Nasolacrimal duct obstruction secondary to sarcoidosis

72-year-old female with sarcoidosis, corneal perforation, and nasolacrimal duct obstruction

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October 22, 2015

INITIAL PRESENTATION

Chief Complaint: Pus, discharge, and blurry vision in the left eye.

History of Present Illness: A 72-year-old African-American female was referred by an outside ophthalmologist for a corneal perforation in the left eye. She had been followed for several months due to a purulent conjunctivitis of the left eye. She reported blurry vision in the left eye for one month. She denied ever having any pain. Three days prior to the referral, corneal thinning was noted and she was started on gatifloxacin 6 times daily. She perforated despite the antibiotics and was referred for evaluation and treatment.

Past Ocular History: No light perception (NLP) vision in the right eye (OD) secondary to sarcoidosis involving the optic nerve status post biopsy of the right optic nerve 28 years prior

Past Medical History:

- Sinonasal sarcoidosis
- Dementia
- Steroid-induced severe psychosis
- Gout
- Anemia

Medications:

- Multivitamin
- Iron supplements

Allergies: No known drug allergies

Family History: Non-contributory

Social History: Non-contributory

Review of Systems: Negative except for what is detailed in the history of present illness

OCULAR EXAMINATION

Visual Acuity with correction (Snellen – Linear)

- OD: NLP
- Left eye (OS): 20/800

Ocular Motility: Full both eyes (OU)

Intraocular Pressure (IOP)**:** Deferred due to the corneal perforation OS

Pupils:

- OD: Large relative afferent pupillary defect (RAPD)
- OS: 4 mm in dark, 3 mm in light, minimal reaction, no RAPD

External: Normal

Slit lamp exam:

- Lids/lashes:
  - OD: Normal
  - OS: Copious purulent discharge on the eyelids; large purulent efflux with pressure over the nasolacrimal sac but not canaliculus [Figure 1]
- Conjunctiva/sclera:
  - OD: Clear and quiet.
  - OS: +1-2 Diffuse injection with copious purulent discharge and white blood cells (WBCs) in the tear film
- Cornea:
  - OD: Clear
  - OS: 4.0 mm x 2.3 mm area of corneal ulceration and thinning near the limbus from 9:30 to 11:30 with a 3.2 mm x 1.0 mm corneal perforation with iris prolapse
- Anterior chamber:
  - OD: Deep and quiet
  - OS: Very shallow
- Iris:
  - OD: Normal architecture
  - OS: Iris prolapse through the corneal perforation
- Lens:
  - OD: 2+ nuclear sclerosis
  - OS: 2+ nuclear sclerosis

Dilated fundus examination (DFE): Dilation was deferred due to corneal perforation on the left eye.

(../cases-i/case222/NLDO-1-LRG.jpg)

Figure 1. *External photograph of the left eye showing copious mucopurulent discharge*

Differential Diagnosis:

- Canaliculitis
- Dacryocystitis
- Bacterial Conjunctivitis
- Blepharitis

CLINICAL COURSE

Due to the large size of the corneal perforation and active presumed bacterial keratitis and conjunctivitis in a monocular patient, corneal cultures were obtained and an emergent mini-penetrating keratoplasty (PKP) was performed. The patient was concurrently placed on a medication regimen consisting of fortified vancomycin, levofloxacin, prednisolone acetate, atropine, and oral trimethoprim sulfamethoxazole. Oral steroids could not be used to control her sarcoidosis as she had previously developed severe psychosis and systemic infection on prednisone.

