

Congenital lacrimal fistulas with secondary infection mimicking acute dacryocystitis: a case report and literature review

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Received: 2021-03-18 Accepted: 2021-12-06

DOI:10.18240/ijo.2022.06.24

Citation: Xu PF, Zhong XC, Shi X, Ye J, Wu H. Congenital lacrimal fistulas with secondary infection mimicking acute dacryocystitis: a case report and literature review. *Int J Ophthalmol* 2022;15(6):1023-1026

Dear Editor,

We reported a rare case of congenital lacrimal fistula with recurrent cellulitis secondary to fistulitis, mimicking acute dacryocystitis. Congenital lacrimal fistula is an anomalous development of the lacrimal drainage system manifested by a patent communicating with skin on one side and the canaliculus, lacrimal sac, or the duct on the other^[1]. Most congenital lacrimal fistulas are asymptomatic, or minimally symptomatic. Some may have symptoms when coughing, blowing the nose, or in the case of hyperlacrimation, such as in windy weather^[2]. However, symptomatic cases typically present with epiphora from the fistula or the eye, especially in patients with nasolacrimal duct obstruction. Rare few patients may, on occasion, demonstrate recurrent inflammatory symptoms, such as dacryocystitis, canaliculitis, blepharitis or fistulitis^[3-5]. The diverse clinical features may result to high rate of misdiagnosis.

The retrospective study was reviewed and approved by the Ethics Committee of the Second Affiliated Hospital of Zhejiang University, and followed the tenets of the Declaration of Helsinki. Written informed consent was obtained from the

individual(s) for the publication of any potentially identifiable images or data included in this article.

A 3-year-old female child was referred to our eye center for evaluation of recurrent cellulitis manifested with eyelid redness and swelling at the right lower eyelid and medial canthal (Figure 1A, 1B). Two weeks before our examination, the swelling developed again, and a subcutaneous mass was formed at the right medial canthal region. Her parents reported three previous episodes of presumed right-sided dacryocystitis within 2y but denied having epiphora in remission stage. During the previous episodes of acute inflammation, the punctums at medial canthal region were ignored, and the patient was treated with repeated stab incision and drainage in other medical institutions. Due to poor outcomes and repeated episodes, the patient once was advised to undergo dacryocystorhinostomy (DCR) surgery. Upon our examination, the patient was firstly found to have bilateral pits located at the medial canthal region (Figure 1C, 1D). A skin scar of the right lower eyelid from previous incision and drainage was found. The child did not suffer from epiphora. No discharge was elicited from the puncta or fistulas when pressure was applied to the lacrimal sac. When lacrimal irrigation through the lower puncta was performed, the fluid passed into the nose and no reflux from the puncta or fistulas was found, which implied that the lacrimal drainage system was functioning normally. By probing of the cutaneous opening of the right side, we revealed that the probe could not reach the lacrimal sac, which suggested that the connection between the fistula tract and the lacrimal sac was closed presumably due to the repeated inflammation. There was no history of systemic diseases, trauma, or eyelid surgeries. No other nasal or ocular anomalies were found, and no family member had a lacrimal fistula. The diagnoses of bilateral congenital lacrimal fistula and right-side cellulitis secondary to fistulitis were made.

The decision to excise the right-side fistula alone was based upon the presence of recurrent fistulitis but free of any other lacrimal symptoms. A fusiform incision was made around the orifice along the skin tension lines. The dilated fistula tract was carefully dissociated and completely excised from the

adjacent tissue with a micro-scissors (Figure 2A, 2B). After that, the lacrimal canaliculi were irrigated with methylene blue to ensure that no damage was done to the lacrimal drainage system during the surgery. Moreover, the passage of fluid from the lacrimal duct to the nose was confirmed by the presence of methylene blue on endoscopy. The incision was closed in 2 layers with 6-0 Vicryl. Histologic examination of the specimen showed fibrous capsule covered with stratified squamous epithelium, indicating that the most possible origination was in common canaliculus (Figure 2C, 2D). After follow up of 13mo, the patient was symptom free with no recurrence of inflammation or fistula (Figure 3). The fistula had healed well with no obvious scarring. The left-side fistula was asymptomatic and kept under observation.

Congenital lacrimal fistula is not common. The incidence of this disorder had been reported to be 1 in 2000 births^[6-7], which might be underestimated due to referral bias. The rate of bilateral lacrimal fistula increased when associated with familial inherited^[8] or syndromal cases^[9-11]. Most of the fistulas are asymptomatic, non-progressive and inconspicuous due to their small size and lack of pigmentation around, and therefore are frequently overlooked^[12]. Symptomatic cases may present with epiphora or mucoid discharge from the fistula^[8,13], the eye or both when associated with functional or anatomical nasolacrimal duct obstruction. In very rare cases, the lacrimal fistulas suffer to secondary infection, and present redness, swelling and pain at the medial canthal region^[3,12], mimicking acute dacryocystitis. In our case, the patients with recurrent cellulitis secondary to fistulitis were firstly misdiagnosed as acute dacryocystitis, and treated with repeated stab incision and drainage.

