Case report

Pseudomembrane to nodule-gamut of ligneous conjunctivitis: A case report

Pavithra Prabhakar[1], Varun Kumar Singh[2] Namrata Rao[1], Sudha Girish Menon[2]

[1]Melaka Manipal Medical College, [2]Kasturba Medical College, Manipal, Manipal Academy of Higher Education, Madhav Nagar, Manipal, Udupi, 576104, Karnataka, India

(Received: February 2022 Revised: June 2023 Accepted: August 2023)

Corresponding author: **Varun Kumar Singh.** Email: varun.singh@manipal.edu

ABSTRACT

Ligneous conjunctivitis is a type of chronic conjunctivitis characterized by the presence of pseudomembranes on the conjunctival surface. It can present as a part of a multisystem disease with involvement of other mucous membranes. A 1-year-old girl presented with features of conjunctivitis. Local examination revealed thick exudates in left tarsal conjunctiva, which was diagnosed as an ulcer with pseudomembrane and sterile culture. The patient presented with a nodular mass in the same site after a year, which showed a pseudomembrane along with sub-epithelial hyaline lobular deposits negative for Congo red, concurring a diagnosis of ligneous conjunctivitis. The patient was prescribed topical lubricating agents and regular follow up. Ligneous conjunctivitis is attributed to a deficiency of plasminogen, which plays a vital part in wound healing. The present case highlights the progression of pseudomembrane into a ligneous lesion in a child who was resisting medication and follows up.

Keywords: Conjunctiva; pseudomembrane; plasminogen deficiency; Congo red; conjunctivitis.

INTRODUCTION

Ligneous conjunctivitis is a chronic disorder characterized by recurrent episodes of conjunctivitis with the formation of fibrin rich pseudomembrane on the tarsal conjunctiva. It also affects other mucous membranes presenting as a multisystem disease. Inherited in an autosomal recessive inheritance pattern, it shows mutation in the plasminogen gene associated with type I plasminogen, which leads to impaired wound healing (1,2).

Case presentation

A 1-year-old female child presented to the outpatient department of ophthalmology with complaints of redness, excessive watering, discharge, and bleeding from the left eye for two months. It was not associated with fever, cough, cold, rashes or any other systemic complaints. There was no history of asthma or any other allergic illness. The patient was on antibiotic eye drops for the last one month before coming to the tertiary center. Topical antibiotics were discontinued in suspicion of toxic conjunctivitis and the patient was advised to install lubricating eye drops every three hours. The patient was called for a review in the next week, and it was noted that the caregivers were not compliant with the prescribed drug regimen. On local examination, there was congestion and edema of the left eyelid, congestion of the superior fornix and thick whitish adherent secretions were seen on eversion of the left upper eyelid. The cornea was clear. The pupil was normal in size and reacting to light. The lens was clear, and the fundus examination was normal. Her systemic examination was normal. The secretions were debrided and sent for histopathological and microbiological examinations. Histopathology showed an ulcerated conjunctival tissue with surface fibrinous pseudomembrane (Fig. 1).

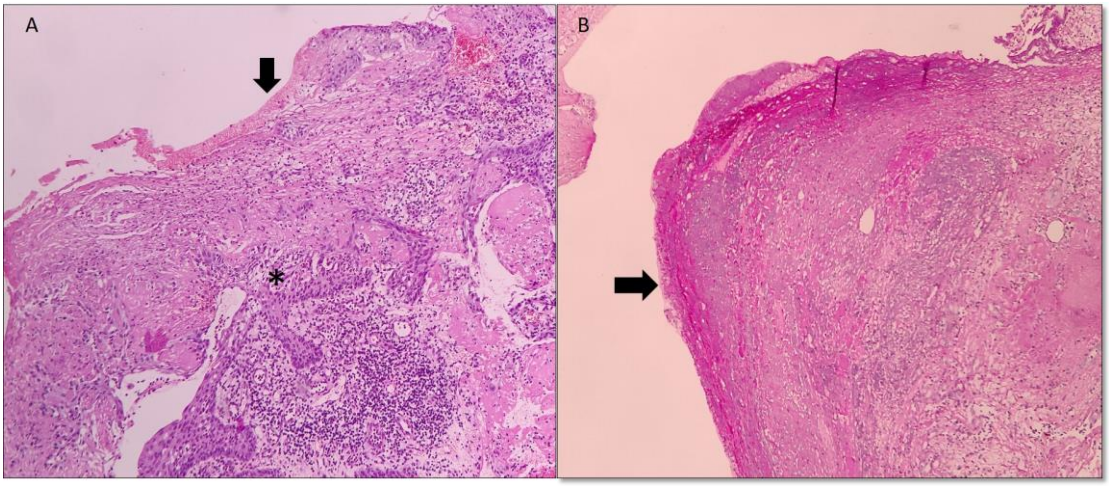

Fig. 1: A, Ulcerated conjunctival mucosal lining (*) with surface pseudomembrane (arrow) (H&E,100x). B, The pseudomembrane is composed of fibrin scaffold and neutrophils (arrow). No fungal elements/viral inclusions/ bacterial colonies were seen (PAS, 100x).

