BRIEF REPORT

PREVOTELLA DENTICOLA DACRYOCYSTITIS AND ABSCESS IN A CHILD WITH GOLDENHAR SYNDROME

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KEYWORDS

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ABSTRACT

Dacryocystitis is the inflammation of the nasolacrimal sac and is due to obstruction of the nasolacrimal system or lacrimal stagnation, leading to favorable environments for infection. Any disruptions to the nasolacrimal pathway can increase the risk of dacryocystitis. Here is a unique case of *Prevotella denticola* dacryocystitis that progressed into cellulitis and abscess formation in a 7-year-old female with a medical history of Goldenhar syndrome, recurrent dacryocystitis, and periodontal disease. The patient presented with standard dacryocystitis symptoms—tenderness to palpation, erythema, and edema to the medial canthus. The patient was treated empirically with vancomycin, ceftriaxone, and clindamycin. She was discharged home on clindamycin with symptomatic improvement and plans for dacryocystorhinostomy. Goldenhar syndrome is a rare congenital craniofacial malformation disorder due to aberrant morphogenesis in structures derived from the first and second branchial arches. *P. denticola* is an anaerobic gram-negative bacillus significant to the oral microbiome. There is currently limited research suggesting the relationship between Goldenhar syndrome and dacryocystitis complicated by *Prevotella denticola*. Based on the patient's medical history, we predict that her complex presentation was secondary to the dissemination of *Prevotella* from her oral cavity. This case report emphasizes the importance of gram-negative anaerobic coverage in complicated dacryocystitis in patients with nasolacrimal defects.

INTRODUCTION

Dacryocystitis, the inflammation of the nasolacrimal sac, is commonly caused by obstruction of the nasolacrimal duct—either due to the backup or stasis of tears.[1] The nasolacrimal system starts with lacrimal tear production at the main lacrimal gland, drains into the common canaliculus, travels through the valve of Rosenmüller, and into the lacrimal sac.[2] From there, lacrimation continues inferiorly into the nasolacrimal duct, through the valve of Hasner, and into the nasal cavity.[2] There is increased risk for dacryocystitis infections in the setting of abnormalities or alterations to the nasolacrimal system.

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The etiology of dacryocystitis is characterized as acute, chronic, congenital, or acquired. There is a bimodal distribution of causes, with newborns and infants susceptible to acute and congenital cases, while adults over 40 years are prone to chronic and acquired situations.[1] The most causative organisms are gram-positive *Staphylococcus aureus*, coagulase negative *Staphylococcus*, and *Streptococcus pneumoniae*. *Haemophilus influenzae* is the most causative gram-negative organism.[1] Chronic dacryocystitis stems from long-term obstruction secondary to systemic diseases like Wegener's granulomatosis or sarcoidosis, tumor-like lesions, and mechanical obstruction.[3] Acquired dacryocystitis is caused by repeated trauma to the nasoethmoid bones, surgeries (endonasal and endoscopic procedures), or thickening of facial structure and bones.[4]

Congenital dacryocystitis is an obstruction in the distal nasolacrimal duct at the valve of Hasner.[1] It is detected when the amniotic fluid buildup from the womb fails to flow through the nasolacrimal system after delivery. Any congenital malformations to the craniofacial morphogenesis can increase risk of obstruction to the nasolacrimal duct. Goldenhar syndrome, also known as oculo-auriculo-vertebral (OAV) syndrome, is a rare congenital disorder defined as the malformation of the structures derived from the

first and second branchial arches during development, including the eyes, mouth, lip, tongue, palate, ear, maxilla, and mandible.[5] Because of the multiorgan involvement, OAV syndrome can lead to increased susceptibility to infection, inflammation, and issues in the craniofacial area.

Dacryocystitis is a clinical diagnosis based on the patient's history and physical exam, specifically the ocular exam—including normal visual acuity, absence of pain in extraocular movements, location of tenderness, and lack of ophthalmalgia. Individuals tend to endorse an erythematous, edematous, and tender mass at the medial canthus of the eye with additional nasal dorsum involvement.[6] The inflammatory symptoms of dacryocystitis follow the path of the nasolacrimal system; therefore, if erythema or edema is present beyond the nasolacrimal system, alternative diagnoses must be considered. When suspected clinically, dacryocystitis is confirmed by culturing the purulent fluid from the abscess. Radiologic imaging is not required for diagnosis, but computerized tomography (CT) scans can rule out more severe cases of orbital cellulitis.[7] Complication of dacryocystitis can be detrimental, with significant sequelae of orbital cellulitis, lacrimal fistulas, brain abscesses, cavernous sinus thrombosis, meningitis, and death.[1]

