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Case Report

Ligneous conjunctivitis: A challenging case to manage

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ABSTRACT

Ligneous Conjunctivitis is an autosomal recessive, chronic form of conjunctivitis characterized by formation of pseudo-membrane particularly over tarsal conjunctiva attributed to plasminogen deficiency. Various forms of medical and surgical treatment modalities have been reported. We are reporting a case of ligneous conjunctivitis in a nine month old baby managed successfully with both topical and intravenous fresh frozen plasma, topical heparin and cyclosporine.

1. Introduction

Ligneous conjunctivitis is a rare form of conjunctivitis with recurrent pseudomembrane formation mostly seen in childhood.[1] Plasminogen plays an important role in its etiology.[2,3] The condition was first noticed by Vongraefe in 1854 and in 1933 the condition received its distinctive name. It tends to occur in young children, mainly females and may affect other members of the same family. The most common site of ligneous conjunctivitis is the inside of the eyelids involving palpaebral conjunctiva, although it can also damage the sclera and cornea, thus impairing vision.[4] The larynx, voice chords, nose, trachea, bronchi, vagina, cervix, and gingiva may also be affected in case of systemic involvement. Besides sight threatening, it may cause mortality due to pulmonary involvement.[4,5] Various treatment options have been reported including surgical management for ligneous conjunctivitis.[5–11] Here we describe the case of a male child with ligneous conjunctivitis managed with fresh frozen plasma, topical heparin and topical cyclosporine without surgical intervention.

2. Case Report

A nine-month old baby boy, product of non-consanguineous parents, presented with recurrent bilateral eyelid swelling, photophobia, watering and whitish membrane under both eyelids noted for 2 months (Figure 1). The baby was treated elsewhere as a case of membranous conjunctivitis and membrane was removed twice. On presentation to our clinic, we removed a portion of the membrane from right upper eyelid under surface anaesthesia and sent the tissue for histopathology. The histopathological report showed mucosa lined by locally atrophic squamous epithelium with subepithelial deposits of eosinophilic material consisting predominantly of fibrin and immunoglobulin and scattered inflammatory cells. Histopathological findings were suggestive of ligneous conjunctivitis. The baby was otherwise healthy clinically, though we could not assess the serum plasminogen level due to lack of facility.

After confirming diagnosis the baby was started with topical cyclosporine (0.05%) twice daily, topical heparin (5000 IU/ml) 6 hourly and topical fresh frozen plasma 1 hourly. The baby was presented after 2 weeks with noticeable improvement and the whitish membrane was

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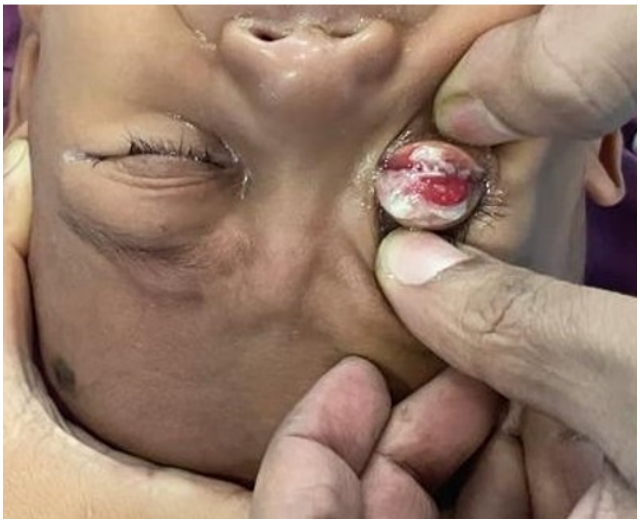

