

Case Report:

Congenital Lacrimal Sac Fistula: A Case Report and Literature Review

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Key Words:Fistula, Congenital,
Lacrimal sac, Surgery**ABSTRACT**

Introduction: Congenital lacrimal sac fistula is a rare anomaly of the lacrimal system in which, an epithelium-lined lacrimal tract is connected to the skin through a fistula. In most cases, it usually presents as an asymptomatic unilateral lesion located inferolateral to the medial canthus. Several studies reported that congenital lacrimal sac fistulas may be associated with systemic anomalies like Down syndrome, renal agenesis, etc. However, in this case, we aimed to report a patient with isolated bilateral congenital lacrimal sac fistula and its surgical management.

Case presentation: A previously healthy 17 years old female presented with the complaint of intermittent watery discharge from her left eye since a few months ago. Physical examination revealed a small orifice in the skin at about 4 mm from the medial canthus on the medial side in both eyes without any signs of inflammation. Probing and irrigation in the left eye disclosed communication with the puncta and nose. Surgical repair was carried out through fistulectomy which successfully controlled the patient's lacrimation.

Conclusions: Recent advances in surgical techniques have shown better rates of success in relieving the patient's lacrimation through fistulectomy than the simple excision methods. Our experience with the case presented here demonstrates that this lacrimal system anomaly can be treated successfully by surgical intervention.

1. Introduction

A Congenital lacrimal sac fistula is a rare anomaly of the lacrimal system in which, an epithelium-lined lacrimal tract is connected to the skin (1). Most cases are unilateral and located inferolateral to the medial canthus. In most fistulas, the lesions are asymp-

tomatic and are found accidentally during routine clinical examinations (2). This report aimed to describe an interesting case of a bilateral congenital lacrimal sac fistula that was presented with unilateral epiphora managed surgically to alleviate the patient's symptoms.

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2. Case Presentation

A generally healthy 17-year-old girl was presented with the chief complaint of intermittent watery discharge from her left eye since a few months ago. The slit-lamp biomicroscopic examination of the anterior segment and funduscopy were unremarkable. Ocular motility was also normal. Her family history was negative for any similar problems. Two small orifices were detected in the skin at about 4 mm from the medial canthus on the medial side in both eyes (Figures 1 and 2). She did not have any complaints about her right eye at the time of referral. Probing and irrigation of the cutaneous opening of fistula in the left eye disclosed communication with the puncta and nose. The opening was cleared with no evidence of any inflammation.

An operation was performed under general anesthesia. We clarified that the fistula was connected to the lacrimal drainage system by simultaneous guided probing of the skin orifice and punctum until they were in contact with each other. The evaluation of the lower lacrimal system revealed the patency of the nasolacrimal duct. In the left eye, fistulectomy was carried out after performing a fusiform skin incision around the fistulous opening. The base of the fistula was closed with a 6-0 Vicryl suture, and the surrounding skin was enclosed using a 6-0 silk suture (Figure 3). The patency of the lacrimal drainage system was confirmed using fluorescein dye irrigation.

In histopathology examination, the excised tracts revealed duct lined by stratified squamous epithelium. At 4 months of postoperative follow-up examination, the patient declared resolution of epiphora, and no skin hole was detectable. She also did not report any complaints about her right eye.

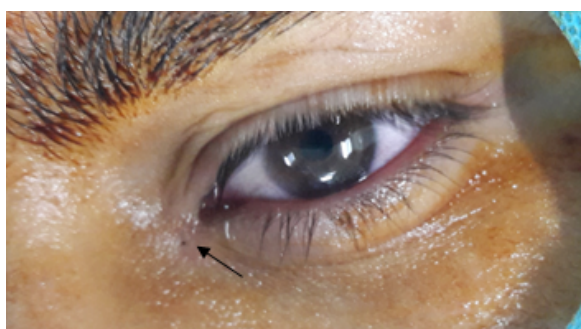
The patient was informed of being reported in medical journals and ensured her confidentiality and written informed consent was also obtained.

3. Discussion and Review of Literature

The ectoderm cord on the surface constitutes nasolacrimal apparatus embryologically to form the canaliculi proximally, and the lacrimal sac and nasolacrimal duct distally. The embedded ectodermal cord is canalized throughout the length of the nasolacrimal apparatus. Aberrant out-budding of the embedded ectodermal cord or incomplete disconnection of the cord from the surface epithelium can lead to exceptionally extra puncta and canaliculi (3). These developmental anomalies of lacrimal fistulas are rare with an estimated occurrence of 1 in 2000 live births (4).

