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Editorial

Congenital Dacryocystocele and Management: Why do we Need a Nasal Endoscope?

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Congenital dacryocystocele (or dacryocele, amniocele, amniotocele) is a rather rare, cystic dilation of the lacrimal sac due to nasolacrimal duct obstruction, which becomes patent either spontaneously, or by massaging, or probing and irrigation of the lacrimal drainage system in the majority of the affected infants. [1-5] A dacryocele causing respiratory distress due to an intranasal cyst that blocks the airways may be life treathening and recognizing the intranasal cyst and its management via marsupialization by the ophthalmic surgeon is crucial [5-7]. I perform and advocate to intervene dacryoceles under nasal endoscopy, in order to avoid an incomplete recanalization in the presence of intranasal cists, also keeping in mind that, this condition occasionally involves both sides, even when the contralateral side appears to be symptomless and normal on external examination [8-10]. Nasal endoscopy, finding its role in the conventional lacrimal surgery routine at present, is a useful tool to decrease the rates of failure and reoperations in simple, complex and complicated congenital nasolacrimal duct obstructions (CNLDO) [11]. Intraorbital extension of dacryocystoceles may be seen but it is a rare complication which may rarely necessitate an early dacryocystorhinostomy [12].

Definition, signs, symptoms and diagnosis

A dacryocele generally appears as a grayish swelling, just below the medial canthal tendon seen at the time of birth or within the first few weeks of life [3-5]. A dacryocele occurs in newborns with CNLDO as a result of consequent obstruction of proximal opening of the enlarged lacrimal sac, where the Rosenmüller valve acts as a one-way valve, in addition to the incomplete canalization of the distal aperture of the lacrimal drainage apparatus at the level of Hasner's valve [2,7,12,13]. The functional obstruction at the proximal opening may also be caused by compression of the dilated lacrimal sac onto the internal common canaliculus and the canaliculi so that these short and fine structures plicate or collapse at the junction site with the sac [2,13,14]. Accumulation of fluid in the drainage system eventually dilates the lacrimal sac into a dacryocystocele, The distal obstruction is anatomical, where the proximal obstruction is functional. [2,5,15-18].

It is seen in 1 of 3,900 live births with higher female predilection up to 73% [2,4,5]. The unilaterality of dacryocystoceles in neonates is between 86 -100%, and some infants may have bilateral obstructions [4,5].

Most common symptoms of dacryocystoceles in the newborns are epiphora and difficulty in breathing which worsens during feeding. Elevated tear meniscus hight over 2 mm. s, and/or overflow of tears in the affected eye with a bluish-gray, firm, nontender mass located at or below the medial canthus denotes a sterile distension of lacrimal sac [2,5,7]. As the retained stagnant fluid becomes infected, dac-

ryocystitis takes place. Secondary infection may supervene within days or weeks if left untreated. In this episode the lacrimal sac and neighboring structures are inflamed. Secondary preseptal or orbital cellulitis may set in.

Dacriyocystitis is seen in 14% to 75% of infants with congenital dacryocystoceles and generally necessitates surgical intervention [4,8,9]. Intranasal cysts, cutaneous fistulae, eyelid and orbital abscesses are other complications [5,12]. Rarely, intraorbital enlargement of the dacryocystocele may cause proptosis and distortion of the globe. The increase in hydraulic pressure within the sac, most commonly expands anteriorly toward the eyelid where it is covered only by skin and orbicularis muscle. The second probable direction of sac distension is inferiorly through the nasolacrimal duct, resulting in an intranasal cystic expansion in a newborn. A "third probable direction" of expansion has been postulated to occur due to anatomical variations of the lacrimal sac fossa with less posterior resistance, resulting in cystic expansion of the sac posteriorly into the orbit. Maturing bones of nasolacrimal sac fossa and duct may prevent the sac from evaginating inside the nose, initially forcing an anterior, cutaneous route, with recurring episodes of spontaneous external drainage through a fistula formation. Increased resistance of the scarred eyelid skin, may cause posterior expanding of the sac into the orbit (Figure 1). Orbital cellulitis, meningitis and rarely with sepsis may follow [12].

