Bilateral Congenital Lacrimal Fistulae: A Case Report and Review of the Literature

Lei Zhuang, MD\textsuperscript{1}, Christin L. Sylvester, DO\textsuperscript{2}, and Jeffrey P. Simons, MD\textsuperscript{1}

Departments of Otolaryngology\textsuperscript{1} and Ophthalmology\textsuperscript{2}
Children's Hospital of Pittsburgh and University of Pittsburgh School of Medicine, Pittsburgh, PA

\section*{ABSTRACT}

The lacrimal system is comprised of the lacrimal glands for tear production and the lacrimal drainage system for draining tears away from the eyes. Congenital lacrimal system anomalies other than nasolacrimal duct obstruction are uncommon. Congenital lacrimal fistulae are a rare developmental anomaly, and when they occur, they are usually unilateral. Fistulae and diverticulae can originate from the common canaliculus, lacrimal sac, or nasolacrimal duct. They can be seen externally as small orifices or pits located inferior and/or medial to the medial canthi. These anomalies are often asymptomatic and may go undetected. However, they can also present with epiphora or discharge.

We report the case of a 4-year-old male with bilateral congenital lacrimal fistulae. The patient presented to our clinic after his parents discovered bilateral pits located inferior and medial to the medial canthi. The pits had first been noticed only a few weeks prior to presentation. There was no history of drainage or infection. The diagnosis of bilateral congenital lacrimal fistulae was confirmed with computed tomographic imaging. No other systemic, nasal, or ocular anomalies were found. The patient was referred to pediatric ophthalmology and because the lacrimal fistulae have been asymptomatic to this point, it was decided to proceed with a course of observation.

We will discuss the embryologic basis for congenital lacrimal fistulae, as well as the typical presentation and possible treatment modalities. The presence of lacrimal fistulae is an indicator to search for a variety of underlying systemic and ocular anomalies.

\section*{CASE REPORT}

A 4-year-old male presented to our otolaryngology clinic a few weeks after his parents made an incidental discovery of bilateral pits located inferior and medial to the medial canthi. There was no history of drainage or infection. The pits were 1 mm in size and overlying the lacrimal sac on both sides of the nose. There was a small amount of dried sebaceous material found in the pits. No discharge was elicited from the punctae when pressure was applied to the lacrimal sacs. Computed tomographic imaging was obtained, confirming the diagnosis of bilateral congenital lacrimal fistulae. The patient was referred to pediatric ophthalmology to rule out associated ocular abnormalities, and no evidence of a complete lacrimal-cutaneous fistula was found on transillumination. The rest of the ocular examination was normal. No other systemic, nasal, or ocular anomalies were found. No other family members were found to have lacrimal fistulae. Since the lacrimal fistulae were asymptomatic, the decision was made to manage the patient conservatively with close observation.

\section*{DISCUSSION}

The nasolacrimal apparatus arises embryologically from a cord of surface ectoderm that invaginates between the maxillary and frontonasal processes, giving rise to the canaliculi proximally, and the lacrimal sac and nasolacrimal duct distally. Congenital lacrimal fistulae are an indicator to search for a variety of underlying developmental anomalies, and when they occur, they are usually unilateral. Fistulae and diverticulae can originate from the common canaliculus, lacrimal sac, or nasolacrimal duct. They can be seen externally as small orifices or pits located inferior and/or medial to the medial canthi. These anomalies are often asymptomatic and may go undetected. In some cases, symptoms such as drainage of a mucoid discharge or epiphora may necessitate surgical excision. While most fistulae are unilateral, familial cases are associated with a higher incidence of bilateral fistulae. The location of congenital lacrimal fistulae is characteristically infero-nasal to the medial canthal angle.

Our review of the existing literature on lacrimal fistulae revealed reports of associations with other abnormalities such as preauricular fistulae, hypospasias, and VACTERL syndrome (vertebral anomalies, anal atresia, cardiac malformations, tracheo-esophageal fistulae, renal anomalies, and limb anomalies). Congenital lacrimal fistulae can also be associated with thalassemia and Down syndrome. Associated ocular pathology including dacrocystitis, lacrimal tract stenosis, and infections of the lower eyelid, as well as hypertelorism and strabismus have also been reported.

A wide range of treatment modalities for symptomatic lacrimal fistulae have been discussed in the literature. They range from nasolacrimal duct probing and cautery of the external osulum to surgical excision of the fistula, either alone or in combination with dacryocystorhinostomy. For patients who are asymptomatic (as in our case) or minimally symptomatic, observation is a reasonable approach.

\section*{REFERENCES}