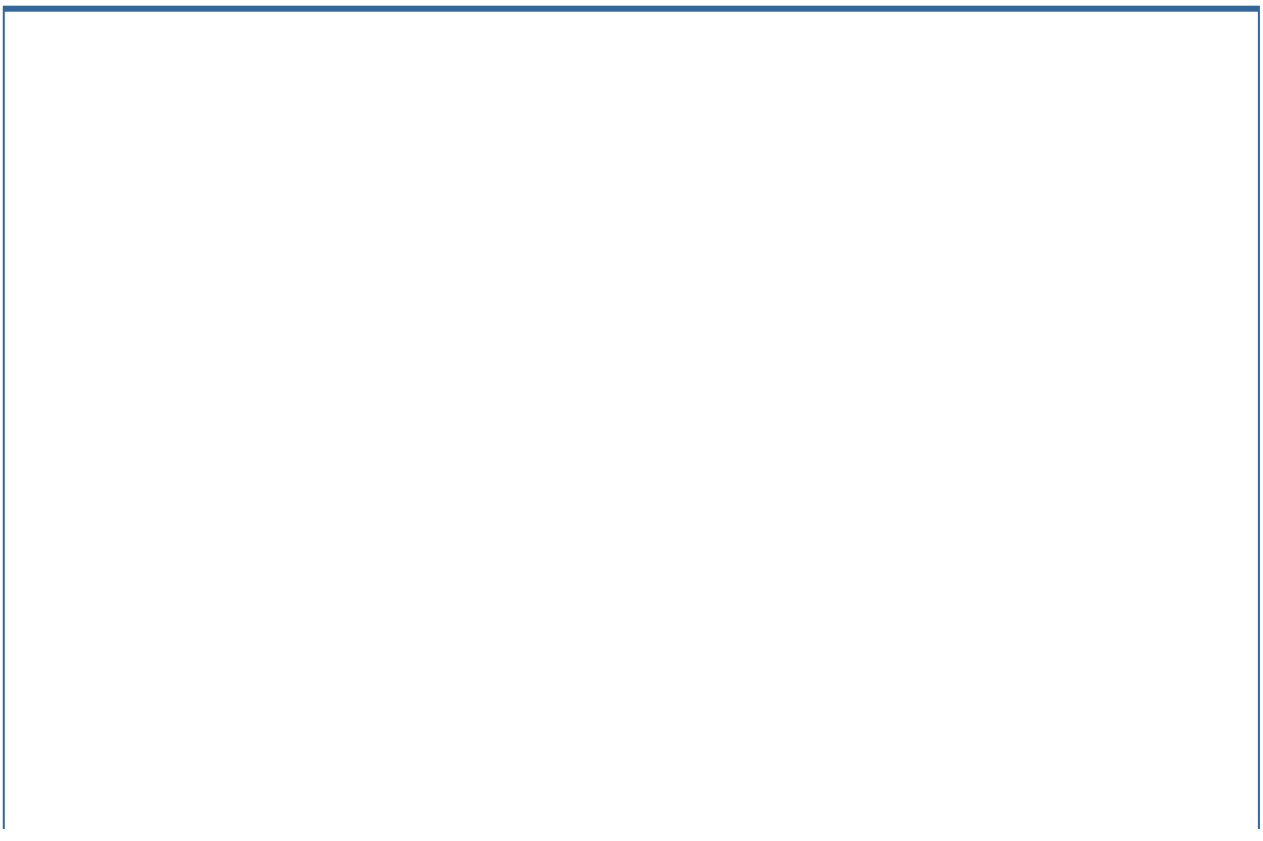
On post-operative day (POD) #1, the corneal perforation appeared to be secure after the mini-PKP and was Seidel negative, however a purulent discharge persisted from the nasolacrimal system. At that point, the oculoplastics service was consulted. On exam, the puncta were not inflamed, and there appeared to be no reflux of material or firmness on compression of the canaliculi. Copious discharge was noted with compression of the nasolacrimal sac, so dacryocystitis secondary to nasolacrimal duct obstruction (NLDO) was suspected. A dacryocystorhinostomy (DCR) was scheduled in one week in order to allow time for the cornea to heal and for targeted systemic antibiotics to decrease the bacterial load.

In the week preceding the DCR, a culture obtained from the cornea grew coagulase-negative staphylococcus. Cultures from the punctal discharge grew *Provotella*, an anaerobe, for which metronidazole was added to her systemic antibiotic regimen.

On POD #6 after the mini-PKP, it was noted that the graft had several loose sutures and was bulging forward. An exam under anesthesia (EUA) was performed at the time of the DCR, and the graft was resutured prior to proceeding with the lacrimal surgery.