Fig. 1: Pseudomembrane formation over palpaebral conjunctiva

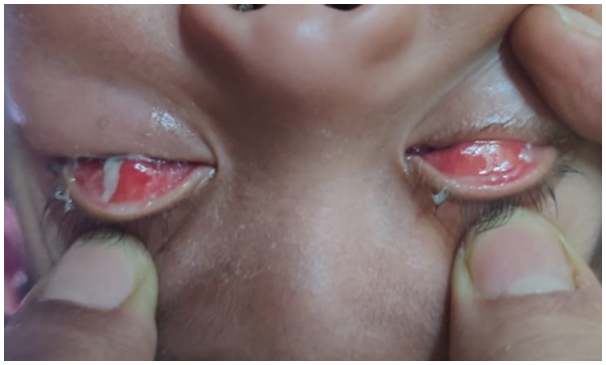

Fig. 2: Marked resolution of membrane after treatment with fresh frozen plasma

almost resolved (Figure 2). Same treatment was continued and within 6 weeks the baby was completely cured without any membrane on the tarsal conjunctiva. Topical fresh frozen plasma was administered every 2 hourly for further one month. Topical cyclosporine (0.05%) twice daily and topical heparin (1000 IU/ml) four times daily were continued as maintenance treatment. One month after cessation of topical fresh frozen plasma the membrane recurred again over tarsal conjunctiva in both eyes. At this stage, the baby was administered with intravenous fresh frozen plasma (20mg/kg) daily for 5 days, then weekly for one month and two weekly for 3 months. Topical cyclosporine (0.05%) and topical heparin (1000 IU/ml) were administered as adjuvant treatment and continued for further 8 weeks after cessation of intravenous fresh frozen plasma. The condition was completely resolved without any symptom of lacrimation and photophobia (Figure 3). The baby was without any treatment for four months till last follow up. No recurrence was noted during this follow up period.

Fig. 3: Baby looks comfortable without lacrimation and photophobia after complete resolution of membrane

3. Discussion

Ligneous conjunctivitis is a rare autosomal recessive disorder secondary to inherited homozygous plasminogen deficiency.[2] Plasminogen is a precursor molecule of plasmin, which is responsible for degradation of fibrin. The cornea is the extrahepatic site for plasminogen synthesis, controlling its concentration in tear fluid.[12] Lack of plasminogen in tear fluid leads to fibrin rich pseudomembrane formation mostly over the palpaebral conjunctiva often described as “woody” induration.

Various treatment modalities have been described in literature.[5–11] Considering plasminogen deficiency as a causative factor for ligneous conjunctivitis, plasminogen eye drop has been reported as a promising treatment option, however its use in clinical practice is limited by its cost and availability.[9,13] Alternatively, topical and systemic use of fresh frozen plasma (FFP) has been found effective.[5–8] Ocak and Bas have reported positive outcome with topical use of FFP alone in case of conjunctival involvement only.[5] However, patients with systemic involvement need to be treated with FFP transfusions. Our case had conjunctival involvement only, though it required both topical and systemic administration of FFP as there was recurrence with topical FFP alone. Systemic application of FFP may have several side effects like immunological reaction (allergy or anaphylaxis), transfusion-related acute lung injury and haemolysis.[8] However, none of these adverse reactions was observed in our patient.

Besides topical and systemic use of FFP, we have also treated our patient with topical heparin. Hiremath et al have reported long term treatment with heparin eye drop over 5 years to prevent recurrence.[10] Heparin acts by accelerating the activity of antithrombin and neutralizing factor Xa and thrombin. Hence it prevents the synthesis of fibrin which contributes to the primary pathogenesis in ligneous conjunctivitis. We started topical heparin at the concentration of 5000 I.U. per milliliter (IU/ml) and later continued at the concentration of 1000 IU/ml to prevent relapse.

Ligneous conjunctivitis is characterized by formation of wood like membrane on palpaebral conjunctiva. Immunohistochemical analysis of this membrane has shown predominance of T cells in inflammatory infiltrate.[7,14] Cyclosporine is an immunomodulator that inhibits T lymphocyte proliferation. Various case reports have suggested that cyclosporine helps in remission and prevention of relapse of inflammatory membrane.[7,11,14] We have also included cyclosporine in our treatment regimen to manage our case.

Surgical excision of membrane followed by amniotic membrane graft has also been reported as an effective treatment option. Amniotic membrane graft promotes epithelization and reduces inflammation and prevents fibrosis.[15] Hence its use may help in prevention of recurrence. Watts et al. have reported successful outcome with amniotic membrane graft along with medical management including FFP and heparin.[6] However, in our case, we excised the membrane only once at our setting for histopathological analysis and then treated medically without any surgical intervention.

The major drawback in managing our case is that we could not assess serum plasminogen level due to lack of facility. However, considering the fact of plasminogen deficiency in the pathogenesis of ligneous conjunctivitis, we initiated the treatment with fresh frozen plasma that yielded positive outcome. Though ligneous conjunctivitis is a rare condition, it can easily be diagnosed through its clinical presentation and sight threatening complications can be prevented through timely and proper management protocol.

4. Source of Funding

None.

5. Conflict of Interest

None.

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