Uncomplicated dacryocystitis is treated conservatively with warm compresses and Crigler massage, a manual decompression of the nasolacrimal duct. In acute settings, Crigler massage is not warranted due to increase in hydrostatic pressure from the massage, causing a vulnerable environment for the spread of infection. Broad-spectrum antibiotics that cover gram-positive and gram-negative organisms, especially antistaphylococcal agents, are considered in the acute phase.[1] Initially, intravenous (IV) therapy of penicillin and cephalosporins are indicated. Vancomycin is the drug of choice in the setting of methicillin-resistant *Staphylococcus aureus* (MRSA) while clindamycin should be considered in the setting of anaerobic coverage.[6] In recurrent cases of dacryocystitis, ophthalmologists should be consulted for potential nasolacrimal probing, a highly successful technique used to irrigate the nasal cavity. In more advanced cases, invasive procedures like balloon dacryoplasty, nasolacrimal intubation, or nasolacrimal stenting are available.[3] Definitive therapy is a dacryocystorhinostomy, a new pathway for the tears to flow through the nasolacrimal sac and into the nose.[3]

CASE PRESENTATION

A 7-year-old Caucasian female with a past medical history of Goldenhar syndrome with mild dysmorphic features, type 1 Duane retraction syndrome, prematurity, and extensive dentoalveolar concern presented to inpatient pediatrics with bilateral infraorbital erythema with pronounced edema in the medial aspect of the right eye that exhibited significant tenderness to palpation and cellulitis-like symptoms (Figure 1). The patient denied any fevers or recent sick contacts. Immunizations were up to date. Her foster mother reported that the patient had a history of lacrimal duct infections that were similar to this presentation but less tender. The most recent episode happened the week prior, and the patient was managed with a 7-day course of amoxicillin. Her social history is significant for substance abuse in her biological mother, neglect and malnourishment by her biological parents, and involvement of the Department of Social Services (DSS).

On admission, the patient was afebrile with vitals showing signs of tachycardia at 127 beats per minute and tachypnea at 24 breaths per minute. The patient's initial labs were significant for signs of inflammation and infection with an elevated C-reactive protein of 12.3 mg/dL (reference range: 0.0-0.6 mg/dL), elevated erythrocyte sedimentation rate of 66 mm/h (reference range: 0-20 mm/h), and leukocytosis at 14.4 x10^3/µL (reference range: 4.5-13.5 x10^3/µL). Aside from her facial erythema and edema, the patient's physical exam was unremarkable. During the first few hours, the patient had a temperature of 102.5°F (39.2°C) that was decreased to 98.1°F (36.7°C) with acetaminophen and ibuprofen.

Dacryocystitis was suspected, and IV ceftriaxone and vancomycin were prophylactically started while awaiting blood culture results. The patient was not adequately responding to the antibiotic therapy; therefore, vancomycin was switched to IV clindamycin for anaerobic coverage. Despite antibiotics and warm compresses, the patient continued to experience progressively worsening right-sided infraorbital tenderness.

FIGURE 1:

(Left) Bilateral infraorbital erythema with notable edema medial to the right eye

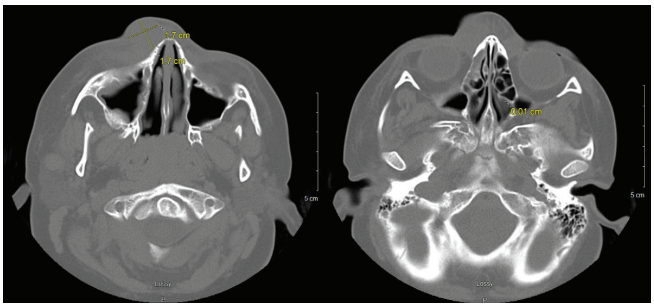
(Middle) Postrupture of abscess and purulent drainage

(Right) Prior to discharge: significant improvement of erythema, edema, and tenderness

Ophthalmology and otolaryngology services were both consulted and recommended a CT scan without contrast of the sinuses. It showed prominent soft tissue density along the right nasal bridge and frontal process of the maxilla that extended along the right premaxillary soft tissues and periorbital region (Figure 2). Most of the infection appeared to be localized over the lacrimal duct without orbital involvement. The intraconal and extraconal fat planes were preserved with no evidence of postseptal extension of the soft tissue density, but moderate circumferential mucosal thickening of the right maxillary sinus with occlusion of its ostium was shown. Additionally, focal dehiscence and disruption of the anterior alveolar cortex were visible and associated with an unerupted right paramedian maxillary tooth.

