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Congenital bilateral dacryocystocoele: A neonatal emergency

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ABSTRACT

INTRODUCTION AND IMPORTANCE: Bilateral congenital dacryocystocoele with intranasal extension is very rare and may lead to mild to severe respiratory distress, depending on the degree of obstruction, in an otherwise healthy newborn. If severe, an urgent surgical intervention may be life saving.

Our aim is to alert paediatric surgeons to this rare condition. Early detection and early treatment with a minimally invasive surgical procedure may be life saving and prevent severe sequelae due to respiratory distress.

CASE PRESENTATION: We present a healthy newborn girl who was admitted to neonatal intensive care with progressive respiratory distress. After a full work-up, she was diagnosed with bilateral dacryocystocoele with intranasal extension and complete obstruction of the anterior nasal cavity. Emergency bilateral endoscopic marsupialization of the cysts with probing of the nasolacrimal duct was performed. The girl recovered without sequelae.

CLINICAL DISCUSSION: In congenital dacryocystocoele, coexistent obstruction of the valve of Rosenmüller and the valve of Hasner is observed. Subsequent intranasal protrusion with obstruction of the inferior nasal cavity may occur. The incidence is unknown, but if bilateral, it is very rare. In most cases, a dacryocystocoele is uncomplicated and may be treated conservatively. However, if intranasal protrusion occurs, and especially if the involvement is bilateral, it is a surgical emergency.

CONCLUSION: Early diagnosis with subsequent minimally invasive surgical treatment of bilateral congenital dacryocystocoele with intranasal protrusion may prevent serious complications due to respiratory distress.

1. Introduction

Bilateral dacryocystocoele with intranasal extension is a rare variant of a congenital lacrimal duct stenosis. In typical cases, a neonate who started well, presents progressive respiratory distress. An urgent, but minimally invasive surgical procedure is needed to prevent serious neurological complications, as a consequence of respiratory distress. Neurological complications may vary from mild to serious encephalopathy, ranging from attention deficits to serious psychomotor retardation and even death, depending on the rate of respiratory distress. If performed by trained surgeons, the procedure is known to be easy and without any complications [1–5].

In this article we describe the case of a newborn infant who was admitted to the neonatal intensive care unit of an academic center with increasing respiratory distress due to congenital bilateral dacryocystocoele.

This work has been reported in line with the SCARE criteria [6].

2. Presentation of case

A female Caucasian – Hispanic neonate was admitted to the department of neonatal intensive care of a tertiary center with increasing respiratory distress. The prenatal period had been uneventful and she was born at term at 39 weeks without complications. Family and psychosocial history were unremarkable. On the second day she presented obstructive respiration with tachypnoea and need for oxygen (nasal cannula 2 L/21 %). Increasing oxygen need (4 L/21–28 %) was reported in the next days with regular episodes of desaturation up to 40 %. Clinical examination revealed severe bilateral nasal obstruction, but no other anomalies. Following nasal decongestion with oxymetazoline 0.1 mg/mL, nasal endoscopy was performed which revealed a bilateral intranasal mass just below the inferior conchae (Fig. 1). Complementary facial CT confirmed the diagnosis of bilateral dacryocystocoele with intranasal extension resulting in obstruction of the total anterior nasal cavity (Figs. 2 and 3).

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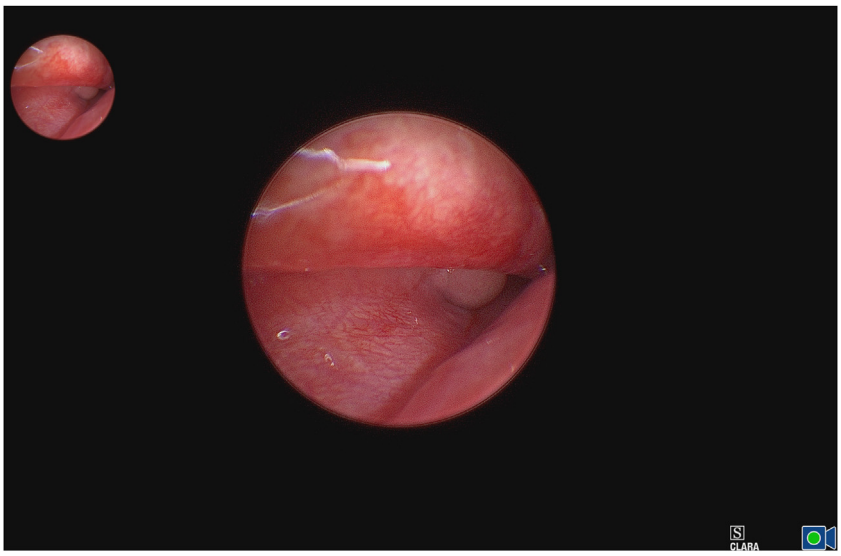

Fig. 1. Peroperative nasal endoscopy: protrusion of dacryocystocoele with medialization of the inferior concha and extension in the inferior nasal passage.