Comprehensive evaluation is of great importance to make definite diagnosis, which can be divided into history taking, local examination, lacrimal system evaluation and ancillary investigation. A detailed history will provide a clue to the appropriate diagnosis in most cases. The history should include details about the onset, frequency, type, intermittency, laterality of the symptoms, any previous treatment, *etc.* History of previous of trauma, medical therapy, or surgical intervention like lacrimal surgery, probing, incision and drainage should be elicited as it has a bearing on differential diagnosis and management decision. Local inspection should include the face and periorbital region. Position, site, size of the punctum, any swelling or mass in the lacrimal sac area, presence of any skin scar from previous surgery or fistula, as well as other ocular and lacrimal anomalies should be noticed. Type of the regurgitated material (watery, mucoid, mucopurulent, blood stained) and where it is coming from (the same or opposite punctum, the fistula, or both) should be noted when pressure over the lacrimal sac^[14]. In our case, a punctum at medial

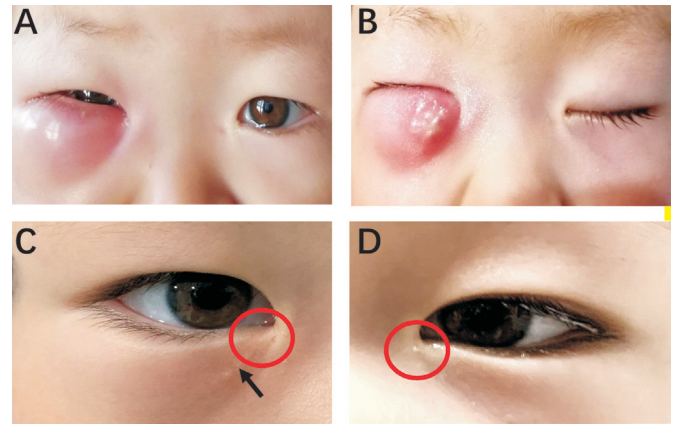


Figure 1 Color photographs of right-side cellulitis and bilateral lacrimal fistulas Two previous episodes of pre-septal cellulitis with swelling and redness of lower eyelid (A, B). The lacrimal fistulas are seen just inferior nasally to the medial canthal angle, the black arrow pointed a skin scar from previous incision and drainage (C, D).

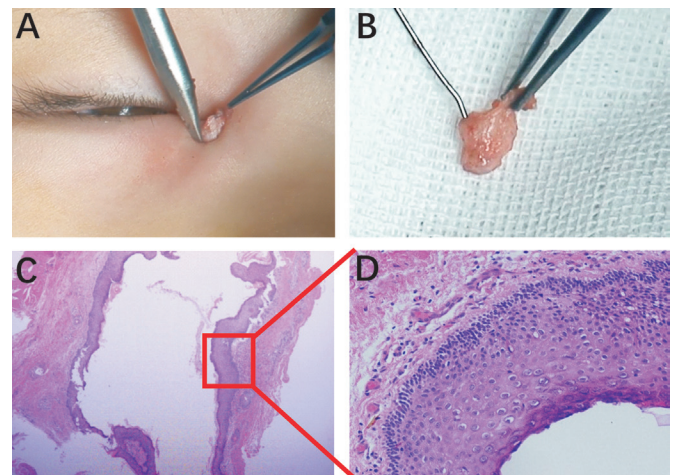


Figure 2 Photographs of the procedure of fistula excision (A, B) and histopathology examination of the fistula (C, D) Clinical photograph showing the dissociation of fistula (A) and the completely excised fistula (B). Histopathology evaluation showing superficial part of the fistula lined by keratinized squamous epithelium (C, D).



Figure 3 Last follow up (13mo) after lacrimal fistula excision.

canthal region was ignored, leading to the several episodes of misdiagnosed and improper treatments.