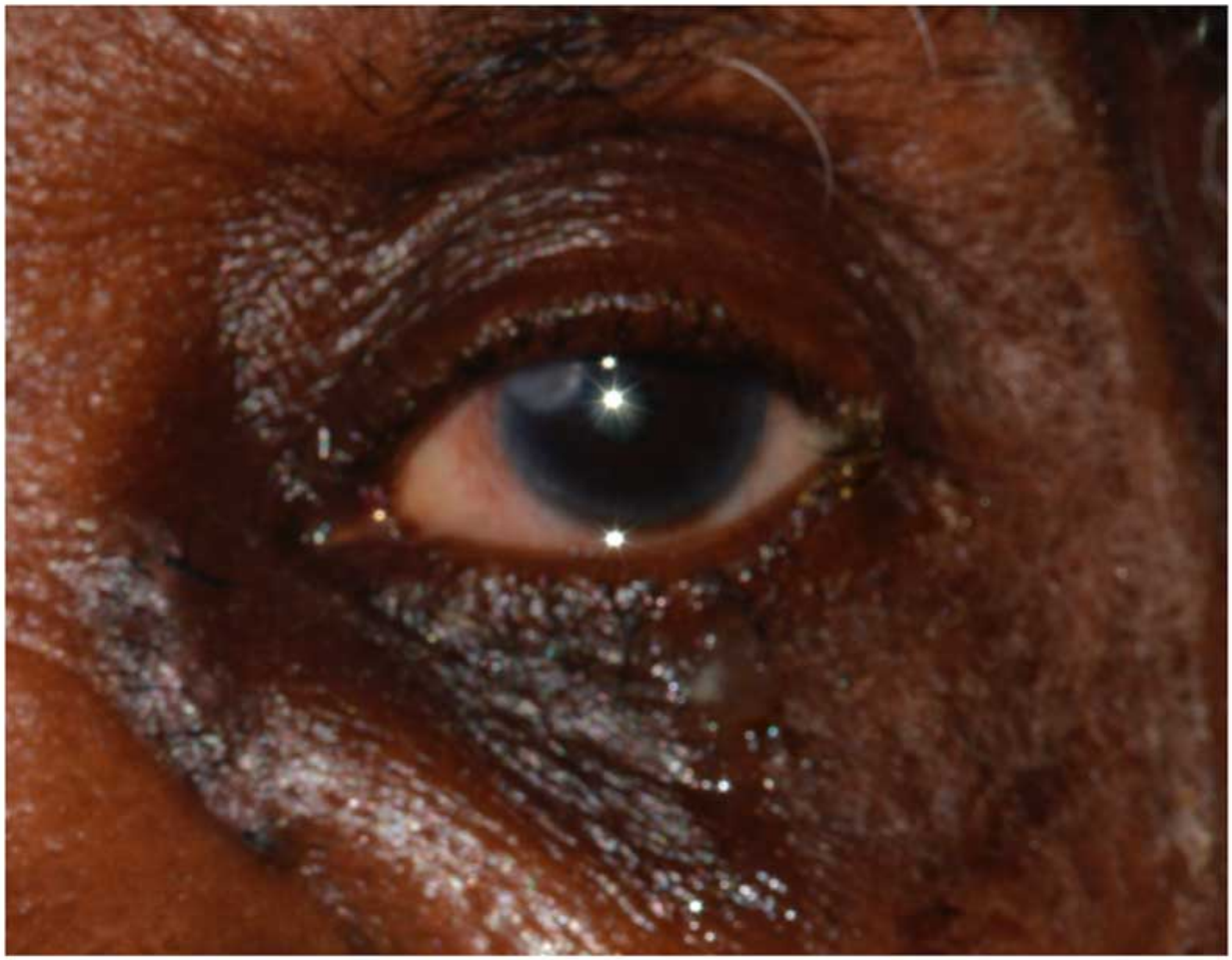
During the patient's DCR, the lacrimal sac was noted to be fibrotic, thickened and surrounded by tissue appearing to be significantly inflamed. Intraoperative frozen sections of the lacrimal sac were sent to pathology and found to be consistent with chronic granulomatous and fibrotic inflammation. Upon further evaluation, the nasal septum was noted to be eroded and necrotic. The surgical plan was reevaluated and discussed with otolaryngology. Rather than forming a traditional ostium for the DCR, a stent was placed just through the nasolacrimal duct in order to dilate the lacrimal system while minimizing manipulation of the nasal mucosa and maintaining more of a barrier between the orbit and necrotic nasal cavity.

Given the monocular status of the patient and the fragility of her corneal graft, ameliorating the purulent dacryocystitis that was the etiology of her corneal perforation was thought to be crucial. The purulence persisted despite the nasolacrimal stenting [Figure 2], so a left dacryocystectomy with cauterization of the puncta and canaliculi were performed. The patient did well post-operatively with resolution of the discharge [Figure 3], no significant epiphora, and with stabilization of the graft [Figure 4] and visual acuity improving to 20/50 8 months post-operatively.