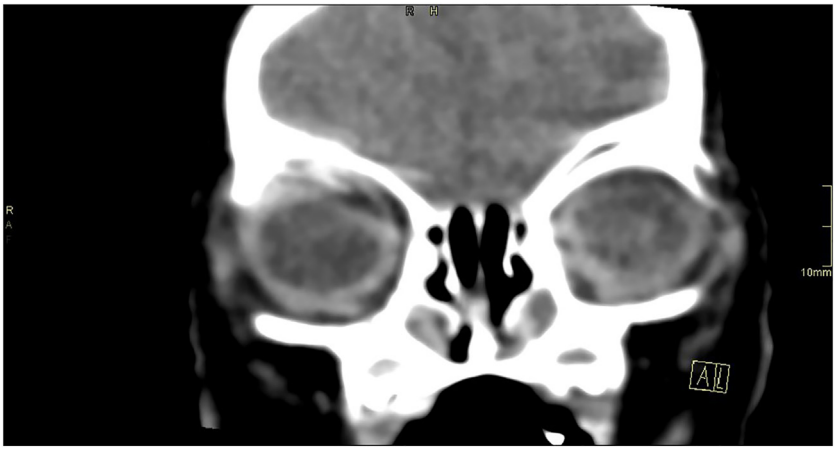

Fig. 2. Facial CT: coronal image: cystic extension of the mucocoele below the inferior concha with almost total obstruction of the anterior nasal cavity.

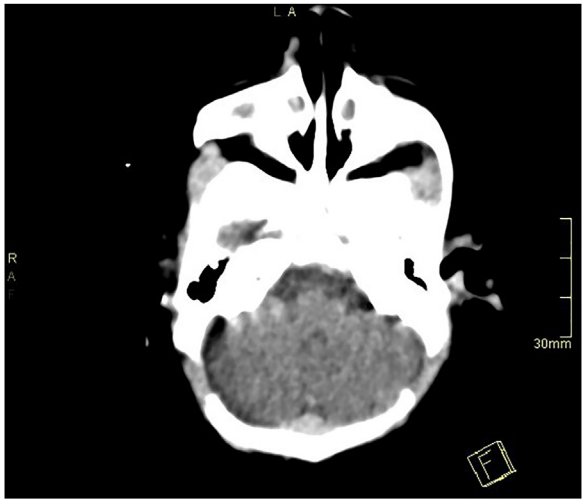

Fig. 3. Facial CT: axial image: bilateral cystic dilation of the nasolacrimal duct.

On day 7 bilateral exploration with marsupialization of the cysts was performed in order to restore the normal physiological function. This procedure was performed under general anaesthesia in an operating theater by a trained Ear Nose Throat (ENT) surgeon (subspecialized in among other things dacryocystorhinostomy surgery) and an oculoplastic surgeon, both active in a tertiary center for many years.

Oxymetazoline 0.1 mg/mL was instilled nasally preoperatively to obtain sufficient nasal decongestion to allow good visualization. The cystocoele was opened in the region of the lower nasal passage with a paediatric 3 mm microdebrider. The lacrimal duct was then dilated with a Muldoon lacrimal dilator and probed with a Bowman lacrimal probe, the duct was opened with a microhook and mucus was drained. No stent was placed.

At the end of the operation tobramycin/dexamethasone ointment was instilled in both eyes to prevent superinfection and postinflammatory swelling of the lacrimal canal. Postoperative treatment with tobramycin/dexamethasone drops 4 times a day was started with tapering off by 1 drop per week. The oxygen was continued (3 L/22 %) during 2 days after surgery until sufficient recovery was observed. No problems with therapy adherence were noted and the postoperative treatment was well tolerated during hospitalization.

Postoperative recovery was uneventful and on day 9 postpartum adjuvant treatment with oxygen was discontinued without problems. On day 11 the girl left neonatal intensive care unit in good condition.

After two weeks, an ambulant combined ophthalmological and ENT follow-up consultation was unremarkable. No further treatment and follow-up were necessary because no more complications were to be expected.

3. Discussion

3.1. Pathophysiology

A dacryocystocoele is a dilation of the lacrimal sac that occurs with obstruction of the proximal and distal parts of the lacrimal drainage system. In congenital forms, a dacryocystocoele is generally formed by coexistent obstruction of the proximal valve of Rosenmüller and the distal valve of Hasner. In the presence of swelling due to mucus in the coele, intranasal protrusion can occur, resulting in obstruction of the inferior nasal passage. In turn, this may cause severe respiratory distress, certainly if it is bilateral [1].

