Ligneous conjunctivitis in a patient of juvenile colloid milia: A rare association

Shubhangi Sudhir Bhave, Sanyogita Swapnil Joshi[1]

We present to you, case of a 10-year-old female with h/o redness, watering since 8 months. Her vision was 20/30 in right eye and 20/70 in left eye. Conjunctiva had plenty of purulent discharge and palpebral conjunctiva was studded with membranous lesions. She was found to have multiple hyperpigmented papulopustular lesions over face, palms and legs. She was started with topical moxifloxacin and lubricating drops. Patient was followed-up after 15 days. At that time her conjunctiva had formation of a woody pseudomembrane. Excision of the lesions and skin biopsy was done and sent for hislopathological examination. Findings of histopathological examination were suggestive of ligneous conjunctivitis and juvenile colloid milia. We have started this patient with long-term cyclosporine drops and tear supplements. In next visit, the membrane was resolved. Hence, we continued with the same treatment, but again the woody membrane recurred.

Key words: Juvenile colloid milia, ligneous conjunctivitis, pseudomembranous conjunctivitis

Ligneous conjunctivitis is a rare form of chronic, recurrent conjunctivitis characterized by fibrinous pseudomembranes on the palpebral conjunctivae. Prevalence in the general population estimated between 0.13% and 0.42% in healthy subjects with heterozygous hypoplasminogenemia. It may be associated with systemic pseudomembranous lesions of the gingiva, ears, tracheobronchial tract, female genital tract, and kidneys. It has also been linked to congenital hydrocephalus and juvenile colloid milium.

Access this article online

Quick Response Code:

Website: www.ijo.in

DOI: 10.4103/0301-4738.158091

Drishti Eye Clinic and Squint Centre, [1]Drishti Eye Clinic, Ramdaspeth, Nagpur, Maharashtra, India

Correspondence to: Dr. Shubhangi Sudhir Bhave, Drishti Eye Clinic and Squint Centre, 5th Floor, Midas Heights, Opposite Tarun Bharat, Central Bazaar Road, Ramdaspeth, Nagpur - 440 010, Maharashtra, India. E-mail: bhave.shubhangi@gmail.com

Manuscript received: 13.09.14; Revision accepted: 16.04.15

Case Report

A 10-year-old female patient resident of Madhya Pradesh presented to us with chief complaints of redness, watering, discharge, irritation in both eyes since 8 months. There was no history of fever, instillation or intake of any drugs into the eyes. There was no history of asthma or allergy.

Her vision was 20/30 in right eye and 20/70 in left eye. Her refraction could not be done as she was unable to open her eyes.

Slit lamp examination showed matting of lashes with crusted discharge. Conjunctiva had plenty of purulent discharge and palpebral conjunctiva was studded with granulomatous, firm and woody membranous lesions [Figs. 1-3]. In both eyes, there was congestion. Ocular surface appeared unhealthy showing superficial punctate keratitis, epithelial defects, and peripheral vascularisation. Anterior chamber was quiet. Pupils were normal in size and reacting to light. Fundus appeared normal in both eyes.

On general examination, she was found to have multiple hyperpigmented papulopustular lesions over face, palms and Dermatologist's opinion was taken. The findings were suggestive of juvenile colloid milia [Fig. 4]. Pediatrician's opinion was taken regarding other associations like upper respiratory tract, middle ear which were found to be normal.

She was started with topical moxifloxacin, lubricating eye drop and gel and was advised set of investigations. Investigations showed raised erythrocyte sedimentation rate (ESR) (28 mm/h). X-ray chest posterior-anterior view showed reticulonodular shadows. This aroused suspicion of miliary tuberculosis by radiologist. But as her total lymphocyte count, Mantoux test were normal and ESR was marginally raised, we excluded the possibility of tuberculosis. Patient was followed-up after 15 days. At that time her conjunctiva was studded with plenty of thick granulomatous lesions and formation of a woody pseudomembrane.