(../cases-i/case222/NLDO-2-LRG.png)

Figure 2: *Pre-dacryocystectomy*



(../cases-i/case222/NLDO-3-LRG.png)

Figure 3: *Post-dacryocystectomy*

(../cases-i/case222/NLDO-4-LRG.jpg)

Figure 4: *Mini penetrating keratoplasty*

DIAGNOSIS

Corneal perforation and chronic dacryocystitis secondary to NLDO from sarcoidosis

DISCUSSION

Etiology/Epidemiology:

Sarcoidosis is a multi-system inflammatory disease of unknown etiology characterized by non-caseating granulomas throughout the body. Orbital and adnexal manifestations of sarcoidosis include granulomatous inflammation causing microarchitectural distortion of the lacrimal gland (63%), eyelids (17%), orbit (13%) and the lacrimal sac (7%) (1). In fact, some studies report sarcoidosis involving the lacrimal sac is an infrequently reported problem, accounting for as little as 1-2% (2). For more information on sarcoidosis, please refer to the EyeRounds case Sarcoidosis affecting the lacrimal gland (135-sarcoidosis-lacrimal-gland.htm).

Chronic inflammatory and granulomatous infiltration of the nasolacrimal duct mucosa can result in an acquired nasolacrimal duct obstruction (NLDO). NLDO can be classified into two groups: primary acquired nasolacrimal drainage obstruction (PANDO) and secondary acquired lacrimal drainage obstruction (SALDO).

Classification of Acquired Nasolacrimal Drainage Obstruction (3)

A. **Primary acquired nasolacrimal duct obstruction (PANDO)**: an idiopathic narrowing of the nasolacrimal duct with associated inflammatory or fibrotic changes of the tissue
B. **Secondary acquired lacrimal drainage obstruction (SALDO):** an obstruction of the nasolacrimal duct secondary to various etiologies such as infection, inflammation, neoplasia, trauma, and mechanics

Infectious

- Infections can cause obstructions anywhere within the lacrimal system, presenting as a punctal abscess, canaliculitis, dacryocystitis, and isolated nasolacrimal duct infections
- Common infection etiologies include actinomyces, staphylococcus, streptococcus, herpes simplex virus, aspergillus

Inflammatory

- **Endogenous causes (by diseases):** sarcoidosis, granulomatosis with polyangiitis (Wegener's granulomatosis), pyogenic granuloma, cicatricial pemphigoid, sinus histiocytosis, and Stevens-Johnson Syndrome
- **Exogenous causes (introduced by other sources):** ophthalmic medications, radiation therapy, systemic chemotherapy, and bone marrow transplantation may all cause punctal and canalicular scarring, stenosis or occlusion (4)

Neoplasia

- **Primary neoplasm (5):**
  - Primary epithelial tumors: papilloma, squamous cell carcinoma, and transitional cell carcinoma
  - Primary non-epithelial tumors: fibrous histocytoma, malignant lymphoma, and malignant melanoma
- **Secondary neoplasm:** squamous cell carcinoma, basal cell carcinoma, adenoid cystic carcinoma, leukemia and lymphoma of adjacent tissues may extend and cause obstruction of the nasolacrimal system
- **Metastasis:** most commonly breast carcinoma; prostate carcinoma and malignant melanoma rarely metastasize to the lacrimal system

Trauma

- **Iatrogenic traumas:** secondary to probing, punctal plugs, and sinus surgery
- **Accidental traumas:** canalicular tears and lacerations, and naso-orbito-ethmoid bone fractures causing nasolacrimal entrapment and damage

Mechanical

- Mechanical NLDO are physical obstructions due to endogenous factors like dacryoliths and migrated punctal plugs or exogenous factors like caruncular masses and sinus mucoceles

Signs and Symptoms

Patients commonly present with epiphora (tearing), mucopurulent discharge, irritation, and blurry vision due to tear accumulation in the conjunctival cul-de-sac. NLDO can be associated with acute or chronic dacryocystitis, lacrimal mucocele, lacrimal abscess, orbital cellulitis, and orbital cellulitis leading to cavernous sinus thrombosis. Further evaluation may show tear meniscus height greater than 2 mm, punctal stenosis, canaliculitis, or reflux discharge from the punctum (3).

