Isolated form of congenital bilateral lacrimal gland agenesis

Dear Editor,

Congenital lacrimal gland agenesis is a rare cause of dry eye syndrome in childhood.[1,2] Lacrimal gland agenesis may occur as an isolated condition or it may accompany salivary gland agenesis and atresia of lacrimal drainage system.[3,4] We present herein a seven-year-old boy with an isolated form of congenital bilateral absence of lacrimal glands.

A seven-year-old boy presented with a four-year history of foreign-body sensation and irritation in both eyes. The visual acuities were 20/20 in the right and 20/100 in the left eye. Slit-lamp examination showed conjunctival hyperemia, corneal punctate epithelial erosions, and mucous filaments in both eyes. Tear break-up time was 2 sec and Schirmer’s I test detected 2 mm of wetting in 5 min in each eye. He was otherwise systemically healthy.

Systemic diseases that can cause dry eye syndrome were eliminated by negative systemic evaluation including rheumatology consultation, chest X-ray, purified protein derivative (PPD), and serum angiotensin-converting enzyme titer. Therefore, we performed lacrimal gland biopsy through a superotemporal extraperiosteal approach. Histopathology showed no evidence of lacrimal gland tissue. Orbital magnetic resonance imaging (MRI) scan revealed absence of both lacrimal glands [Fig. 1].

The patient was subsequently treated with permanent occlusion of both puncti and topical artificial tears. After two years of follow-up, the patient is clinically comfortable with improved vision of 20/50 in the left and 20/20 vision in the right eye.

Lacrimal gland begins to develop in the lower eyelid at the end of the second month of the intrauterine life. Orbital and palpebral parts are formed in the fifth month, but the full differentiation occurs in three to four years after birth. Thus, any disarrangement in very early intrauterine life may cause lacrimal gland agenesis.

Dry eye syndrome is an uncommon diagnosis in children. This disease can be associated with systemic disorders such as Sjögren’s syndrome, Riley-Day syndrome, ectodermal dysplasia syndromes, hypovitaminosis A due to congenital small bowel atresia, and graft-versus-host disease following bone marrow transplantation.[5] Delayed diagnosis of any of these diseases can cause significant corneal problems and amblyopia. Detailed systemic workup is necessary to detect the specific cause of childhood dry eye syndrome. Patient history is especially important for the diagnosis. Photophobia, chronic eye redness and blepharospasm from early childhood should alert the clinician to search for lacrimal glands with appropriate imaging techniques. Very few previous articles have reported salivary gland absence in association with congenital lacrimal gland agenesis. Thus, this condition should be ruled out clinically. Our patient had normal saliva production clinically and the history did not reveal any symptom of dry mouth during the entire follow-up period. Additionally, MRI findings showed normal salivary gland architecture.

Isolated form of congenital lacrimal gland agenesis is a rare condition but should be considered in the differential diagnosis of childhood dry eye syndrome. In children with dry eye syndrome without any systemic findings, it should be emphasized that noninvasive measures such as MRI can be primarily undertaken for the possible diagnosis of lacrimal gland agenesis before performing any invasive procedure.

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References

1. Milunsky JM, Lee VW, Siegel BS, Milunsky A. Agenesis or hypoplasia of major salivary and lacrimal glands. Am J Med Genet 1990;37:371-4.
2. Kim SH, Hwang S, Kweon S, Kim TK, Oh J. Two cases of lacrimal gland agenesis in the same family--clinicoradiologic findings and management. Can J Ophthalmol 2005;40:502-5.
3. Ferreira AP, Gomez RS, Castro WH, Calixto NS, Silva RA, Aguiar MJ. Congenital absence of lacrimal puncta and salivary glands: Report of a Brazilian family and review. Am J Med Genet
3. In the case which underwent endophthalmitis, you have
1. There had been changes in the technique of AGV
2. Those cases which were not included in the study because
4. There are no definite data on failure rates because of
5. Mac Cord Medina F, Silvestre de Castro R, Leite SC, Rocha EM,
2. The success rate is indeed influenced by including or
3. The endophthalmitis case had developmental glaucoma
5. You are correct in pointing out that a control group was

References

4. Caccamise WC, Townes PL. Congenital absence of the lacrimal

5. Mac Cord Medina F, Silvestre de Castro R, Leite SC, Rocha EM,

6. Preoperative data previous to
5. There is no control group to show the comparison of your

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