European Journal of Ophthalmology / Vol. 15 no. 4, 2005 / pp. 500-503

SHORT COMMUNICATION

Cerebrospinal fluid leakage during dacryocystorhinostomy in a patient with meningoencephalocele

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PURPOSE. To report a rare case of cerebrospinal fluid leakage during dacryocystorhinostomy in a patient with Möbius syndrome and meningoencephalocele and to explain our experience in the management of this complication.

METHODS. A 9-year-old girl with a history of surgically repaired fronto-ethmoidal meningoencephalocele and bilateral canthopexy was diagnosed with Möbius syndrome and underwent sequential bilateral dacryocystorhinostomy and silicone intubation because of dysgenesis of the lacrimal drainage pathway.

RESULTS. Both dacryocystorhinostomies resulted in cerebrospinal fluid leakage, which spontaneously ceased with conservative management.

CONCLUSIONS. Dacryocystorhinostomy surgery in patients with a history of trauma or surgery or congenital defects around the base of the nose or lacrimal system may rarely result in cerebrospinal fluid leakage; thus an ophthalmologist should be familiar with its management. (Eur J Ophthalmol 2005; 15: 500-3)

KEY WORDS. Dacryocystorhinostomy, Meningoencephalocele, Cerebrospinal fluid leakage, Möbius syndrome

Accepted: December 19, 2004

INTRODUCTION

Cerebrospinal fluid (CSF) rhinorrhea is often secondary to trauma or surgery involving the skull base due to leakage of CSF from the subarachnoid space into the paranasal sinuses or nose. Occasionally CSF rhinorrhea can occur spontaneously, the etiology of which is not well understood but hypotheses include congenital defects of the cribriform plate, bone erosion by CSF pressure waves, and focal atrophy of the olfactory nerve (1). Several cadaver studies of dacryocystorhinostomy (DCR) have shown that the osteotomy site is close to the cribriform plate and direct injury of the cribriform plate may occur (2, 3). However, in a similar cadaver study it was found that the average distance between the osteotomy site and the cribriform plate is 25 mm, and CSF leakage during DCR is not due to direct injury to the cribriform plate; however, torsional and tractional movements during osteotomy may extend the fracture into the cribriform plate and cause CSF leakage (4).

CSF leakage is a rare complication of DCR and orbital surgery, which is often mild with spontaneous remission before it can be diagnosed (2, 5, 6.) Rarely, intraoperative CSF leakage may be evident and since a neurosurgeon is often not available in the operating room, an ophthalmologist should be familiar with its management.

We report a rare case of CSF leakage during DCR in a patient with Möbius syndrome and frontoethmoidal meningoencephalocele in addition to our experience in the management of this condition.

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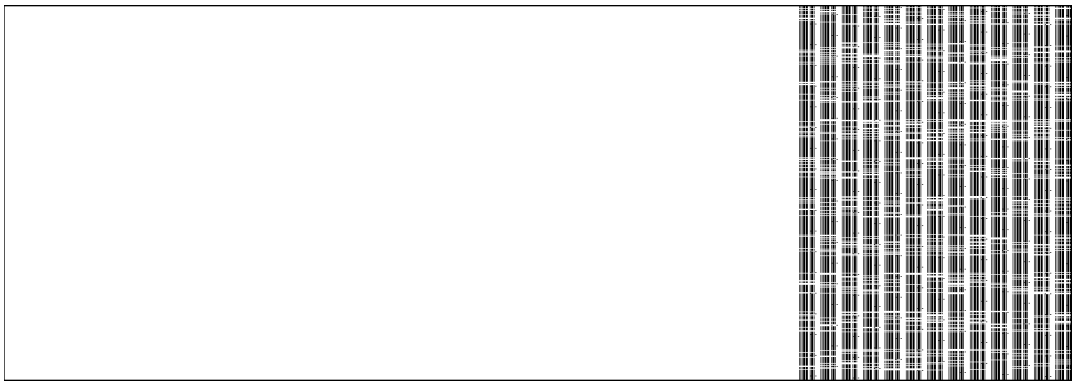

Fig. 1 - Preoperative appearance. A medium-sized mass is visible on the nasal bridge, more prominent on the right side.