Diagnostic Tests

These tests help to determine the location of the obstruction.

A. **Probing and Irrigation:** This is the gold-standard test for evaluating NLDO. Saline is injected into each canaliculus with a 21 or 23 gauge lacrimal cannula on a 3cc syringe (6). Reflux of saline through the same canaliculus indicates obstruction of the canaliculus. Reflux from the opposite punctum indicates obstruction in the nasolacrimal sac or duct. Easy passage of saline into the nose without reflux demonstrates an anatomically patent nasolacrimal system. Slight resistance to irrigation and/or some degree of reflux can indicate a partial obstruction.

B. **Fluorescein Dye Disappearance Test**: A drop of fluorescein dye or paper strip with fluorescein is placed between the eyelid and the eye. After 15 minutes, the dye should be observed using the blue slit-lamp light disappearing into the tear ducts. A large residual of dye left in the eye may indicate an obstruction of the lacrimal system.

C. **Regurgitation Test:** Placing compression over the canaliculi with subsequent efflux of fluid or discharge may indicate obstruction of the canaliculi or canaliculitis. Efflux with compression of the lacrimal sac may indicate lacrimal duct obstruction or dacryocystitis. It is important to note the type of regurgitated material (watery, mucoid, mucopurulent, blood stained) and whether it is coming from the same or opposite punctum.

D. **Dacryocystography (DCG)**: DCG is an imaging study in which contrast dye is injected into the lacrimal system and simultaneously imaged with CT or MRI. It can be beneficial in detailing anatomic obstruction.

NLDO and Corneal Ulcer

The lacrimal apparatus consists of a secretory and drainage system that functions to provide lubrication of the cornea and conjunctiva. The tears secreted by the lacrimal gland spread over the corneal surface with the blinking motion. The tears may disappear due to evaporation or gather in the lacrimal lake near the medial canthus to be drained by the punctum. An obstruction of the drainage system causes tear stasis and an increased number of pathogens within the tears themselves. With an increased number of pathogens circulating in the tears, corneal ulcers are more likely to occur.

Treatment of NLDO secondary to sarcoidosis:

Small, retrospective case series have shown both external and endonasal dacryocystorhinostomy to be beneficial in NLDO secondary to sarcoidosis.

A retrospective case series (2) was performed to investigate the surgical outcomes of external dacryocystorhinostomy (EX-DCR) performed in patients with sarcoidosis. The study included 13 sides of 9 sarcoidosis patients. A subset of patients was also given intralesional triamcinolone. Initial EX-DCR was successful in 10 of 13 (87%) surgeries with an average follow up of 31 months. All the patients receiving intralesional triamcinolone had resolution of epiphora (5 surgeries, 4 patients). This success rate is compared to only 3 of 8 (38%) surgeries in the group without intralesional triamcinolone. The study concluded that EX-DCR surgery can successfully treat NLDO associated with sarcoidosis and that the use of intralesional triamcinolone may provide an added beneficial.

Endoscopic powered-type Endonasal DCR (EN-DCR) and non-endoscopic mechanical EN-DCR, alternative treatments to EX-DCR, has been shown to also be useful in NLDO secondary to sarcoidosis. The surgical outcomes of EN-DCR performed in 18 sides of 12 sarcoidosis patients were investigated retrospectively (7). The success rate was 100% after an average of 11.3 months follow up. While the results of the study are impressive, the low number of cases, short follow up, and variability in steroid and treatment regimen limited the study.

Additional studies comparing EX-DCR and EN-DCR, as well as the role of steroids are warranted.

EPIDEMIOLOGY OR ETIOLOGY	SIGNS
• Infectious • Inflammatory • Neoplastic • Traumatic • Mechanical	• Epiphora • Elevated tear meniscus height (>2mm) • Distention of the nasolacrimal sac in the inferior medial canthal region • Discharge – if dacryocystitis present
SYMPTOMS	TREATMENT/MANAGEMENT
• Epiphora • Mucopurulent discharge • Inferior medial canthal swelling	• Dacryocystorhinostomy (DCR)

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Suggested Citation Format

Scott J., Beebe JM, Shriver EM. Detachment (PVD). EyeRounds.org. posted Oct 21, 2015; Available from: http://EyeRounds.org/cases/222-NLDO.htm

last updated: 10/21/2015

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