Surgical Drainage of Lymphangiectasia Haemorrhagica Conjunctivae

Jeffrey M. Goshe, MD,* Sean Platt, MD,* Gabrielle Yeaney, MD,† and Arun D. Singh, MD*

Purpose: To report a case of nonresolving lymphangiectasia haemorrhagica conjunctivae (LHC) successfully treated with surgical drainage.

Methods: Retrospective case report.

Results: A 17-year-old white girl presented with a history of a large red lesion affecting her right eye. Approximately 1 year earlier, she had noticed a small painless “red spot” affecting the inferior aspect of the conjunctiva of her right eye. Three months before presentation, she noticed a sudden increase in the size of the lesion. There was no history of trauma, and she was not a contact lens wearer. Her medical history was remarkable for a remote seizure disorder which had been stable on levetiracetam. Magnetic resonance imaging/magnetic resonance angiography of the brain and orbits was unremarkable. Anterior segment fluorescein angiography did not show any flow in the lesion, indicating a lymphatic origin of channels. Surgical drainage of the lesion was performed while sparing the affected conjunctiva resulting in an excellent cosmetic outcome. Incisional biopsy confirmed the diagnosis of LHC. The patient remained free of recurrence 6 months after the procedure.

Conclusions: Nonresolving LHC can be managed successfully with conjunctival sparing surgical drainage with an excellent cosmetic outcome.

Key Words: lymphangiectasia haemorrhagica conjunctivae, conjunctival lymphangiectasis, conjunctival hemorrhage

(L)ymphangiectasia haemorrhagica conjunctivae (LHC) was originally described by Leber in 1880. LHC is characterized by spontaneous hemorrhagic engorgement of conjunctival lymphatic channels. Most cases are localized and resolved over days to weeks. We report a unique case of severe nonresolving LHC causing cosmetic deformity successfully managed with conjunctival sparing surgical drainage.

CASE REPORT

A 17-year-old white girl presented for evaluation of a chronic “red spot” affecting her right eye. Her medical history was remarkable for a seizure disorder which had been successfully managed with oral levetiracetam for the previous 5 years. She originally noticed a small red lesion on the inferior bulbar surface of the right eye 1 year earlier, which she described as a “cut.” During this time, she was evaluated by an outside ophthalmologist who noted a small prominent conjunctival vessel inferiorly with prominent cystic changes of lymphatics circumferentially. Appearance remained stable for 9 months when, 3 months before presentation, she noticed a sudden and dramatic increase in the size of vascular channels on the ocular surface. There was no pain, associated trauma, recent viral illness, or other symptoms coincident with the change in appearance. The patient was observed for 3 months during which time the lesion remained unchanged.

The patient was referred for a second opinion at the Cole Eye Institute. Vision was 20/20 OU. Intraocular pressure measured 18 and 22 mm Hg. Pupil size and reactivity, extraocular motility, and confrontational visual fields were all within normal limits. Slit-lamp examination was remarkable only for a large circumferential network of blood-filled vessels extending from the limbus in a 4–8-mm wide zone (Figs. 1A, B). When manipulated gently with a cotton-tipped applicator, the superficial component was freely mobile, but the deeper vessels were fixed.

Based on the location and characteristics, a lymphatic origin of vessels was suspected. Although suspicion for an arteriovenous fistula was low, because of the patient’s history of a seizure disorder, magnetic resonance imaging/magnetic resonance angiography of the brain and orbits was ordered and found to be unremarkable. Anterior segment fluorescein angiography was performed to identify the presence of any communicating arterial or venous feeder vessels. Angiography revealed normal filling of surrounding conjunctival arterioles and venules, whereas the lesion did not show any hyperfluorescence, confirming no-flow sequestration of blood in lymphatic channels (Fig. 1C).

Based on chronicity and severity of the lesion, the patient and her family were interested in excision to restore the normal cosmetic appearance of her eye. Conjunctival excision of the interpalpebral portion of the lesion was planned with amniotic membrane grafting to cover conjunctival defect. If necessary, a second-staged procedure would be performed to resect the superior and inferior aspects. Surgery was performed under general anesthesia. After sterile preparation, an 8-mm conjunctival incision was created temporally. After reflecting the conjunctival flap with sharp dissection, it was...
observed that some of the lymphangiectatic channels had partially drained of blood and appeared less prominent. Rather than excising the conjunctiva, it was decided to empty the vessels of their contents by making multiple small incisions and applying pressure with surgical sponges (see Video, Supplement Digital Content 1, http://links.lww.com/ICO/A455). Focal cautery was applied to larger channels. A 2-mm strip was excised for histopathology (Fig. 2), and the conjunctival flap was reapproximated with fibrin glue. A 22-mm bandage contact lens was applied, and topical antibiotics and steroid eye drops were instilled. The contact lens was left in place for 2 weeks and then removed. The postoperative course was uneventful, and the patient was very satisfied with the cosmetic result (Fig. 3A). There were no recurrences as of 6 months after the procedure, although dilated lymphatic vessels remained visible clinically and with optical coherence tomography (Figs. 3B, C).

**DISCUSSION**

LHC is a rare but potentially overlooked condition resulting from acute or subacute hemorrhagic dilation of conjunctival lymphatics. Most cases seem to be limited to 1 quadrant, although circumferential cases have been reported. Few published studies indicate that it is typically self-limited, although some patients may suffer from multiple recurrences. The exact mechanism by which blood gains access to the lymphatic channels is not well understood. Previous authors have speculated that failure of lymphatic valves may allow retrograde flow from the associated venous vessels.

Initial management of LHC is typically conservative as appearance is often minimally disfiguring, and many lesions resolve spontaneously over days to weeks. Previous authors have described the use of Argon laser to cauterize the channels connecting blood vessels and lymphatics. Our case was remarkable both in the severity of presentation and chronicity of the lesion. Because there were no visible feeding vessels both clinically and angiographically, we believed that surgical excision would be the best option to provide optimal cosmetic results. This technique has not been described for LHC, although surgical excision of symptomatic lymphangiectasis (nonhemorrhagic) has been previously reported with good results. Unexpectedly, our initial dissection demonstrated the relative ease with which the blood could be drained from lymphatic channels, thus allowing the surrounding conjunctiva to be spared.
Judicious application of cautery may help to decrease the risk of recurrence, similar to the Argon laser technique described previously.

To date, we are not aware of any reports of LHC being managed with surgical drainage. Although it is prudent to first allow time for spontaneous clearing, we believe that the technique of multiple incisions with manual drainage can provide a simple minimally invasive method to restore normal cosmesis for patients with nonresolving LHC.

REFERENCES

FIGURE 3. A and B, Slit-lamp photographs demonstrating appearance 4 months after surgical drainage. Note the prominent conjunctival lymphatics visible inferiorly. C, Anterior segment optical coherence tomography showing persistent dilated conjunctival lymphatic channels.