Case report

A 9-year-old girl presented to our clinic complaining of bilateral tearing and inability to close the eyelids. Best-corrected visual acuity was 20/40 in the right eye and counting fingers at 50 cm in the left. Other findings were 45 Δ left esotropia, bilateral cranial nerves VI and VII palsy, lagophthalmos, and adherent leukoma of the left eye due to previous corneal perforation secondary to exposure keratopathy (Fig. 1). The patient had a history of frontal craniotomy by a neurosurgeon when she was 2 years old. The patient also had a history of bilateral medial canthopexy with miniplate by a plastic surgeon.

Radiography revealed the location of the residual cribriform plates to be lower than normal (Fig. 2). Computed tomography (CT) scan showed a soft tissue mass medial to the medial rectus muscle with pressure effect on the lateral wall of the right ethmoidal sinus and nose in association with a horizontal defect of both nasal bones up to the left medial orbital wall (Fig. 3). Also, a large defect involving both nasal bones and a smaller defect in the bony orbital roof were observed on three-dimensional CT scan (Fig. 4).

Cytogenetic evaluation for possible genetic disorders was reported as a mosaic of 46,XX/47,XX (+Mar) (Mar: an extra acrocentric [18 P-like] chromosome), with ratio of 4/1.

The diagnosis was Möbius syndrome, and to help reduce lagophthalmos and improve the lacrimal pump, she underwent bilateral sling surgery of the lower lids with fascia lata, which resulted in reduction of the lagophthalmos but persistence of epiphora.

One year later, she underwent DCR and silicone intubation on the left side and mild CSF leakage was observed during the surgery, which ceased spontaneously after wound closure. Postoperatively, epiphora of the left eye decreased and improved completely after removal of the silicone tube.

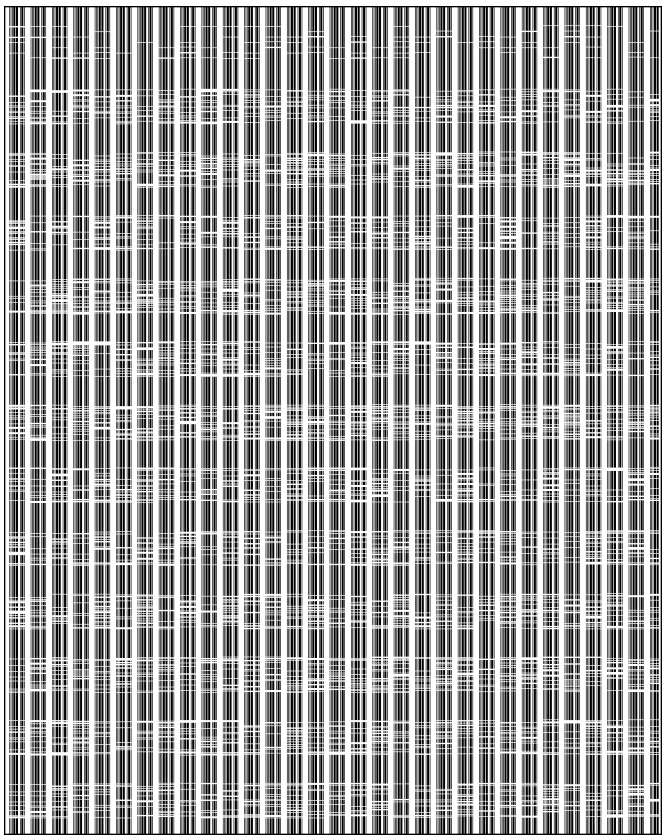

Fig. 2 - Preoperative plain radiography shows infradisplaced cribriform plates (arrow).

