Congenital dacryocystocele: case report and treatment

Dacriocistocele congênita: relato de caso e conduta

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ABSTRACT

The dacryocystocele represents a rare congenital anomaly in the medial region of the orbit caused by distal obstruction (Hasner valve) and proximal (valve Rosenmüller) of the lacrimal system causing dilation of the lacrimal sac. Mucocele is called when the content is mucus and amniocele when the content is filled with amniótico fluid. The incidence is only 0.1% in children with nasolacrimal duct obstruction. It is commonly unilateral and more frequent in women with familial predisposition.

The diagnosis is made by clinical features: tense cystic lesion below the medial canthal tendon, blue-gray, pink or red color with epiphora since birth. However we can use image tests to diagnose this congenital anomaly such as tomography computerized, magnetic resonance and ultrasonography.

Keywords: *Lacrimal duct obstruction/diagnosis; Lacrimal duct obstruction/congenital; Lacrimal duct obstruction/therapy Lacrimal duct obstruction/ultrasonography; Epidermal cyst; Dermoid cyst; Case reports*

RESUMO

A dacriocistocele representa uma rara anomalia congênita da região medial da órbita, causada pela obstrução distal (ao nível da válvula de Hasner) e proximal (ao nível da válvula de Rosenmüller) da via lacrimal, com subsequente dilatação do saco lacrimal. Recebe o nome de mucocele, quando seu conteúdo representa muco, ou amniocele, quando o seu conteúdo é preenchido por fluido amniótico. Acomete somente 0.1% das crianças, com obstrução do ducto lácrimonasal, sendo comumente unilateral e mais frequente no sexo feminino e com predisposição familiar.

O diagnóstico é realizado pelas características clínicas: lesão cística tensa, abaixo do tendão cantal medial, de coloração azul-acinzentada, rósea ou vermelha acompanhada por epífora desde o nascimento. No entanto podemos utilizar exames de imagem para diagnosticar esta anomalia congênita tais como: tomografia computadorizada, ressonância magnética e ultrassonografia.

Descritores: Obstrução dos ductos lacrimais/congênito; Obstrução dos ductos lacrimais/diagnóstico; Obstrução dos ductos lacrimais/terapia; Obstrução dos ductos lacrimais/ultrassonografia; Cisto epidérmico; Cisto dermóide; Relatos de casos

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Introduction

Dacryocystocele is a rare congenital anomaly of the medial region of the orbit[1] caused by distal (at the level of the valve of Hasner) and proximal (at the level of the valve of Rosenmüller) obstruction of the lacrimal system, with subsequent dilation of the lacrimal sac. It is called mucocele when its contents are mucus and amniocele when it is filled with amniotic fluid. The condition is present in only 0.1% of children with nasolacrimal duct obstruction; it is usually unilateral, it has a familial predisposition, and it is more frequent in females.[1-3]

Diagnosis is based on clinical features,[4,5] which include a tense cystic lesion below the medial canthal tendon with a blue-grayish, pink, or red colour accompanied by epiphora since birth. However, imaging studies such as computed tomography, magnetic resonance imaging, and ultrasound imaging can be used to diagnose the condition.[6-9]

The cyst is clearly visible on prenatal ultrasound scans[10,11] as an anechoic collection of fluid in the medial edge of the orbital cavity that does not communicate with the skull or the eye globe. It is observed in the last trimester of pregnancy. Spontaneous resolution occurs in 50% of cases before birth. Postnatal complications include epiphora, dacryocystitis, conjunctivitis, cellulitis, and respiratory distress.[12-14] The differential diagnosis includes anterior or posterior meningoencephalocele, haemangioma, epidermoid cyst, dermoid cyst, nasal glioma, and lymphangioma.

There is no consensus on the treatment of congenital dacryocystocele.[15,16] It can be initially treated with Crigler massage,[17] topical and systemic antibiotics, and warm compresses. Because dacryocystocele is highly susceptible to infection, antibiotic prophylaxis is indicated. If conservative treatment is ineffective after a few weeks, lacrimal probing is conducted. If initial probing is ineffective, silicone intubation, balloon dacryocystoplasty, or surgical marsupialisation of the nasolacrimal cyst should be performed.

Ophthalmologists should be aware of the symptoms of nasal obstruction leading to respiratory distress, which can be life-threatening. In such cases, endoscopic marsupialisation of the nasolacrimal cyst is recommended. In cases of dacryocystocele progressing to acute dacryocystitis, systemic antibiotic therapy is indicated to prevent serious complications such as meningitis, brain abscess, and sepsis.

