in either eye, and the temporal exudative detachment had resolved (Fig. 1B). The patient was subsequently placed on adalimumab and monitored for more than 1 year without recurrence.

This patient most likely had migratory orbital inflammation that occurred when she was off of immunosuppressive therapy for RA. Its association with the patient’s RA is highly implied given the time of presentation and otherwise negative work-up. Clinical decline did not reverse until she was started on high-dose intravenous steroids. Previous reports of orbital myositis have been linked to psoriatic arthropathy and lupus, and management with oral prednisolone has been shown to be beneficial. This suggests that systemic autoimmune conditions can have ocular manifestations and that treatment of the systemic inflammation also can alleviate ocular symptoms.

Previous case reports support the role of an inflammatory process in migratory inflammation. In one study, patients with orbital myositis of an isolated extraocular muscle and recurrent myositis of the contralateral orbit or eyelid, biopsy revealed perivascular lymphocytic aggregates and histiocytes. Treatment with oral steroids helped alleviate symptoms.

In cases of suspected orbital inflammation, prophylactic antibiotic treatment should be initiated, and infectious etiologies should be ruled out prior to starting steroids because infectious and noninfectious orbital cellulitis can present initially in the same manner.

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References


Footnotes and Disclosure

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Use of topical allogenic fresh-frozen plasma drops in the treatment of ligneous conjunctivitis

Ligneous conjunctivitis is a rare inflammatory condition characterized by chronic and recurrent deposition of fibrin-rich pseudomembranes on the palpebral conjunctiva. Firm membranes with a characteristic ‘woody’ induration usually occur bilaterally presenting since childhood. The formation of fibrinous pseudomembranes is caused by a mutation in the PLG gene, leading to deficient levels of type 1 plasminogen. Plasminogen is a precursor of plasin, a molecule involved in the degradation of fibrin. Pseudomembrane formation has been reported after trauma, surgery, contact with ocular foreign bodies, and infection. Lesions may also occur in other mucous membranes.

There is no standardized treatment for ligneous conjunctivitis. Several topical and systemic therapeutic options have been used to prevent pseudomembrane recurrence post-operatively. These include antibiotics, corticosteroids, immunosuppressants, heparin, plasminogen and fresh frozen plasma as well as amniotic membrane transplantation, with varied outcomes. They characteristically tend to recur shortly after surgical excision. Pseudomembranes may be debilitating and may result in vision-threatening complications.

A two-year-old female patient presented to our Pediatric Ophthalmology clinic with bilateral pseudomembranes and clear, watery discharge. The patient was otherwise healthy. She was born term and had no complications during pregnancy or delivery. Immunizations were up-to-date. There was no previous ocular history, and family history was not contributory. No history of consanguinity was reported.

On examination, visual acuity was 6/6 bilaterally using Cardiff cards. There was mild left upper lid swelling. Thick pseudomembranes were present along the palpebral conjunctival surfaces bilaterally. The rest of the anterior and posterior exam was unremarkable bilaterally.

Cultures of the palpebral conjunctiva were positive for *Hae-mophilus influenza*. The patient was treated with topical tobramycin, fluorometholone (FML), and tobramycin/dexamethasone.

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After two weeks, there was complete resolution of the pseudomembranes on the right palpebral conjunctival surfaces. In the left eye, a thick, pale yellow, firmly adherent pseudomembrane persisted over the entire upper and three quarters of the lower palpebral conjunctival surface. The lesions on the left palpebral conjunctiva were surgically excised in the operating room with minimal bleeding. The patient was given topical tobramycin/dexamethasone. Unfortunately, pseudomembranes recurred two weeks post-operatively on both left palpebral surfaces (Figure 1).

Figure 1—Colour photograph of the left eye of a two-year-old female with ligneous conjunctivitis showing: A) worm’s-eye view of a thick pseudomembrane covering most of the (everted) upper palpebral conjunctival surface; and B) en-face view of the same large pseudomembrane over the upper palpebral conjunctiva and a smaller pseudomembrane over the lower palpebral conjunctiva post-resection on a regimen of topical tobramycin/dexamethasone. Credit: Dr. Dorothy Bautista, 2016.
Histopathological examination demonstrated acellular fibrinous material with chronic inflammation, in keeping with ligneous conjunctivitis. Plasminogen level was 0.2U/mL (normal range 0.8-1.2U/mL). The diagnosis of Type 1 Plasminogen deficiency was confirmed by genetic testing, which demonstrated a homozygous pathogenic mutation in the PLG gene, c.112A>G (previously denoted as p. Lysine19-Glutamic Acid or p. Lys19Glu), where a negatively charged amino acid is switched for a positively charged one.