At age 12, obstruction in the right canaliculi was confirmed by irrigation test and scintigraphy. The patient underwent DCR and silicone intubation on the right side. We tried to perform the surgery under supervision by a neurosurgeon, but this could not be arranged. Intraoperatively, after skin incision and during dissection of the orbicularis muscle, a large cystic mass was observed in the inferonasal part of the right orbit and some clear fluid leaked into the wound, but the exact site of the leakage

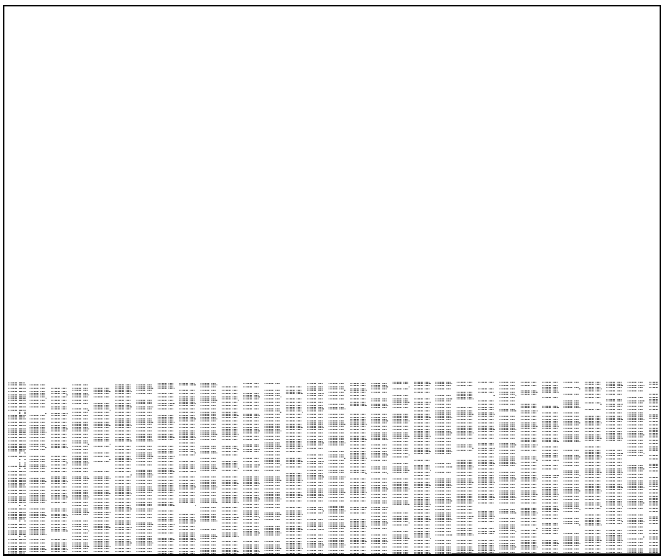

Fig. 3 - Axial computed tomography scan showing the meningoencephalocele on the nasal bridge with extension to the medial space of the right orbit.

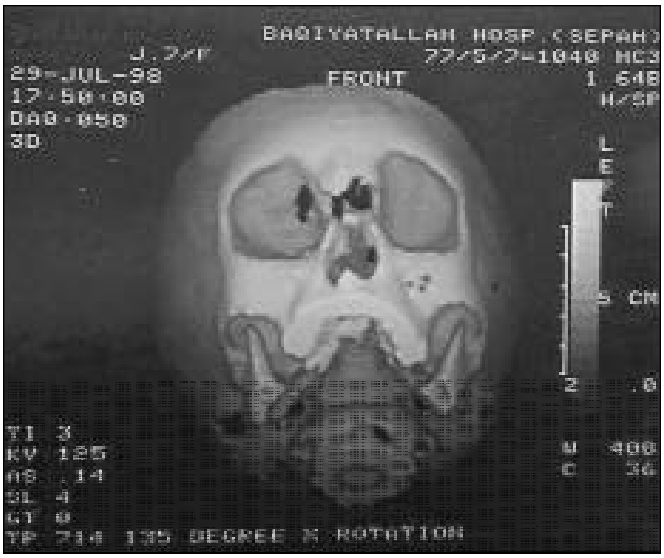

Fig. 4 - Three-dimensional computed tomography scan of bony structures of the skull showing defects on the nasal bridge and right orbital roof.

could not be determined. Surgery was continued while keeping the cyst retracted from the site of surgery. The fibrosed lacrimal sac was anatomically indistinct beneath the cystic mass, and its dissection from the bone had a high risk of cyst perforation. Therefore osteotomy was performed anterior to the lacrimal sac and slightly lower than the level of the previous canthopexy. Both the superior and inferior canaliculi were open only at their proximal 3 to 4 mm, thus using a lacrimal probe a canal was forcefully made along each canaliculus through the fibrosed tissues above the mass toward the osteotomy. Then the silicone tube was passed through each canaliculus, the artificially produced canal, and the osteotomy into the nose. At the conclusion, the orbicularis muscle and skin were sutured separately. Leakage of the clear fluid into the wound was continuously observed during surgery, which persisted as rhinorrhea without any wound leakage even after wound closure. Comparison of glucose concentration in the leaking fluid with simultaneous blood glucose confirmed the leaking fluid to be CSF.