Excision of the lesions was planned. At the time of excision, membrane was easily peeled off and there were no underlying attachments. After the excision of membrane, hemostasis was achieved and supratarsal injection of triamcinolone acetate was given in both eyes. During same procedure, biopsy of skin lesions was taken and sent for hislopathological examination. Her vaginal

mucosa was examined after obtaining informed written consent from parents during the procedure and was found to be normal.

Histopathological examination of conjunctival lesion showed mucosa lined by focally atrophic squamous epithelium with submucosal eosinophils, hyaline in nodular fashion and scattered inflammatory cells. Small blood vessels were present [Fig. 5]. Histopathological findings were suggestive of ligneous conjunctivitis.

Histopathological examination of skin lesions showed dystrophic calcification in epidermal and subepidermal zone [Fig. 6]. According to pathologists and dermatologist, calcification can occur in any form of pathology after certain

Figure 1: Upper lid showing thick, woody membrane covering upper tarsus

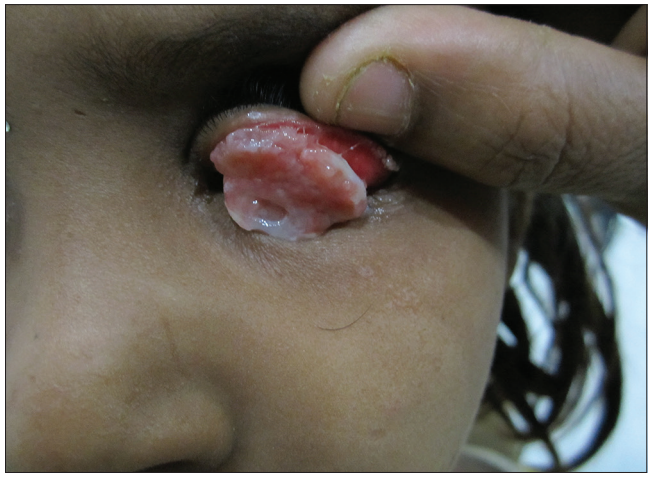

Figure 2: Woody membrane covering upper tarsus

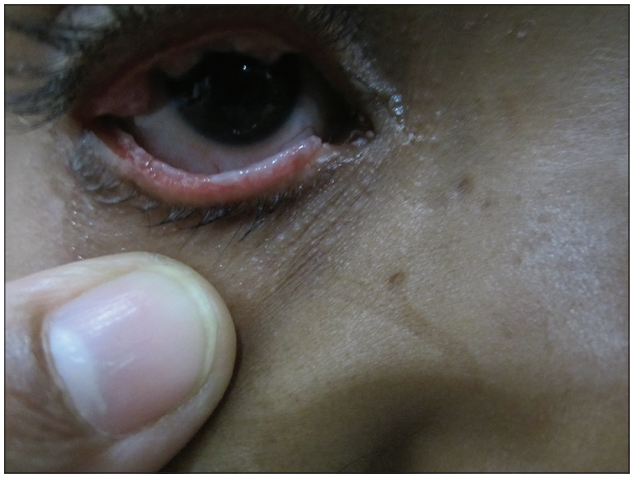

Figure 3: Lower lid showing raw areas

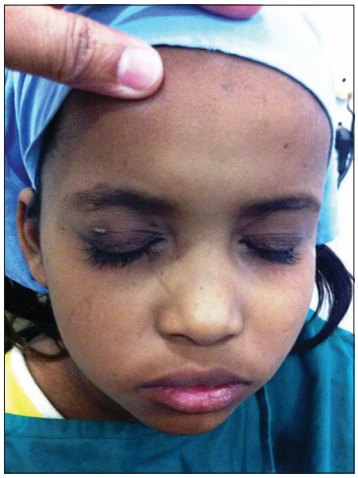

Figure 4: Skin of face showing papulopustular lesions

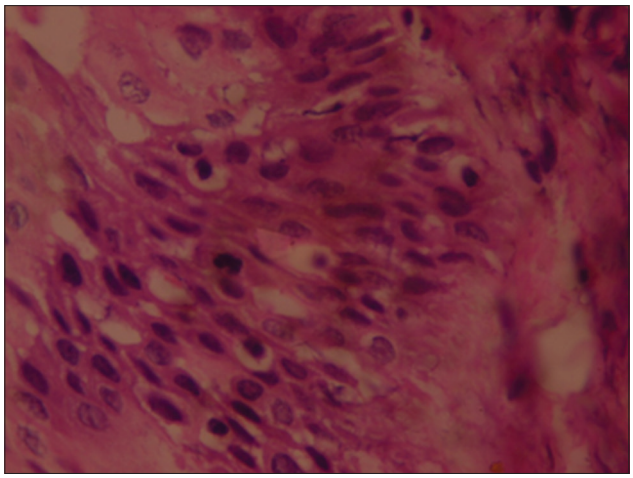

Figure 5: Focally atrophic squamous epithelium

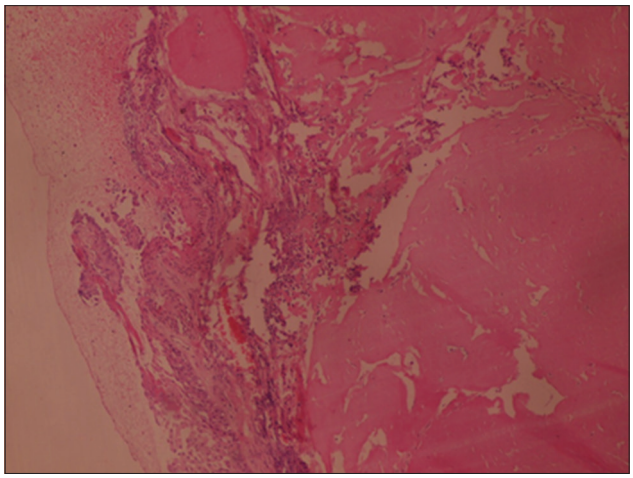

Figure 6: Submucosal eosinophlic hyaline in nodular fashion

time, though literature doesn't mention calcification in juvenile colloid milia.

Hematologist's opinion was taken for starting topical treatment with fresh frozen plasma. Patient was resident of remote village and poor. Treatment with fresh frozen plasma was not possible in this patient. So, we have started this patient with long-term cyclosporine drops and tear supplement.

After 1-month the patient was reviewed. The symptoms were relieved. On examination the thick woody membrane had resolved. She was continued with the same treatment and reviewed after 3 months. At this time, the woody membrane over conjunctiva had recurred because definitive treatment with plasminogen could not be started. This confirmed our diagnosis ligneous conjunctivitis.