Although most cases of these fistulas remain asymptomatic, a complaint of watery discharge is often delayed for many years because of the evaporation of small amounts of tear leakage (5). In our case, the patient had a history of intermittent epiphora for many months before referring, so the case was discovered rather late. Up until a few years ago, a simple skin excision with electrocauterization was usually used to alleviate the patient's symptoms with little success, but recently, better results can be obtained with simple fistulectomy in most cases (6). In this case, fistulectomy alone relieved the patient's lacrimation.

Anomalies of the lacrimal apparatus can cause partial or complete obstruction of the lacrimal drainage system. These anomalies include punctal atresia, canalicular atresia, dacryostenosis, sac diverticula, and congenital fistula, present in one eye or extremely rare in both eyes (7, 8). The lesions are mostly located inferolateral to the medial canthus; however, atypi-



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Figure 1. A small skin hole of lacrimal fistula located approximately 4.0 mm medial to the medial canthus (arrow) of the left eye in an Iranian girl



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Figure 2. A small asymptomatic skin hole of lacrimal fistula located medial to the medial canthus of the right eye



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Figure 3. Fistulectomy in the left eye

Closure of the opening using 6-0 chromic, and closure of the skin using 6-0 silk sutures

cal cases of congenital lacrimal sac fistula have also been reported, located on the temporal side of the lateral canthus (9). Lacrimal fistula can be associated with systemic anomalies. Singh et al. and Keserü et al.

separately described two cases of congenital bilateral lacrimal fistulae with nasolacrimal duct obstruction in a patient with Down syndrome managed by bilateral probing and fistulectomy (10, 11).

Bilateral lacrimal fistula has been previously reported in a patient with the VACTERL (vertebral anomalies, anal atresia, cardiac malformations, tracheoesophageal fistula, renal anomalies, and limb anomalies) (17). Some of these associations with systemic anomalies are listed in Table 1.

4. Conclusion

Our case demonstrates that this lacrimal system anomaly can be treated successfully by surgical intervention.

Table 1. A few previously reported congenital lacrimal fistula (CLF) associated with systemic anomalies

Author(s)	Publication Year	Associated Anomalies	Case(s)
Sullivan et al. (12)	1992	Down syndrome	Five cases of unilateral CLF
Al-Salem et al. (13)	2014	Down syndrome	Two patients out of 15 cases had CLF
Lee et al. (14)	2012	Down syndrome	Report of 8 cases of CLF in 198 Down syndrome patients
Keserü et al. (11)	2010	Down syndrome	A case of unilateral dacryocystitis with CLF
Singh et al. (10)	2013	Down syndrome	A 3 years old female with bilateral CLF
Satchi et al. (15)	2010	Down syndrome	One case out of 23 patients with CLF
Harrison et al. (17)	2002	VACTERL	A 19 years old man with VACTERL associated with bilateral CLF
Mukherji et al. (16)	1972	Preauricular sinuses	A case with fistula
Satchi et al. (15)	2010	Preauricular sinuses	Two cases out of 23 patients with CLF
Ghosh et al. (18)	2015	Ectrodactyly-ectodermal dysplasia-clefting syndrome	A 17 years old male with lobster claw deformity and CLF
Elmann et al. (19)	2015	Ectrodactyly-ectodermal dysplasia-clefting syndrome	A 9 years old girl with Lobster claw deformity and CLF
Tien et al. (20)	2006	Ectrodactyly-ectodermal dysplasia-clefting syndrome	A case of lobster claw deformity with bilateral CLF
Altun et al. (21)	2015	Renal agenesis and ectopic pelvic kidney	A 12 years old boy with renal agenesis and CLF
Turan-Vural et al. (22)	2012	Uterus didelphys and renal agenesis	A 30 years old female with uterus didelphys, renal agenesis, and CLF
Onaran et al. (23)	2009	CHARGE syndrome	A case of multisystem anomalies with CLF
Michaelides et al. (24)	2002	Hypospadias and hypertelorism	A case of urogenital and orbital anomalies with CLF
Akdemir et al. (25)	1991	Orbital meningocele	A case of right naso-orbital meningocele associated with bilateral CLF

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CHARGE: Coloboma, Heart defects, Atresia choanae, Growth Retardation, Genital abnormalities, and Ear abnormalities; CLF: Congenital Lacrimal Fistula; VACTERL, Vertebral anomalies, Anal atresia, Cardiac malformations, Tracheoesophageal fistula, Renal anomalies, and Limb anomalies.

Ethical Considerations

Compliance with ethical guidelines

The study was done in compliance with ethical guidelines and the Declaration of Helsinki. Informed consent was obtained from the study case.

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Authors' contributions

All authors contributed in preparing this article.

Conflicts of interest

The authors declared no conflict of interest.

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