Table 1. Infants with osteopetrosis who underwent optic canal decompression reported in literature

Case	Age, months	Sex	Indication for surgery	Visual acuity		VEP		Follow-up	Ref. No
				Pre-op	Post-op	Pre-op	Post-op		
1	6	F	OA + OC narrowing	NA	NA (stable)	NA	NA	Died at 15 mos	1
2	4	M	OA + OC narrowing	NA	NA	Abnormal	Improved	Died	4
3	7	F	OA + OC narrowing	NA	NA	Abnormal	NI	Died at 2 mos	4
4	8	M	OA + OC narrowing	NA	Normal	Abnormal	Improved	4 months	4
5	1	M	OA + OC narrowing after 2 BMTs	NA	NA	Abnormal	NI	Died at 6 mos	5

BMT, bone marrow transplantation; *NA*, not available; *OA*, optic atrophy; *OC*, optic canal; *VA*, visual acuity; *VEP*, visual-evoked potential.

patients, 4 showed improvement in VEP; however, there was no documentation of quantitative assessment of visual acuity (Table 1). To our knowledge, ours is the first case of OCD via endoscopic approach in an infant with osteopetrosis.

Prophylactic deroofing of the optic canal has been reported in other conditions associated with excess bony growth of the optic canal; however, the procedure carries a risk of vision loss.[9] The general consensus is to intervene surgically only in patients with sudden or progressive deterioration of vision.[10] A recent metanalysis comparing surgery and observation in patients with fibrous dysplasia showed that expectant management resulted in better visual outcomes than OCD in asymptomatic patients with radiological evidence of optic nerve compression.[10]

Although visual impairment in our patient was multifactorial, she had postoperative improvement in behavioral indicators of visual function as witnessed by the parents and vision teachers as well as improved VEP, visual acuity, and nystagmus. In centers with an experienced team of skull base surgeons, endoscopic OCD should be considered in children with optic neuropathy secondary to osteopetrosis. Regular ophthalmology evaluation is crucial to recognize early visual dysfunction in these children. The importance of electrodiagnostic testing and neuroimaging cannot be overemphasized if the child shows any change in visual behavior.

Literature Search

PubMed was searched on November 30, 2017, for English-language results, using the following terms: *osteopetrosis*, *optic atrophy*, and *optic canal decompression*.

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Fresh frozen plasma (Octaplas) and topical heparin in the management of ligneous conjunctivitis

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Ligneous conjunctivitis is a rare form of chronic recurrent membranous conjunctivitis with reduced plasminogen activity. It is

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characterized by the formation of characteristic firm ("woody") membranes on the tarsal conjunctiva. Similar lesions may occur on other mucous membranes. When treated with local excision, ligneous conjunctivitis is invariably associated with recurrences. Various therapeutic modalities, including topical heparin, cyclosporine, fresh frozen plasma (FFP), plasminogen, and amniotic membrane transplantation have been reported to reduce postoperative recurrences. We present 2 cases of recurrent ligneous conjunctivitis in children successfully managed with surgical excision under cover of FFP transfusion, amniotic membrane grafting, and combined with concomitant postoperative administration of topical heparin, steroids.

Case 1

A 6-year-old girl of Arab descent, the product of consanguineous parents, presented to the pediatric ophthalmology unit of the University Hospital, Oman, with bilateral yellowish-white masses under both eyelids, noted since she was 2 years of age. Surgical excision of the masses was followed by recurrence. Topical steroids, antibiotics, and mast cell stabilizers had failed to reduce the size of the lesions. She was known to have congenital plasminogen deficiency. Her visual acuity was 20/20 in in each eye. The eyelids were thickened, and firm, sessile, yellowish white ("woody") lesions were observed on the tarsal conjunctiva of both eyes (Figure 1).

Plasminogen level was 0.24 IU/ml (normal range, 0.730–1.270 IU/ml). She was treated with topical prednisolone 1% every 4 hours, cyclosporine 0.05% twice daily, and olopatadine once daily for 6 months. The lesions failed to respond to treatment and further surgery was planned. Fresh frozen plasma (Octaplas; Octapharma, Vienna, Austria) 20 ml/kg was administered intravenously (IV) twice on the day before surgery. Conjunctival lesions were excised, and synthetic amniotic membrane (Ambio dry 2; IOP Ophthalmics, Costa Mesa, CA) was sutured onto the raw palpebral conjunctiva with interrupted 6-0 polyglactin 910 sutures. Fresh frozen plasma (20 ml/kg) was administered intravenously during surgery and twice daily for the first 3 days postoperatively. Following these infusions, 10 ml/kg of FFP was administered intravenously once daily for the next 3 days. Heparin eye drops (5000 IU/mL, Farmigea SpA, Pisa, Italy) were instilled every 30-60 minutes for the first 3 postoperative days followed by every 1-2 hourly for 14 days (during the day for 12 hours) and 4 times daily for 2 months. Cyclosporine 0.05% eye drops twice daily, prednisolone drops 1%, and erythromycin eye ointment were also administered postoperatively in both eyes.