Discussion

Ligneous conjunctivitis is a rare form of chronic, recurrent conjunctivitis characterized by fibrinous pseudomembranes on the palpebral conjunctivae. It may be associated with systemic pseudomembranous lesions of the gingiva, ears, tracheobronchial tract, female genital tract, and kidneys. It has also been linked to congenital hydrocephalus and juvenile colloid milium.[1,2]

Colloid milium, which is characterized by the deposition of amorphous, eosinophilic granular deposits in the superficial dermis, has subtypes including the juvenile and adult variants. The juvenile variant, develops in children before puberty and has a familial incidence. Patients present with discrete papules measuring 0.2–1.5 cm in diameter, yellow–brown in color, appear translucent and when punctured characteristically express gelatinous material. The underlying tissues often feel indurated. Juvenile colloid milium predominantly affects the face, in particular the cheeks, nose, and around the mouth. It occurs due to vascular fragility due to infiltration of the blood vessel walls by colloid material. Juvenile colloid milia may present with gingival deposits and ligneous conjunctivitis as a result of infiltration of these tissues by colloid-like material.

Ligneous conjunctivitis may be inherited in an autosomal recessive pattern due to mutations in the plasminogen gene and associated with type I plasminogen deficiency (hypoplasminogenemia), leading to impaired wound healing.[3,4]

The conjunctivae are susceptible to frequent exposure to irritants, like dust, local infection, minor injury, and trauma, especially in young children.[1] This trauma induces the wound-healing cascade, which begins with fibrin matrix deposition. The fibrin acts as scaffolding for granulation tissue and then collagenous tissue for wound remodeling and eventual healing. Plasmin acts to degrade the fibrin clots, and it is formed from cleavage of the plasminogen. Plasminogen deficiency leads to impaired healing of injured mucosa with accumulation of a serofibrinous material that forms the fibrin-rich pseudomembranes, which harden giving the characteristic "woody" appearance.