The pseudomembranes remained stable on a topical regimen of FML and cyclosporine. A trial discontinuation of steroid drops resulted in a prolonged episode of keratitis, which eventually resolved with increased lubrication and resumption of the steroid drop regimen.

At age five, the patient began to develop mild ptosis of the left upper eyelid and mild astigmatism secondary to pseudomembrane size (Figure 2A). The patient also presented increased intraocular pressure and early lens opacity in the left eye secondary to prolonged steroid use. Corrective glasses were prescribed, and the patient was placed on timolol.

A second surgical excision of the left upper palpebral conjunctival pseudomembrane was performed. The pseudomembrane on the left lower palpebral conjunctiva was too small to warrant excision. In post-operative week one, there was a small recurrence of the pseudomembrane on the left upper eyelid; however, the pseudomembrane completely regressed by post-operative week two, leaving a small linear scar.

The patient was placed on a post-operative regimen of topical allogenic fresh frozen plasma (FFP) drops hourly, cyclosporine drops four times a day, and tobramycin/dexamethasone drops four times a day with a prolonged taper over five months.

FFP was prepared from pooled plasma by the Canadian Blood Services. FFP was stored frozen in vials, which contained approximately 3-4 doses each. The vials were stored in a refrigerator once opened.

The taper involved continuing FFP drops hourly only while awake and decreasing to every 3 hours while asleep on post-operative day five. The frequency of FFP was slowly decreased over five months. Tobramycin/dexamethasone and cyclosporine drops were discontinued after three weeks and four months, respectively.

Ten months post-operatively (i.e. five months after discontinuation of topical drop regimen), there was scarring of the left upper palpebral conjunctiva but there was no sign of pseudomembrane recurrence. The pseudomembrane on the left inferior palpebral conjunctiva decreased in size and remained stable (Figure 2B). The patient had corrected distance visual acuity of 6/6 bilaterally, with normal ocular alignment, normal motility, and excellent stereopsis. Corneas were clear bilaterally, and the early lens opacity noted in the left eye was unchanged.

Many aspects of our patient’s presentation followed a course characteristic of ligneous conjunctivitis. The patient presented with a first episode of ligneous conjunctivitis in childhood, carried a homozygous mutation in the PLG gene, possessed a low level of plasminogen, and had recurrence of pseudomembranes after surgical resection. In this patient, infection was identified as a precipitating factor in the development of pseudomembranes, which has been reported in the literature.1,2

Homozygous mutations in p. Lys19Glu have been associated with a higher residual PLG activity; and therefore, a milder clinical course of Type 1 Plasminogen deficiency. This may explain the complete absence of systemic disease, characterized by pseudomembrane formation in other organ systems, in our patient. The patient continues to have annual evaluations to monitor for the formation of pseudomembranes in the oral cavity, nasopharynx, respiratory tract, gastrointestinal tract, genitourinary tract, and the cerebral ventricular system.

The size of pseudomembranes may predict response to treatment. During our patient’s original presentation, the smaller pseudomembranes on the right palpebral conjunctiva completely resolved with a course of topical antibiotics and steroids, whereas the larger one on the left palpebral conjunctiva persisted. This is an interesting outcome as ligneous conjunctivitis-associated pseudomembranes have been extensively reported to be non-responsive to antibiotic and steroid therapy.2,5 In addition, the pseudomembrane present on the left lower palpebral conjunctiva, which was not resected a second time due to its small size, decreased in size over five months on a course of topical allogenic fresh frozen plasma, cyclosporine, and tobramycin-dexamethasone with taper. This is also an interesting outcome as fresh frozen plasma has only been reported to prevent recurrence of pseudomembrane formation after surgical excision, but the ability to decrease the size of existing pseudomembranes has not been reported.2,5

The use of topical fresh frozen plasma drops may be a suitable option for the treatment of ligneous conjunctivitis flare-ups. Our case suggests that topical fresh frozen plasma is successful in preventing the recurrence of pseudomembranes3,5 without the need of its subconjunctival or systemic administration.2,4 The results of our case also suggest that topical fresh frozen plasma may be useful in reducing the size of existing pseudomembranes.

Topical administration of fresh frozen plasma decreases the likelihood of adverse outcomes including anaphylaxis, transfusion-related acute lung injury, and haemolysis related to its systemic administration.4 The use of topical medication also eliminates the need for hospitalization, intravenous access, and the short half-life of intravenous fresh frozen plasma.