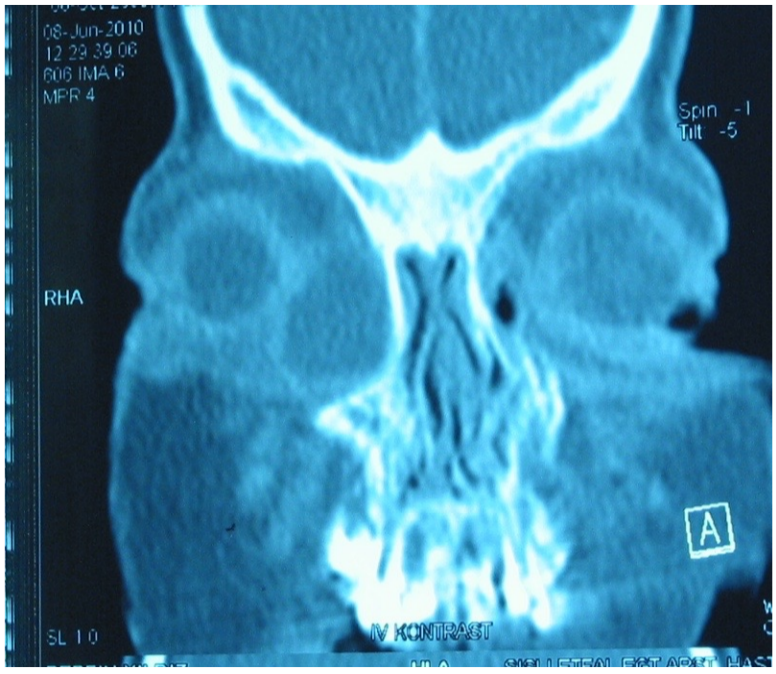

Figure 1: *Coronal CT imaging section of the infant with intraorbital extension of the dacryocystocele who did not respond to probing and marsupialization of intranasal cyst.*

Typical clinical presentation of congenital dacryoceles and the relatively short course prior to management minimize the need for imaging studies in the majority of infants [19]. However, ultrasound (US), computerized tomography (CT) scanning, magnetic resonance imaging (MRI) can be informative in where orbital invasion, with or without infection is suspected, and/or neoplasms or meningoencephaloceles are in differential diagnosis. The sonographic appearance of a dacrycystocele is a medial cystic mass with a fluid content communicating with the dilated nasolacrimal duct [19-23]. Contrast material in the lacrimal drainage system as in CT-DCG and MR-DCG give also provide more definitive results [17,19]. Gadolinium chelate drops instilled in cul-de-sac in MR-DCG avoids catheterization of the

lacrimal system gives details but require general anesthesia [24]. Dacryoendoscopy has been used described by Kakizaki., *et al*. to view the internal wall of a dacryocystocele, demonstrated injection and hemorrhages on the inner wall of the cyst with inflammatory debris. It aided in taking the biopsy and marsupialization of the cyst [25].

The differential diagnosis of medial canthal swellings include diverticulae of lacrimal drainage apparatus, supernumerary sacs, epidermoid and dermoid cysts, mucoceles of paranasal sinuses, anterior encephaloceles, vascular aneurisms, tumors impinging on the sac [26-29]. Encephaloceles or menigoceles can present as a faint pulsating proptosis and a mass typically above the medial canthal level. On CT scan the osseous defect can usually be demonstrated [27]. Sudoriferous cysts, the apocrine gland cysts of Mol, appear as a fluctuant swelling beneath the lower eyelid similar to dacryocystitis without inflammation. Aneurisms and neoplasms are solid firm masses diagnosed on using MRI scans, better with contrast enhancement [24,26].

Management of congenital dacryocystoceles

Congenital dacryocystoceles are conservatively addressed with warm compresses and topical ant biotherapy during the postnatal several weeks. Resolution rate with conservative therapy is reported from 17% to 80% which included massaging [4,5,11,16,17,30,31] Firm pressure on the dacryocystocele by an experienced ophthalmologist at the first visit, in order to direct the fluid content of the lacrimal sac inferiorly aims to break the membraneous tissue to open at the valve of Hasner by hydrolic pressure [5,30]. If the pressure overcomes the resistance at the inferior meatal soft tissues, the cyst decompresses, and the symtoms disappear.

If the condition persists after conservative measures are taken, lacrimal probing is generally the first step surgical intervention [5,16,31-33]. At this stage I recommend probing to be performed under nasal endoscopy whenever possible, with attention to the kinked canaliculi so that a false passage and canalicular trauma is avoided. In some cases the contents of the sac may regurgitate from the puncti if the canalicular kink is straightened. Probing should open the membraneous mucosa at the level of Hasner's valve and marsupialize the nasal cystic protrusion if present. Nasal endoscopy minimizes the possibility of a false mucosal opening and ensures the visualization of the inferior turbinate and the intranasal cist and its marsupialization. Urgent probing is recommended if dacryocystitis occurs. Successful probing under endoscopy can prevent the incidence of secondary dacryocystitis and cellulitis, which typically presents if treatment is delayed, or after an unsuccessful intervention, which may necessitate another secondary procedure under general anesthesia [11]. In Mansour., *et al.* s series of dacryocystoceles, probing without nasal endoscopy resulted in 78% successful resolution, where in 36 infants of 54 had dacryocystoceles complicated with dacryocystitis [4].