Case Report

White female child, born and raised in São Paulo, seen at the Lacrimal Apparatus Unit of UNIFESP from the 7th day of life with congenital dacryocystocele on the right side (Figure 1). Ophthalmic examination (inspection and palpation) showed a blue-grayish tense cystic lesion below the medial canthal tendon accompanied by epiphora since birth. Biomicroscopy showed no mucopurulent discharge or bulbar conjunctival hyperaemia in the right eye; the Milder test was intensely positive. The fluorescein appearance test in the oropharynx was negative. Eye ultrasound (Figure 2) found a round, well-defined preseptal orbital lesion in the medial canthus with a hypoechoic content. The lesion measured 3.1 × 3.6 × 2.9 mm (depth × height × width). There was no evidence of posterior extension or communication with the eye globe. The features of the lesion were compatible with dacryocystocele.

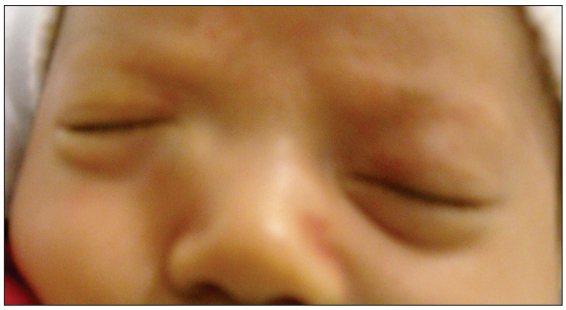

Figure 1. Child with congenital dacryocystocele on the right side.

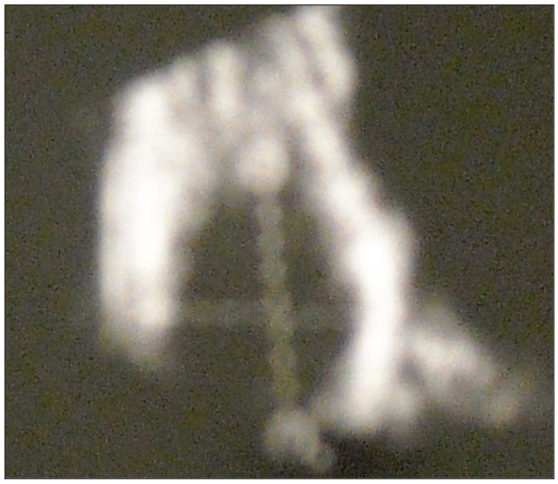

Figure 2. Right eye ultrasound.

The child received conservative treatment with Crigler massage, warm compresses, and systemic antibiotics (cephalexin suspension 250 mg/5 ml, 2 ml every 6 hours for 7 days). The condition resolved after 4 weeks.

Discussion

Dacryocystocele is caused by an accumulation of amniotic fluid in the lacrimal sac (amniocele), which becomes dilated and clearly visible on prenatal ultrasound scans in the last trimester of pregnancy. The lesion appears as an anechoic, fluid-filled cystic area on the internal edge the orbital cavity.[18] However, the amniotic fluid by itself is not able to dilate the lacrimal sac, especially when the bulging occurs days after birth. The term mucocele is used when the lacrimal sac is filled with mucus. Dilation of the lacrimal sac is more likely caused by a combination of mucus, amniotic fluid, lacrimal fluid, and bacterial proliferation, but its contents are often sterile.[19]

In newborns, dacryocystocele is diagnosed clinically. Diagnostic tests such as transillumination, ultrasound, computed tomography, magnetic resonance imaging, dacryocystography, and rhinoscopy may be required to clarify the diagnosis. MRI can help characterise the contents of the lesion, while TC is used to visualise bone anomalies affecting the nasolacrimal duct.[20]

Prenatal visualisation of dacryocystocele by Doppler ultrasound helps identify associated malformations.[21]

Cystic extension of dacryocystocele into the nasal cavity is not uncommon and can cause respiratory difficulty during sleep and nursing in bilateral cases. Therefore, all children with

congenital dacryocystocele should undergo rhinoscopy performed by an otolaryngologist to exclude an associated intranasal cyst.[22]

There is no consensus on the treatment of dacryocystocele. Conservative treatment is initially recommended (lacrimal sac massage, topical and/or systemic antibiotic therapy) during the first months of life.[23-25] Lacrimal probing is recommended when conservative treatment is ineffective or when the child presents severe infection or respiratory distress.[26,27] According to the literature, early probing can prevent infections and sequelae (tissue distortion in the inner canthus, induction of corneal astigmatism, and anisometropic amblyopia).[28]

In the presence of an intranasal cyst, the child can be referred to an otolaryngologist for cyst marsupialisation.[29,30]

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