A systematic assessment of fistula and lacrimal drainage system helps to choose the proper treatments. Various investigations are suggested to visualize the anatomy of congenital lacrimal fistula and the patency and function of lacrimal drainage

system, such as fluorescence dye disappearance test (FDDT), lacrimal irrigation, dacryocystography and so on. FDDT is a noninvasive test for evaluating the excretory function with high specificity and positive prediction value^[15-16]. However, in some cases of fistulas, the FDDT may not show patency of the lacrimal system, since fistulas with high outflow can drain all the contrast and the lacrimal system may stay out of the fluorescein. Lacrimal irrigation is an anatomical test which checks for the patency of the lacrimal duct. The type of regurgitation and where it is from should be noted and well interpreted. However, when a fistula is present, lacrimal irrigation may not be efficient because the liquid can flow throughout the fistula. Thus, it can be necessary to block the fistula before injecting. Dacryocystography can outline the lacrimal outflow pathway and the area of blockage with radiopaque dye injection^[17-18]. In some cases, dacryocystography can clearly demonstrate the presence of accessory canaliculus causing the lacrimal fistula as well as its origin. Radiological investigations for evaluation of the lacrimal system are indicated in selected cases where other anatomical and physiological tests cannot provide a conclusive diagnosis^[14,19-20]. In our case, lacrimal irrigation implied that the lacrimal drainage system was functioning normally. Thus, we did not perform dacryocystography or radiological investigation.

Most patients with congenital lacrimal fistulas are asymptomatic and usually remain undetected, and it is uncommon for asymptomatic congenital fistula to cause epiphora suddenly. It is universally acknowledged that patients who are asymptomatic or have very minor symptoms should merely be kept under observation^[21-22]. While for symptomatic cases, there are no definite guidelines for treatment. Al-Salem *et al*^[23] reported that lacrimal sac massage can lead to resolution of tearing and avoid the need of surgery in four cases with combined ocular and fistula tearing. What's more, two of the four were found to have spontaneous occlusion of their fistulas when gently probing through the cutaneous opening^[23]. They presumed that diversion of tears from the fistula through the natural duct resulted in drying and occlusion of the tract, which might be similar to spontaneous canalicular occlusion resulting from punctal eversion. The most important reason to keep a fistula open is the outflow of fluids. If the obstruction of lacrimal drainage system was resolved, the fistula might be spontaneously closed. However, the result is limited to small case series, and further researches from larger samples are needed to verify this link.

Surgery generally is the treatment of choice, before which the patency of the nasolacrimal duct should be determined. Fistulectomy alone is preferable and much more effective

than simply cutting or cauterizing the ostium of the fistula in patients who had no evidence of nasolacrimal obstruction^[23-24]. This approach has shown excellent success rates ranging from 91% to 100%^[7,12]. The recurrence was attributed to incomplete excision of the epithelial lining of the fistula^[6,25]. During the closed approach of fistulectomy, adequate skin incision was performed to allow direct visualization of the anatomy and the fistula to be completely excised down to its base with closing of the proximal end. Sullivan *et al*^[7] believed that closed fistula excision is more difficult and potentially damaging to the common canaliculus or lacrimal sac than an open approach where the anatomical relationships can be properly defined. To ensure that the lacrimal outflow system is not compromised or injured, the canaliculi should be probed and the nasolacrimal system should be irrigated after fistula removal. Therefore, irrigation with methylene blue or other type of dye may be a good choice to check if the lacrimal drainage system is damaged after fistula removal, as mentioned in our report. Moreover, dacryoendoscopy enables the clear and direct observation of lacrimal passage without invasive manipulations like incisions to the skin or nasal mucosa^[26-27]. Heichel *et al*^[28] and Yamada *et al*^[29], respectively, employed the use of dacryoendoscopy diagnostically and therapeutically in congenital lacrimal fistulas. Thus, dacryoendoscopy can offer additional therapeutic option by detecting the origin of fistula accurately and simultaneously closing and excising the fistula^[30]. However, additional management for associated nasolacrimal duct obstruction should be performed, where required. For cases with associated nasolacrimal duct obstruction, lacrimal duct intubation or DCR should be performed in addition to fistula removal, depending on the type of abnormalities present^[23,31]. In our present case, the patient was performed with fistula excision without lacrimal duct intubation or DCR due to no nasolacrimal duct obstruction.

In this study, we described an uncommon congenital lacrimal fistula with recurrent fistulitis, which should be considered in differential diagnosis of acute dacryocystitis. Although the rate of lacrimal fistulitis is very rare, it still deserves detailed history taking and careful clinical investigation for definite diagnosis and correct management.

ACKNOWLEDGEMENTS

Authors' contributions: Xu PF and Zhong XC collected the clinical data of the case and wrote the manuscript under the instructions of Wu H and Ye J. Shi X contributed in revising the manuscript and tried to find a suitable magazine to publish the passage. Wu H was the major surgeon who conducted the surgery. All authors read and approved the manuscript.

Conflicts of Interest: Xu PF, None; Zhong XC, None; Shi X, None; Ye J, None; Wu H, None.

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