Complete resolution was noted postoperatively in each eye (eFigure 1). The frequency of heparin and prednisolone eye drops was tapered very slowly over 2 months. Histopathology revealed subepithelial amorphous hyaline-like eosinophilic material with an inflammatory cellular infiltrate negative for amyloid.

Case 2

An 18-month-old white boy, the product of a nonconsanguineous relationship, was diagnosed with bilateral hemorrhagic conjunctivitis and preseptal cellulitis at the age of 3 months that improved after treatment with intravenous antibiotics and steroid-antibiotic drops. Six weeks following resolution of the conjunctivitis, his parents felt that the left eye had not completely recovered. On examination at the ophthalmology department at the University Hospital of Wales, two pedunculated, vascular masses were noted to arise from the left upper tarsal conjunctiva. This was diagnosed as a pyogenic granuloma (or lobular hemangioma) and treated with choramphenicol and dexamathasone eye drops followed by excision of the masses. Histology revealed squamous epithelium with granulation tissue. Three months after excision the granuloma recurred and was again excised. Ligneous conjunctivitis was suspected at this stage. Plasminogen levels of 0.25 IU/L (normal, 2.5–4.2 IU/L) were reported. Within 3 months the lesion had recurred despite topical steroids (Figure 2A). Repeat excision was undertaken with a regimen of FFP at 20 mg/kg administered intravenously before and immediately following surgery. A total of 9 doses were administered over 1 week. In addition, the child received two-hourly heparin eye drops during the day (5000 IU/mL), chloramphenicol and dexamethasone eye drops 4 times daily for 1 week, followed by heparin eye drops only 4 times daily for 4 weeks, tapered over 4 weeks. Histology demonstrated ulcerated epithelium with neutrophils and fibrin with eosinophilic deposits. Follow-up at 1 year has showed no recurrence (Figure 2B).

Discussion

Ligneous conjunctivitis is caused by type 1 plasminogen deficiency which may be associated with other mucosal involvement, hydrocephalus, pericardial infusion and infertility.[1-3] Ligneous conjunctivitis may be triggered by local injury or local and systemic infections.[1,4] The absence of plasmin activity results in impaired fibrinolysis and formation of fibrin-rich, cellular inflammatory infiltrative, wood-like sessile or pedunculated membranes.[5] The cornea is an extrahepatic site of plasminogen synthesis, controlling the concentration of plasminogen in tear fluid.[6]

Both cases of ligneous conjunctivitis reported here were successfully treated with surgical excision, perioperative FFP transfusions, and topical heparin. Surgical excision alone can trigger a recurrence of the lesions.[7] Postoperative control of inflammation is important for successful treatment. Several treatment strategies have been tried. Cyclosporine A decreases the severity and recurrences following surgical excision.[8] Heparin has been used to prevent recurrence of membranes.[9] Topical plasminogen and recombinant tissue plasminogen activators are known to be effective in the management of ligneous conjunctivitis but they are expensive, and are not commercially available.[10,11] Amniotic membrane facilitates epithelialization and reduces inflammation and scarring.[12] In our patients perioperative intravenous FFP transfusions improved the hypoplasminogenemia, intensive use of topical heparin, and amniotic