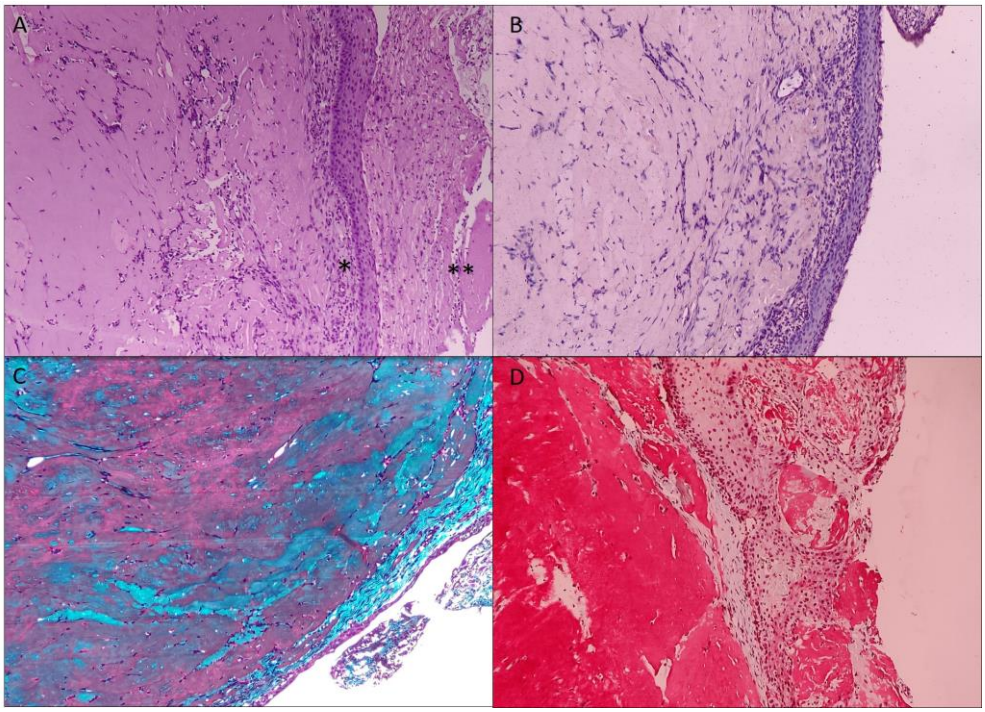

Fig. 2: A, Biopsy from nodule with atrophic conjunctival lining (*), surface pseudomembrane (**) and sub-epithelial hyaline deposits (H&E, 100x). The deposits were negative for congo red (B), and predominantly composed of fibrin highlighted by Masson's trichrome (C) and Martius scarlet blue (D).

Microbiological investigations yielded a sterile culture. The patient was advised to lubricate eye drops four times a day and a review after 15 days; however, the patient was lost to follow up. The patient came back a year later with similar complaints and examination revealed a thick white nodular mass measuring 3 x 2 cm on the left tarsal conjunctiva raising a clinical suspicion of pseudomembranous conjunctivitis/ granulomatous lesion. Histopathology showed an atrophied conjunctival mucosal lining with surface fibrinous pseudomembrane. The sub epithelium showed pink amorphous eosinophilic material resembling amyloid but was negative for Congo red. This eosinophilic material was confirmed to be a hyaline mass of fibrin on staining with special stains like Masson's trichrome and Martius Scarlet blue (Fig. 2). Keeping the clinical and pathological features a diagnosis of ligneous conjunctivitis was made. The patient was discharged with lubricating eye drops and advised for regular follow up.

DISCUSSION

Ligneous conjunctivitis is considered one of the most common manifestations of a multisystemic disorder called hypoplasminogenemia, characterized by reduced to absent levels of plasminogen antigen and its activity. Inherited in an autosomal recessive condition, it can manifest in both homozygous as well as compound heterozygous state. It can affect other mucous membranes like gingiva, respiratory tract, middle ear, female genital tract and kidney (1,3). An association with juvenile colloid milia and Dandy walker syndrome have been reported in literature (4,5).