FIGURE 2:

CT without contrast of sinuses



CT showed prominent soft tissue density along the right nasal bridge and frontal process of the maxilla measuring 1.7 x 1.7 cm that extended along the right premaxillary soft tissues and periorbital region—right greater than left.

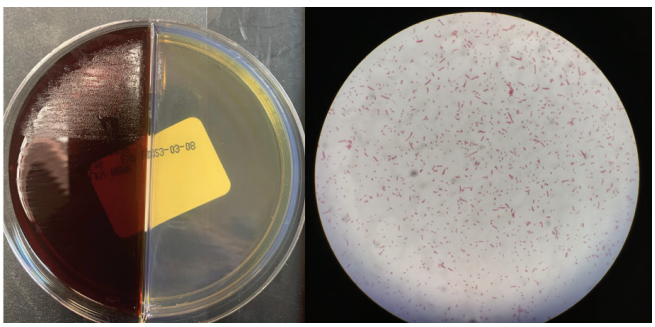
Moderate applied warm compress pressure was placed over the infected area leading to the rupture of the abscess and purulent drainage. Initial blood culture showed moderate gram-positive cocci and wound cultures exhibited preliminary results of anaerobic isolates. After the abscess ruptured, the patient responded to the antibiotic therapy, and her mother reported that the patient was back to her baseline—without nasolacrimal tenderness or purulent drainage. Therefore, the patient was discharged home with a 7-day course of oral clindamycin with plans to consider ocular ductal plasty reconstruction or dacryocystorhinostomy of her left ductal duct with her ophthalmologist in the future.

The patient's abscess cultures macroscopically showed growth of pinpoint colorless colonies on Laked Brucella Blood agar with Kanamycin and Vancomycin (LKV) but had a notable absence of growth on Bacteroides Bile Esculin agar (BBE) under anaerobic conditions for 48 hours. Microscopically, gram stains of isolated colonies revealed short gram-negative coccobacilli rods. Prevotella denticola was confirmed on matrix-assisted laser desorption/ionization-time of flight (MALDI-TOF) mass spectrometry (Figure 3).

(Left) Light growth of pinpoint colorless colonies on LKV agar and absence of growth on BBE agar under anaerobic conditions for 48 hours. (Right) Gram-negative coccobacilli rods seen on isolated colonies. Several organisms show morphologic elongation and ballooning degeneration, likely attributable to cell wall–active antimicrobials at the time of sample collection.

FIGURE 3:

Macroscopic and microscopic lab results



DISCUSSION

This patient has a notable medical history of Goldenhar syndrome, recurrent dacryocystitis, and dentoalveolar concerns and presented to the hospital with acute dacryocystitis and preseptal cellulitis. We believe that the *P. denticola* infection complicated her acute dacryocystitis due to her history of craniofacial anomalies and extensive periodontal disease.

The patient's Goldenhar syndrome presented with mild dysmorphic facial features at birth—a narrow palpebral fissure and a right-sided preauricular skin tag. Her Goldenhar syndrome is complicated by right esotropia, also known as type I Duane retraction syndrome. The craniofacial malformations associated with Goldenhar syndrome are secondary to disrupted migration and proliferation of the mesenchymal and neural crest cells of the first and second branchial arches. This is believed to be secondary to a disruption in the embryonic vascular supply between weeks 4 and 8 of gestation.[5] Risk factors for Goldenhar syndrome are sporadic and multifactorial, incorporating chromosomal aberrations and environmental causes. Reported maternal conditions associated with Goldenhar syndrome include maternal diabetes, multiple gestations, assisted reproductive techniques, hormonal therapy, maternal hypothyroidism, vasoactive drug use, tobacco use, cocaine abuse, and malnourishment.[5] Upon further review, we found that the patient had a history significant for these risk factors.