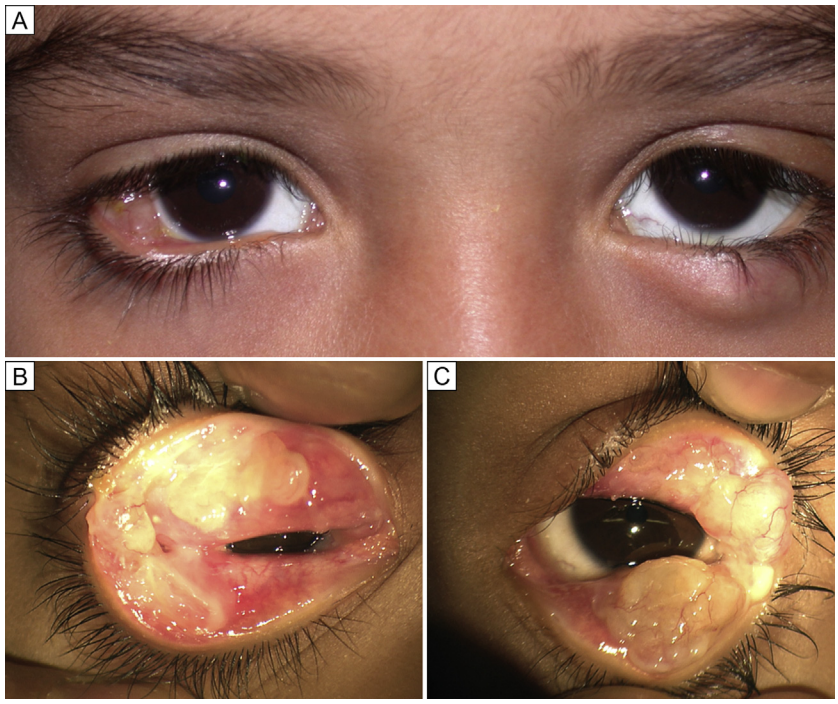

FIG 1. Case 1. Clinical photograph (A) showing bilateral lid swelling and mechanical ptosis due to tarsal conjunctival lesions. Preoperative photographs of the right eye (B) and left eye (C) showing florid sessile conjunctival lesions in the upper and lower tarsal conjunctiva.

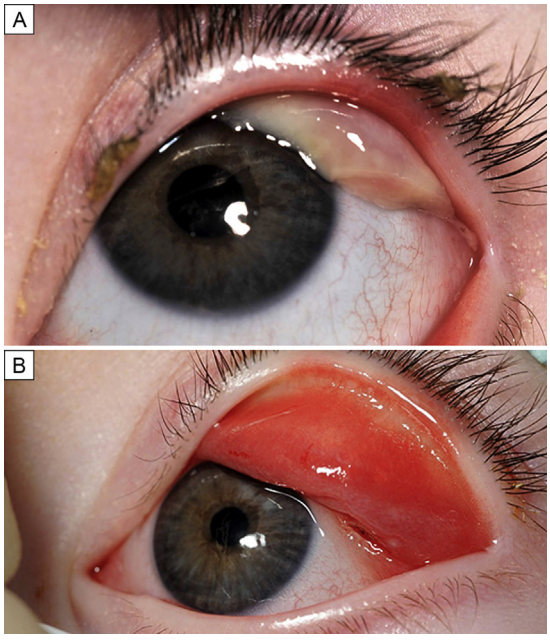

FIG 2. Case 2. Clinical photograph showing pedunculated mass in the left upper eye lid (A) and no recurrence 1 year after excision (B).

membrane transplant (case 1) successfully prevented recurrence of the conjunctival lesions. The frequency and duration of heparin and dexamethasone was based on the clinical response of re-epithelialization.[9] The successful treatment of case 2—with excision of the recurrent lesions, intravenous FFP, topical steroids, and heparin—prompted the initiation of a similar treatment for case 1, who had hitherto been treated without success using cycloporine, steroids, and olopatadine. The addition of FFP and heparin resolved the lesions. Hence, this may suggest that use of cyclosporine is not useful in the treatment of these cases. It is possible that FFP alone with meticulous surgical excision and due consideration to the type suture material used (less pro-inflammatory than polyglactin 910) would have been effective. To our knowledge, administration of FFP alone, without membrane excision, has not been reported.

Further work on the formulation of a stable sterile preparation of plasminogen eye drop holds promise for the management of this rare form of sight-threatening membranous conjunctivitis.[13] In the absence of a commercially available plasminogen preparation, and because recurrences are still possible, our success with intravenous FFP together with, topical medications, and meticulous surgical excision of the membranes offers a reasonable plan for the treatment of ligneous conjunctivitis.

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Orbital wooden foreign body manifesting as hyperdensity on computed tomography

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Detection of wooden foreign bodies within the orbit can be difficult on imaging, including computed tomography (CT). When visible, wood appears iso- or hypodense and can mimic air or fat. We report the case of a 3-year-old boy with progressive orbital cellulitis following a penetrating wood injury to the right lower lid. CT imaging revealed a linear hyperdensity contiguous with an orbital abscess. He underwent a right anterior orbitotomy with abscess drainage, during which exploration revealed a 2.3 cm retained wooden foreign body. The appearance of wood as hyperdense on imaging is unusual. In the appropriate context, high clinical suspicion for retained wood should be maintained with any irregularity on CT imaging.