Plasminogen deficiency is associated with impaired wound healing. The conjunctiva is frequently exposed to irritants like dust, local infection, and minor trauma especially in young children. Following this, there is fibrin exudate which initiates the wound healing cascade by acting as a scaffold for ingrowth of granulation tissue and fibroblasts followed by healing. These fibrin clots are lysed by plasmin, which is an activated form of plasminogen. In its absence, there is excess accumulation of fibrin leading to the formation of fibrin rich pseudomembrane on the conjunctiva (1,3,6). These ligneous lesions often contain foreign bodies like hair shafts, cotton fibers and silica. Surgical removal of pseudomembranes, pterygium, strabismus surgeries and treatment with tranexamic acid, an anti-fibrinolytic agent has also been documented to be initiators of ligneous conjunctivitis (1,2).

The clinical differentials of ligneous conjunctivitis include other causes of pseudomembranous conjunctivitis like viral, bacterial, toxic and allergic conjunctivitis (1,3). It should also be differentiated from amyloidosis as histopathological examination shows subepithelial deposition of amorphous pink eosinophilic material resembling amyloid; however, it does not stain the characteristic salmon color with Congo red nor shows birefringence on polarizing microscopy as in this case (2,4,5). Several management strategies have been tried to treat ligneous conjunctivitis; surgical debridement is the most common modality. Surgical manipulation is however a known risk factor for development of pseudomembrane, thus is seldom the lone treatment modality. By far the optimal management of this

condition involves a topical application of a fibrinolytic agent followed by surgical removal of the membrane and prolonged usage of topical heparin, which reduces the risk of local recurrence (1). Other agents like topical corticosteroids, cyclosporine have been tried with limited success and recurrences (1,7,8). Other potential treatment options being evaluated include topical heterologous serum and intravenous lys-plasminogen especially in cases of a multisystem disease have shown promising results in long term disease control (1,8,9).

CONCLUSION

Ligneous conjunctivitis is a rare form of chronic conjunctivitis, which can present in any age group and may be the first presentation of a multisystem disease. Attributed to the deficiency in plasminogen, it is inherited in an autosomal recessive pattern. The present-day strategy of treatment includes topical fibrinolytic agent, followed by debridement and long-term topical heparin helps in local control of the disease.

CONFLICT OF INTEREST

The authors declared they do not have anything to disclose regarding conflicts of interest with respect to this manuscript.

REFERENCES

1. Schuster, V., Seregard, S. Ligneous conjunctivitis. Surv Ophthalmol. 2003;48(4):369-388.
2. Iqbal, B., Gahlot, A., Arun, S., Magdum, R., Mushtaq, I. Ligneous conjunctivitis: A rare case report. Trop J Med Res. 2015;18(2):119.
3. Raimann, R., Moya, R., Anguita, R., Kobus, R., Pérez, M., Gonzalez, P. Clinical, histopathological, and genetic aspects in one case of ligneous conjunctivitis. Ophthalmic Genet. 2018;39(5):642-644.
4. Bhave, S.S., Joshi, S.S. Ligneous conjunctivitis in a patient of juvenile colloid milia: A rare association. Indian J Ophthalmol. 2015; 63(4):350-352.
5. Srirampur, A., Ramappa, M., Chaurasia, S., Vemuganti, G. Ligneous conjunctivitis in a Dandy– Walker syndrome: A rare case report. Indian J Ophthalmol. 2019;67:143-145.
6. Schuster, V., Hugle, B., Tefs, K. Plasminogen deficiency. J Thromb Thrombolysis. 2007;5:2315-2322.
7. Ang, M.J., Papageorgiou, K.I., Chang, S.H., Kohn, J., Chokron Garneau, H., Goldberg, R.A. Topical plasminogen as adjunctive treatment in recurrent ligneous conjunctivitis. Ophthal Plast Reconstr Surg. 2017;33(2):e37-e39.
8. Pergantou, H., Likaki, D., Fotopoulou, M., Katsarou, O., Xafaki, P., Platokouki, H. Management of ligneous conjunctivitis in a child with plasminogen deficiency. Eur J Pediatr. 2011;170(10):1333-1336.
9. Martins, T.G., Martins, T.G., Martins, D.G. K.M. Treatment of ligneous conjunctivitis with heterologous serum. Einstein (São Paulo). 2019;17(3):eRC4714.