Case Report

Bilateral Congenital Lacrimal Fistula in Down Syndrome

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ABSTRACT

Congenital lacrimal fistulae are rare in Down syndrome and bilateral presentation is very unusual. It can be associated with nasolacrimal duct obstruction. We report a 3-year-old female with Down syndrome who presented with watering and discharge from both eyes and bilateral fistulous openings present inferonasal to the medial canthus. Upon examination, the lacrimal sac regurgitation test was positive on both sides. Our case report documents a distinctive case of bilateral congenital lacrimal fistulae in association with Down syndrome. It was managed successfully by primary fistulectomy and nasolacrimal duct probing.

Key words: Down Syndrome, Fistulectomy, Lacrimal Fistula, Nasolacrimal Duct

INTRODUCTION

Bilateral congenital lacrimal fistulae constitute a rare anomaly of the lacrimal system. In this, an epithelium-lined tract connects the skin to common canaliculus, lacrimal sac or nasolacrimal duct. It has been systemically associated with thalassemia, preauricular fistulae, hypospadias, Down syndrome and VACTERL (vertebral anomalies, anal atresia, cardiac malformations, tracheo-esophageal fistula, renal anomalies, and limb anomalies) in previous reports. In the following report we present a case of bilateral congenital lacrimal fistulae associated with Down syndrome and its surgical management.

CASE REPORT

A 3-year-old female presented with watering and mucoid discharge from both eyes and from small skin holes on both sides. She was a known case of Down’s syndrome with mental retardation and delayed developmental milestones with no family history of similar complaints. On examination she had telecanthus and had typical mongoloid facies with upward slant of lateral canthus. Bilateral fistulous openings were seen inferonasal to medial canthus. Lacrimal sac regurgitation test was positive on both sides.

Evaluation of the lacrimal system was performed under general anesthesia. The right eye had a direct fistulous tract 4.5 mm in length communicating with the lower canaliculus [Figure 1]. Two No. 1 Bowman’s probes, one from the puncta and the other from the fistula were simultaneously guided till they touched each other. In the left eye the fistulous tract was communicating with the lower portion of lacrimal sac [Figure 2]. On bilateral probing, soft stops were encountered at the beginning of nasolacrimal ducts which were overcome with minimal force. Patency of the nasolacrimal duct was confirmed by syringing and retrieval of fluorescein dye from the nose on suction. Both the tracts were excised partially by performing a fusiform incision around the fistulous ostium along the skin tension lines. The tract was held with a tooth forceps and was separated from the surrounding tissue with Vannas scissors and 3/4 was excised. The base of the fistula was cauterized and no regurgitation of fluid was noted on syringing. Surrounding tissue and skin was sutured with 6-0 vicryl suture. Histopathology of the excised tracts showed duct lined by stratified squamous epithelium.
At 4 months follow-up, the patient was symptom free and no fistulous openings were visible.

COMMENT

Congenital lacrimal fistulas are rare developmental anomalies of the nasolacrimal excretory system with an estimated incidence of one in 2,000 births.\(^5\) There is no sex or race predilection. Most of the fistulas are unilateral and typically located inferonasal to the medial canthal angle. The majority remain asymptomatic with patent nasolacrimal ducts.

Ours is a rare case of congenital bilateral lacrimal fistulae with nasolacrimal duct obstruction in a pediatric patient with Down syndrome. To our knowledge (recent PubMed search on Down syndrome), there is only one case with bilateral congenital lacrimal fistula reported by Keserü \textit{et al.}\(^6\) Sullivan \textit{et al.}\(^7\) reported five cases of Down’s syndrome, all of which were unilateral and managed conservatively or with fistulectomy. None of the patients reported in these previous publications\(^6,7\) had associated nasolacrimal duct obstruction. Nasolacrimal duct obstruction can be managed with probing and/or dacryocystorhinostomy depending on the age of the patient. Bilateral fistulous tract excision along with bilateral nasolacrimal duct probing was effective in our patient.

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