Orbital wooden foreign bodies are uncommon but can be associated with high rates of infection.[1] Unfortunately, retained wood is often difficult to diagnose with imaging, because wood tends to mimic the appearance of air or fat on computed tomography (CT), and magnetic resonance imaging (MRI) does not always detect organic foreign bodies. We describe a case of poorly responsive cellulitis of the right lower lid 4 days after a penetrating wood injury with evidence of a an orbital abscess and linear hyperdensity on imaging.

Case Report

A 3-year-old boy rolled off a sled that was being pulled over a slight decline; a stick found embedded in his right lower lid was pulled out by his mother. He was immediately brought to the emergency department, where the wound was cleaned; an ophthalmology consultation found no globe rupture. The following day he started amoxicillin clavulanate by his pediatrician for a red and swollen right lower lid; when this failed to demonstrate improvement in less than 24 hours, the boy was brought to the emergency room, where 2 doses of intravenous ampicillin/sulbactam were administered. The following day, his pediatrician switched him to oral clindamycin. After continuing to show progression overnight, the patient presented to the Harkness Eye Institute for evaluation. On review of systems, there was no fever or chills, no change in appetite, no change in activity level and no pain. On ophthalmic examination, visual acuity was 20/30 in each eye without correction, there was no relative afferent pupillary defect, and extraocular motility was full in both eyes. The marginal reflex distance 1 (MRD1) was 1 mm on the right and 4 mm on the left with a marginal reflex distance 2 (MRD2) of 2.5 mm on the right and 6 mm on the left. There was significant edema and erythema of the right lower lid with a circular scab medially and slight bruising along the central tear trough. Medially the right lower lid was firm and tender without fluctuance. There was mild edema without erythema of the right upper lid. The anterior segment was unremarkable, specifically with no conjunctival injection. There was no optic nerve edema and the posterior segment was unremarkable.

Given the concern for retained wood or other foreign material in the right lower lid, the patient underwent CT of the orbits with contrast. The CT revealed a right

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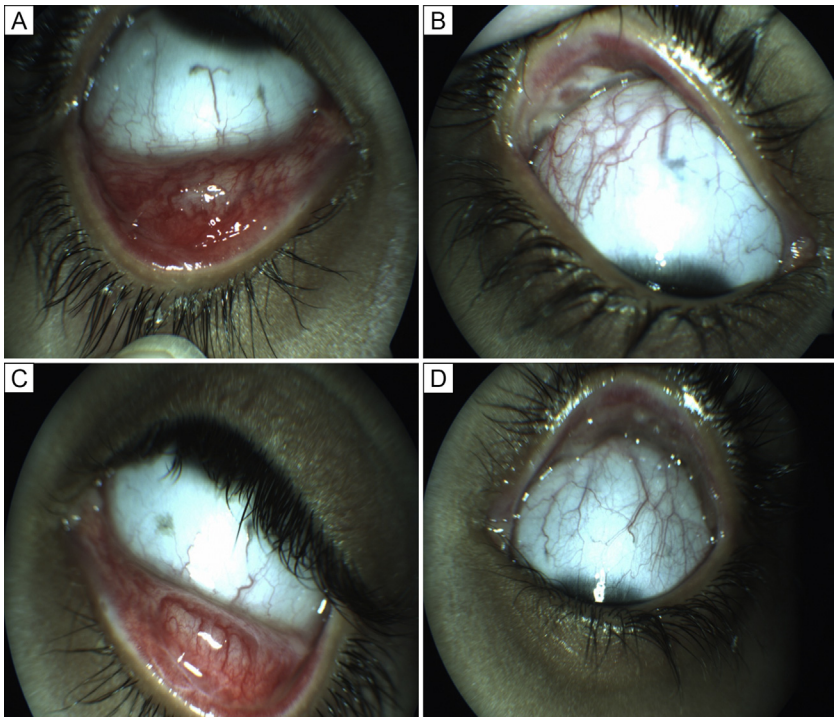

eFIG 1. Patient 1: six month postoperative photographs of the right (A,B) and left (C,D) eyes showing clear upper and lower tarsal conjunctiva in the right eye, and early reformation of soft membrane on upper tarsal conjunctiva of the left eye.