According to infectious disease consultation, intravenous vancomycin (15 mg/kg) and ceftriaxone (50 mg/kg) were started prophylactically at the end of the surgery. The patient was instructed to remain in a semi-sitting position based on the recommendation of a neurosurgeon. On the first night after surgery, the patient had a fever of 38 ºC, which increased to 38.5 ºC the next day associated with nausea and headache while the CSF rhinorrhea continued. Close monitoring with supervision of a neurosurgeon continued. Rhinorrhea ceased spontaneously on the third day while fever, nausea, and headache continued for 1 week. Epiphora decreased during the second week and improved completely after 3 weeks. At the present time, 3 years after the last operation, she has no complaint of epiphora or new neurologic deficit and is a candidate for strabismus surgery.

DISCUSSION

In DCR surgery, it is important that the surgeon be familiar with the anatomy of the area, normal variations, and changes that may occur with special disorders or previous operations to prevent complications. An ordinary DCR requires a bony nasal window of about 20 mm in length and 15 mm in height with or without removal of the anterior arm of the medial canthal ligament. While performing DCR, the surgeon should consider the following points (2, 3, 5):

1. The superior border of the bony window and the inferior surface of the cribriform plate are usually very close to each other.
2. Occasionally the floor of the frontal sinus is also near the superior border of the bony window.
3. The posterosuperior part of the bony window is closest to the cribriform plate.

4. As age increases, ethmoidal air cells enlarge and cause thinning of the bone on the sides of the cribriform plate. Also, the frontal sinus enlarges and covers a larger area superior to the bony window.

Torsional movements during osteotomy, enlargement of the bony window at its posterosuperior border, younger age, and dissection of the medial canthal tendon are associated with an increased risk of CSF leakage in DCR surgery (2-5).

In a report of two cases with CSF leakage after DCR surgery, one of them had no intraoperative problem, but on the day after surgery, the patient developed headache and serosanguineous rhinorrhea without any sign of meningitis; the leak ceased spontaneously after 3 days with conservative treatment. In the second case, serosanguineous fluid leaked intraoperatively from an undetermined site during bilateral DCR. The operation continued, but postoperatively the patient had severe headache and serosanguineous rhinorrhea without fever, and the leak spontaneously ceased after 5 days (2). However, in our case, the anatomy of bones and soft tissues in the surgical area was abnormal. Obstruction of the lacrimal system was confirmed by irrigation test, probing, and scintigraphy. By ordinary CT scan and three-dimensional CT scan an adequate anatomic image of bones and soft tissues was obtained. If anatomic imaging of the soft tissues had not been adequate, MRI could have been helpful.

As we knew the abnormal anatomy of the surgical area, we tried to avoid severance of the meningocele, but unfortunately it was injured and serosanguineous fluid was observed in the wound, which was definitely CSF. Our previous knowledge of the surgical anatomy of the area was very important and helpful to keep off the meningocele while continuing surgery. Because the defect in the nasal bones had already been repaired by a miniplate and canthopexy had been performed against it, osteotomy was performed slightly lower and anterior to the miniplate to avoid injury to them.

Another point of the operation was the passage of silicone tube through the fibrosed tissue instead of the lacrimal sac because the lacrimal sac was an anatomically indistinct fibrosed tissue adherent to the meningocele area, making flap preparation impossible. Results of this procedure showed that it can be successful.

In conclusion, during DCR in a patient with a history of trauma or surgery around the nasal bones or lacrimal sac, or even in an anatomically normal person, CSF may rarely leak. When this occurs, the ophthalmologist should call a neurosurgeon if available; otherwise he or she should repair the defect of the dura as much as possible taking care to stay in a familiar area. CSF leakage in DCR is usually mild and ceases spontaneously, but its significance is the potential development of fatal purulent meningitis via ascending infection through the dural fistula. Prophylactic antibiotics should be started intraoperatively and postoperative consult with a neurosurgeon is mandatory.

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