Our case adds to the growing body of evidence suggesting that the use of frozen plasma, in combination with other topical medications, represents an acceptable treatment for the management of ligneous conjunctivitis. Fresh frozen plasma has shown to be a more accessible and cost-efficient treatment compared to topical plasminogen.2,5
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Figure 2—Colour photograph of the everted left eye of a five-year-old female with ligneous conjunctivitis showing: A) a chronic thick  
pseudomembrane covering most of the upper palpebral conjunctiva on a regimen of topical FML and cyclosporine post-recurrence  
three years prior; and B) scarring of left upper palpebral conjunctiva but no sign of pseudomembrane recurrence ten months post-  
resection after a five-month regimen of slowly tapered topical allogenic fresh frozen plasma drops, cyclosporine and tobramycin/  
Unilateral vision loss in a patient with essential thrombocythemia

Essential thrombocythemia is a chronic myeloproliferative neoplasm that develops when megakaryocytes in the bone marrow produce an excess number of platelets. This neoplasm that develops when megakaryocytes in the bone marrow produce an excess number of platelets. It is a chronic myeloproliferative disorder associated with a number of thrombotic and embolic complications, and prior cases of vision loss have been documented. We describe a rather unique case of essential thrombocythemia presenting with unilateral vision loss in a patient.

A 73-year-old male was referred to the ophthalmology clinic for a 1-week history of decreased vision in the left eye. The vision loss was described as a “green” film that obstructed his entire vision, with sparing of his superonasal visual field. Ocular history was remarkable for bilateral cataract surgery performed 30 years prior. His medical history included hypertension, dyslipidemia, coronary artery disease treated with a coronary artery bypass graft, right carotid endarterectomy, and a left parietal stroke 3 years earlier. His medications included atenolol, ramipril, alendronate, ranitidine, rosuvastatin, folic acid, and Aspirin. The patient denied any headaches, scalp tenderness, jaw claudication, eye pain, or weight loss.

Examination revealed a visual acuity of 20/60 OD improving to 20/40 with pinhole correction and counting fingers OS. There was a left afferent papillary defect (relative afferent papillary defect). Colour testing with Ishihara plates revealed 11/15 in the right eye and 0/15 in the left eye. Extraocular movements were full bilaterally. Slit-lamp examination demonstrated well-centred posterior chamber intraocular lenses with no evidence of posterior capsule opacification. Intraocular pressure was 24 and 25 mm Hg in the right and left eyes, respectively. Dilated fundus examination showed pale optic nerve swelling with associated rare disc hemorrhage. Compared with the right eye, there was also arteriovenous engorgement (Fig. 1). Optical coherence tomography also revealed disruption of the inner retina and outer nuclear layers, loss of foveal contour suggesting mild retinal edema, and left optic nerve head edema. Arteriovenous phase fluorescein angiography revealed decreased mid-peripheral choroidal perfusion and mild disc leakage in the left eye (Fig. 1).

Goldmann visual field demonstrated a dense left central scotoma.

Laboratory testing at time of presentation demonstrated a normal C-reactive protein level, erythrocyte sedimentation rate, and B12 level. However, a complete blood count revealed an elevated white blood cell count (20.6 × 10^9/L) and significantly elevated platelets (102 × 10^10/L). Previous bloodwork demonstrated that the patient’s platelets had been elevated for more than 1 year (63.9 × 10^10/L) prior to his presentation to the ophthalmology department. Subsequent computed tomography angiography revealed a chronic left parietal infarct in keeping with his previous cerebrovascular accident and severe (>95%) stenosis of the left common carotid artery bifurcation. A bone marrow biopsy was performed to rule out thrombocytemia but was inconclusive. However, subsequent genetic testing for a JAK-2 mutation was positive, thereby confirming a diagnosis of essential thrombocytemia.

Essential thrombocytemia is a chronic myeloproliferative neoplasm characterized by excess platelet production by megakaryocytes in the bone marrow and can result in thrombohemorrhagic complications and possible progression to myelofibrosis or acute leukemia. In this rare hematologic disorder, spontaneous thrombotic events are common, and systemic consequences include cerebrovascular accident, transient ischemic attack, deep vein thrombosis, pulmonary embolism, coronary artery ischemia, and hepatic, portal, splenic, and mesenteric vein thrombosis. The World Health Organization criteria for essential thrombocytemia includes a platelet count of ≥450 × 10^9/L and a JAK-2 mutation. JAK-2 is a nonreceptor tyrosine kinase that serves a vital role in transducing signals from various class I cytokine receptors; mutations in this gene disrupt normal functions that are for myelopoiesis. Notably, the JAK-2 mutation can be identified in more than 50% of patients who present with essential thrombocytemia.

Previous reports in the literature have discussed the presentation of essential thrombocytemia in association with ocular manifestations, most commonly presenting with central retinal vein occlusion, branch retinal vein occlusion, or...