There is limited research discussing the association between lacrimal gland abnormalities and Goldenhar syndrome. When documented, the association is hypothesized to be related to dysembryogenesis. During the seventh week of gestation, the nasolacrimal system forms as a cord of ectodermal cells extending from the nasal cavity.[8] Throughout gestation, this system develops and canalizes. The last portion of the nasolacrimal system to recanalize is the inferior meatus, the most common site of obstruction in lacrimal defects.[2] When reported, lacrimal anomalies related to Goldenhar syndrome were found in the form of obstruction to the nasolacrimal duct or common canaliculus.[8] We believe the disruption of embryonic vascular supply during early gestation contributes to nasolacrimal malformations associated with Goldenhar syndrome.

Dacryocystitis has been reported to potentially be complicated by more serious infections like orbital cellulitis, meningitis, lacrimal abscesses, and cavernous venous sinus thrombosis.[1] This patient's case was complicated by a lacrimal abscess. With *S. aureus*, *S. pneumoniae*, and *H. influenzae* being the most causative organisms for dacryocystitis, the patient was started on broad-spectrum, empiric gram-positive, and gram-negative coverage of vancomycin and ceftriaxone. However, the patient's symptoms were not improving, so vancomycin was switched to clindamycin, an antibiotic that covers both MRSA and anaerobes. Anaerobic coverage was crucial in this scenario as her culture grew *P. denticola*, supporting the severity of the patient's presentation. *Prevotella* spp. produces beta-lactamase, which contributes to its virulent antimicrobial resistance.[9] Although susceptibility patterns of *Prevotella* spp. are limited, most are susceptible to piperacillin-tazobactam, imipenem, meropenem, tigecycline, and metronidazole.[9] Intrinsic resistance of *Prevotella* spp. to beta-lactam antibiotics coupled with recurrent treatment of dacryocystitis with amoxicillin may contribute to the development of opportunistic infection through altered

homeostasis of the natural oral microbiome.

The *Prevotella* spp. is classified as an anaerobic gram-negative bacilli and is one of the core mucosal anaerobes of the oral, respiratory, and gastrointestinal microbiome.[10] This species contributes to the production of bacterial biofilm in the oral cavity and can be involved in chronic inflammation and the progressive destruction of periodontal tissue.[11] When the mucosal microbiome is compromised, the reduction or overabundance of *Prevotella* spp. can cause inflammatory infections.[10] An increase in *Prevotella* spp. is seen in periodontitis, bacterial vaginosis, and low-grade systemic inflammation due to the mucosal spread of the bacteria throughout the body.[10]

In terms of periodontal infections, the patient had an extensive dentoalveolar procedure done 2 years ago with dental restorations. She had eight tooth extractions and 12 pulpotomies with stainless steel crowns. Evidence-based research has supported the gut-retina microbiome axis hypothesis, describing the relationship between the oral microbiome and ocular infections. Periodontal disease is known to have an association with inflammatory disease states. At this time, although the exact pathophysiologic mechanism is unknown, current literature supports three hypotheses including transmigration, pro-inflammatory molecule production, and hematogenous spread throughout the nasolabial triangle.[12] The nasolabial triangle, bound by the right and left oral commissures to the nasal bridge, is highly vascularized and full of intricate anastomoses, which can easily communicate infections between the oral cavity and the nasolacrimal system.[12] We suspect that *P. denticola* from the patient's oral cavity disseminated via this route to her nose and laid dormant until it had the opportunity to disrupt the nasal microbiome and proliferate.

CONCLUSION

We discussed a 7-year-old female with a medical history notable for Goldenhar syndrome, multiple dental surgeries, and recurrent dacryocystitis infections previously treated with amoxicillin, who presented acutely for bilateral infraorbital edema, erythema, and tenderness. She was hospitalized for acute dacryocystitis complicated by *P. denticola*, and she was successfully treated with IV ceftriaxone and clindamycin for broad-spectrum and anaerobic coverage. We hypothesized that her anatomic anomalies associated with Goldenhar syndrome, her history of periodontal disease, and her chronic history of dacryocystitis contributed to the spread of *P. denticola* to her lacrimal sac. We could not find any case reviews or reports describing acute dacryocystitis complicated by *P. denticola* infection in patients with Goldenhar syndrome and periodontal disease. Furthermore, there is limited research regarding Goldenhar syndrome and dacryocystitis complicated by opportunistic organisms. This case accentuates the significance of considering gram-negative and anaerobic coverage in complicated acute recurrent dacryocystitis for successful clinical outcomes, especially in those with developmental malformations of the craniofacial region.

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