It affects infant or child and common in females, presenting with recurrent conjunctivitis symptoms including discharge and redness of the conjunctiva.

Examination shows thick, firm pseudomembranous structures on the palpebral conjunctiva. Approximately 50% of cases are bilateral, and affected individuals have a predilection for upper eyelid involvement followed by lower eyelid and bulbar involvement.[1]

The initial signs of ligneous conjunctivitis are mucoid discharge and redness of the conjunctivae. This is followed by palpebral conjunctival pseudomembrane formation, and it progresses to the mucosal thickening with a wood-like consistency, which replaces the normal eyelid mucosa. Multiple recurrences are common and coincide with as fever, upper respiratory tract infection, ear infections, or in females, urogenital tract infection.[1] Involvement of the airway can be life-threatening.

The diagnosis of ligneous conjunctivitis can be challenging. It should be differentiated from following causes of pseudomembranous conjunctivitis:

- Viral conjunctivitis
- Bacterial conjunctivitis
- Toxic conjunctivitis, e.g., secondary to medication use
- Allergic or vernal conjunctivitis any inflammatory condition would respond to triamcinolone and cyclosporine. But in our case thick woody membrane and histopathological changes confirmed the diagnosis
- Amyloidosis.

Management includes topical plasminogen concentrate,[3] topical fresh frozen plasma,[5] with systemic fresh frozen plasma,[6,7] topical heparin,[5,8-10] with topical corticosteroids or alpha chymotrypsin,[1] topical cyclosporine A with amniotic membrane,[10] systemic low-dose lys-plasminogen,[1,3] surgical excision of pseudomembrane, which has high recurrence rates.

Complications are recurrence after surgical excision, corneal scarring, vascularization and perforation and steroid induced glaucoma.

References

1. Schuster V, Seregard S. Ligneous conjunctivitis. Surv Ophthalmol 2003;48:369-88.
2. Chowdhury MM, Blackmore S, Williams S. Juvenile colloid milium associated with ligneous conjunctivitis: Report of a case and review of the literature. Clin Exp Dermatol 2000;25:138-40.
3. Mehta R, Shapiro AD. Plasminogen deficiency. Haemophilia 2008;14:1261-8.
4. Schuster V, Mingers AM, Seidenspinner S, Nüssgens Z, Pukrop T, Kreth HW. Homozygous mutations in the plasminogen gene of two unrelated girls with ligneous conjunctivitis. Blood 1997;90:958-66.
5. Ku JY, Lichtinger A, Yeung SN, Kim P, Cserti-Gazdewich C, Slomovic AR. Topical fresh frozen plasma and heparin treatment of ligneous conjunctivitis in a Canadian hospital setting. Can J Ophthalmol 2012;47:e27-8.
6. 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;170:1333-6.
7. Gürlü VP, Demir M, Alimgil ML, Erda S. Systemic and topical fresh-frozen plasma treatment in a newborn with ligneous conjunctivitis. Cornea 2008;27:501-3.
8. Hiremath M, Elder J, Newall F, Mitchell S, Dyas R, Monagle P. Heparin in the long-term management of ligneous conjunctivitis: A case report and review of literature. Blood Coagul Fibrinolysis 2011;22:606-9.
9. De Cock R, Ficker LA, Dart JG, Garner A, Wright P. Topical heparin in the treatment of ligneous conjunctivitis. Ophthalmology 1995;102:1654-9.
10. Tok OY, Kocaoglu FA, Tok L, Burcu A, Ornek F. Treatment of ligneous conjunctivitis with amniotic membrane transplantation and topical cyclosporine. Indian J Ophthalmol 2012;60:563-6.

Cite this article as: Bhave SS, Joshi SS. Ligneous conjunctivitis in a patient of juvenile colloid milia: A rare association. Indian J Ophthalmol 2015;63:350-2.

Source of Support: Nil. **Conflict of Interest:** None declared.