In our clinical practice, we prefer to perform lacrimal probing under general anesthesia and endoscopic visualization, as this allows the advantages of detailed visualization of the inferor turbinate anatomy and intranasal cysts, and enable precise manipulation of delicate nasolacrimal tissues, with a 98% success rate [Unpublished data by Kaynak., *et al.* The high success rate of intervention under general anesthesia and endoscopy is also attributed to be able to change the surgical plans intraoperatively, where additional pathologies are detected, at the same session to avoid a second intervention at a later setting. Silicone stenting of the lacrimal drainage system is done where a complex stenosis at the duct or canaliculi is encountered at the same session. The frequent coexistence of intranasal cysts caused by extension of dacryocystoceles into the inferior nasal cavity require marsupialization of the cysts under direct or endoscopic visualization. Recurrence with mucocele is reported by Raflo., *et al.* in an infant with a dacryocystocele and coexisting intranasal cyst, where marsupialization of the cyst wall was not done at the time of probing [6]. As recommended by Ali MJ., *et al.* simple marsupialization may not be adequate in these cases in whom the massive redundant mucosa would require additional superior and inferior relaxing cuts on the anterior and posterior flaps, so that they can be reflected like an open book [34]. These findings comply with Kashkouli's work who recommends to start the initial intervention under endoscopy with full range of surgical equipment and continue with the treatment which is adequate at the same session [11].

Another reason to intervene dacryoceles under nasal endocopy is probable bilaterality of intranasal cysts. A unilateral dacryocystocele at inspection does not exclude the possibility of bilateral intranasal cysts, therefore both nasal cavities should always be evaluated [35-38]. Two cases of unilateral CDCs treated with probing and cyst marsupialization developed contralateral dacryocele after the intervention at 6th and 10th days, respectively [39].

In another series, 4 of 22 patients developed a sequential CDC [10].

Intraorbital extension of congenital dacryoceles must be handled with care. Massaging is not recommended especially in long standing cases with dacryocystitis. Possibility of extereme bony tinning from chronic inflammation may potentially cause introduction of the sac contents into the orbit with manual pressure on the sac. Two iatrogenic cases, where massaging lead to dacryocytocele extension into the orbit with abbcess formation is reported presented by Bernardini., *et al.* and Baker and Allen [12,40]. CT or MRI imaging should be considered when intraorbital extension is suspected. In addition, probing must be performed with care to prevent a false entry to the orbit and secondary introduction of the infectious agents into the orbit [5,12]. In cases of orbital extension, transconjunctival marsupialization of the large dacryocystoceles followed by probing, is the first choice for surgery. DCR may also be done if probing and marsupialization of the cyst fails. In severe refractory, or untreated cases of recurrent dacryocystitis within a dacryocystocele, a lacrimal-cutaneous fistula can develop, that may lead to cicatricial changes or ectropion of the lower eyelid. A seven month old baby girl who presented to our hospital, with a fistula and had history of recurrent dacryocystitis attacks since birth fistulating several times that resulted in sicatricial lower eyelid ectropion and intraorbital extension of the dacryocystopyocele with the history of unknown procedure performed in another healthcare center. Under systemic antibiotic therapy, the patient underwent lacrimal probing and intranasal cyst marsupialization at the same surgical session. Dacryocystitis recurred, in a few weeks without fistulisation. A large saccal cyst extending to the orbit was detected in CT, in communication with the nasolacrimal duct (Figure 1). External DCR was performed at 7th month of age, with a nasojugal incision hidden in the preformed sicatricial folds with partial dacryocystectomy to reduce sac walls to anastomose proper sized saccal flaps to the nasal flaps. No recurrence of symptoms were noted Endonasal DCR approaches could also be used in this case. However, external DCR was preferred because of the large sac volume. During dacryocystorhinostomy, a partial dacryocystectomy to reduce the dilated sac walls was performed to allow anastomosis of more properly sized sac flaps to the nasal flaps [5,12].

Accumulation of fluid in the enlarged sac compartments may persist and cause dacryocystitis despite the drainage of some tears. Early imaging and prompt intervention should be done when signs of intraorbital extension of the dacryocystocele is observed. A patient with bilateral congenital punctal agenesis and CNLDO that leaded to formation of dacryocystoceles in years during childhood had undergone conjunctivo-dacryocystorhinostomy (C-DCR) performed by Song and coauthors. In rare cases, dacryocystoceles in the setting of significant congenital punctal agenesis may require conjunctivodacryocystorhinostomy (CDCR) [41]. Santo and coauthors treated 13 year old girl a giant dacryocystocele, in a patient with lacrimo-auriculo-dento-digital syndrome an association with alacrima, and agenesis of the lacrimal punctate, which started to enlarge at 6 years of age via transconjunctival orbitotomy to excise the dacryocystocele, and simultaneous right endonasal antrostomy [15].

The management of congenital dacryocystoceles differs according to the presence of infection extent of the cyst, and other anatomic findings or complications. The main purpose of management is to maintain establish the drainage of tears through the patent and functional lacrimal apparatus and to resolve the infection. The heterogenic properties of CNLDOs and the complications of dacryoceles such as dacryocystitis, intranasal cists and orbital extension require a wide spectrum of management options ranging from conservative therapy to C-DCR. Therefore, nasal endoscopy is recommended during probing at the first session with or without marsupialization of the sac to decrease the possibility of consequent interventions under general anesthesia and increase the success rate of management.

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