Obstruction of the nasolacrimal duct resulting in a dacryocystocoele is rare. The incidence is unknown, but it is rare, certainly if it is bilateral. To our knowledge, less than 25 cases with bilateral intranasal extension have been reported so far [1]. Approximately 14 % of congenital dacryocystocoeles are bilateral [1,3].

3.2. Diagnosis

Bilateral dacryocystocoeles usually present in the later perinatal period with recurrent episodes of desaturation due to variable obstruction of the anterior nasal cavity. Since neonates are obligatory nasal breathers during the first 2–6 months, nasal obstruction can result in severe respiratory distress [1]. Depending on the degree of respiratory distress, there may be need for supportive oxygen therapy or even an acute need for intubation in extreme cases [1].

This is in contrast with unilateral dacryocystocoeles, which often only present later in the postneonatal period, in combination with typical feeding problems and sleep disorders. In this case, the normal contralateral nasal passage is sufficient for breathing in the early neonatal period [1].

On examination a dacryocystocoele, if well filled with mucus, is visible as a local bluish swelling, inferior to the medial canthus of the eye, with epiphora and mucopurulent secretions [2]. Remarkably, this characteristic was absent in our early neonatal history.

If a neonate presents with secondary respiratory distress due to nasal obstruction after a good start, clinical examination should differentiate between the following aetiologies: nasal congestion due to mucus swelling, intranasal masses (including haemangioma/lymphangioma, dermoid cysts and (meningo-)encephalocoeles) and nasal anatomic anomalies [1,3–5].

If a dacryocystocoele with intranasal extension is suspected, ophthalmologic and ENT examination, and complementary bilateral nasal endoscopy are necessary [2]. If the examination and endoscopy are inconclusive, a CT scan is useful to confirm the clinical diagnosis [4]. CT reveals a typical triad of cystic dilation of the lacrimal sac, dilation of the nasolacrimal duct and intranasal cystic mass [2].

3.3. Treatment

In the event of an uncomplicated dacryocystocoele, initial treatment is conservative with warm compresses and massage. In 90 % of cases spontaneous resolution occurs by the age of 1 year [2].

Dacryocystocoeles with intranasal extension require surgical treatment and if present bilaterally, this constitutes an emergency. Three surgical options have been described:

- Blind probing of the nasolacrimal duct. This results in perforation of Rosenmüller's valve and proximal decompression of the nasolacrimal system [1,2].
- Nasal endoscopy with marsupialization of the intranasal cyst. This results in decompression of the system distally and permits drainage of the mucopurulent secretions [1,2].
- Combined endoscopic marsupialization and probing of the nasolacrimal duct. This results in decompression of the system proximally and distally and subsequent recovery of the normal physiological function. At present this is the treatment of choice for the prevention of recurrence [1,2,4].

These procedures are minimally invasive and very easy if performed by a trained ENT endoscopist together with an oculoplastic surgeon (with special interest in orbit/lacrimal duct).

In our case, the choice was emergency combined treatment with endoscopic marsupialization and probing of the nasolacrimal duct. Clinical recovery was rapid and satisfactory without sequelae. It is extremely important to perform surgery in the early stages in order to avoid complications due to respiratory distress.

4. Conclusion

If a neonate presents with respiratory distress, bilateral dacryocystocoele with intranasal extension should always be considered in the differential diagnosis. Presentation depends on the degree of nasal obstruction and can vary from intermittent respiratory distress to severe respiratory distress with need for urgent intubation. CT is useful to confirm the diagnosis if nasal endoscopy is inconclusive. Prompt diagnosis and surgery can avoid severe sequelae due to respiratory distress.

Declaration of Competing Interest

The authors report no declarations of interest.

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Ethical approval

No need for ethical approval. This case report highlights a known but very rare pathology, no new technique or treatment was used.

Consent

Written informed consent was obtained from the patient's parents (neonate) for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

Julie Imschoot: data collection, drafting manuscript, discussion.
Wouter Bauters: imaging, revision manuscript.
Thibault Van Zele: surgeon, revision manuscript.
Virginie Ninclaus: surgeon, revision manuscript.

Registration of research studies

Not applicable.

Guarantor

Julie Imschoot is appointed as the guarantor of this case report. Julie Imschoot accepts full responsibility for the work and/or the conduct of the study, had access to the data, and controlled the decision to publish.

Provenance and peer review

Not commissioned, externally peer-reviewed.

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