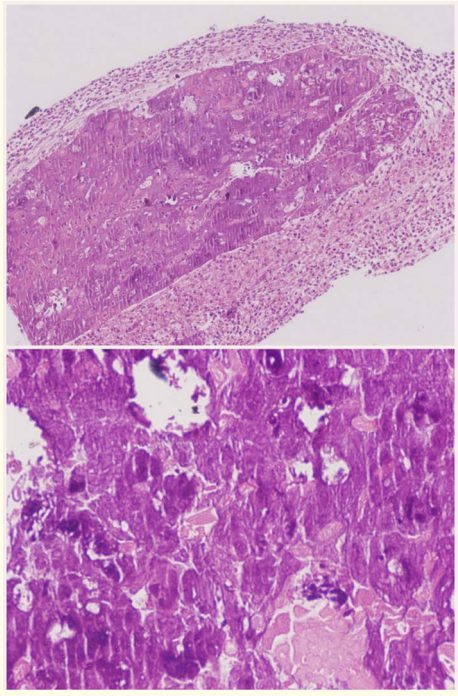

Figure 3: *Histopathological specimen of the membranous lesion showing acute inflammatory exudates and areas of calcification.*

Discussion

This case exhibited several characteristics of ligneous conjunctivitis such as recurrent pseudomembrane and response to surgical excision. Indispensable characteristics of vernal keratoconjunctivitis like itching and extensive papillary formation were not observed. Inspite of lack of woody hardness of conjunctiva, clinical findings led to the diagnosis of ligneous conjunctivitis. Definite histological diagnosis could not be ascertained, because of lack of common histological characteristics among previously reported cases of ligneous conjunctivitis [6]. The treatment of ligneous conjunctivitis includes initial topical application of a fibrinolytic agent (plasminogen concentrate) plus plasminogen activator which softens fibrin-rich pseudomembranes and facilitate removal of the membrane. To prevent regrowth, intensive local treatment with heparin and corticosteroids is recommended. If no improvement is seen, topical cyclosporine and steroids is recommended for longer duration i.e. upto 1 year.

In our case, patient showed satisfactory response to topical cyclosporine and removal of membranous material with no requirement of topical fibrinolytic agent or heparin. Though the patient was given low dose steroids in acute phase, however, long term need of steroids was not required. Thus, topical cyclosporine can be a better alternative than topical steroids, reducing the complication in the form of corticosteroid-induced glaucoma seen in some cases reported earlier.

Conclusion

Ligneous conjunctivitis should be considered a differential diagnosis of allergic and vernal keratoconjunctivitis. Although a rare disease; diagnosis may be tricky due to paucity of knowledge about histopathological characteristics. Topical cyclosporine may be an effective alternative to corticosteroids and fibrinolytic agents for its treatment.

Key Message

Although a rare disease, ligneous conjunctivitis should be considered a differential diagnosis of allergic and vernal keratoconjunctivitis; diagnosis may be tricky due to paucity of knowledge about histopathological characteristics. Topical cyclosporine may be an effective alternative to corticosteroids and fibrinolytic agents for its treatment.

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