
INTERMITTENT PALPEBRAL OEDEMA FOLLOWING SURGERY FOR NASOFRONTAL ENCEPHALOCELE

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ABSTRACT

Purpose: To report a case of intermittent palpebral oedema secondary to a cerebrospinal fluid (CSF) leakage, following encephalocele surgery.

Clinical case: A 9-year old girl consulted with a history of intermittent swelling of the left upper and lower eyelid. The symptoms started at the age of one. As a relevant fact in her medical history, a neurosurgical repair of a nasofrontal encephalocele at the age of 9 months was withheld. Elaborate imaging studies demonstrated a leakage of cerebrospinal fluid (CSF) into the preseptal periorbital soft tissues, originating from a bony defect in the medial orbital wall. The patient was treated conservatively.

Conclusion: A CSF leakage in the orbital or periorbital region is a rare finding. In the literature, similar defects have been reported after trauma or associated with neoplasia. To our knowledge, this is the first case of periorbital CSF leakage after encephalocele-surgery reported in the literature.

KEY WORDS:

Cerebrospinal fluid, encephalocele, leakage, preseptal, periorbital, palpebral oedema.

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INTRODUCTION

Leakage of cerebrospinal fluid (CSF) in the orbital or periorbital structures is a rare finding. We describe a case of CSF leakage in the periorbital soft tissues, presenting as an intermittent palpebral oedema, originating from a fistula after encephalocele surgery.

CLINICAL CASE

A 9-year old girl consulted our office with a history of intermittent swelling of the left upper and lower eyelid. The symptoms started at the age of one. Oedema typically occurred in the morning, especially after sleeping on the left side, and disappeared within one hour after being awake. There were no signs of inflammation, hyperthermia, pain or other complaints. As a relevant fact in her medical history, a neurosurgical repair of a nasofrontal encephalocele at the age of 9 months was withheld. The intermittent palpebral swelling started insidiously after surgery and became more frequent over time. The visual acuity was 9/10 in both



Fig 1: palpebral swelling of the left eye on a picture that the mother took in the morning.

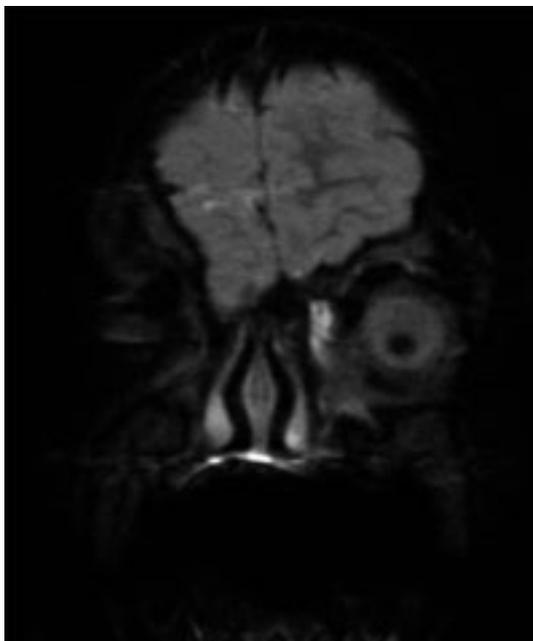


Fig 2: the late phase of the MRI cysternography, showing a leakage of CSF in the nasal periorbital soft tissues of the left eye.



Fig 3: CT-scan showing a bony defect anterior in the medial wall of the left orbit, situated between the frontal and maxillary bone.

eyes after treatment for refractive amblyopia of the left eye. The cyclopegic refractive measurements were S-2.00 for the right eye and S-3.25 C-1.75×158° for the left eye. Biomicroscopy of the anterior segment, fundus evaluation and ocular motility testing revealed no pathological finding. There were no indications of exophthalmia, pulsations or swelling after valsalva manoeuvre. Several pictures demonstrate the left palpebral oedema and its diffuse character (Fig 1).

The neurological and general physical work-up were unremarkable. Magnetic resonance imaging (MRI) of the orbit revealed a thickening of the subcutaneous fat in the nasal part of the left orbital region. After consulting the neurosurgical department, a MRI cisternography after intrathecal injection of Gadolinium was performed. For this examination, 0.2 mL of 0.5 mol/L gadobenate dimeglumine (MultiHance, Bracco) was diluted in 5 mL of 0.9% saline solution and injected directly into the lumbar subarachnoidal space. In the late phase images, a leakage of intrathecal contrast into the anterior nasal part of the periorbital soft tissues was detected (Fig 2). A 3D computed tomography (CT) -scan revealed a bony defect in the anterior part of the medial orbital wall of the left eye. The defect is located between the frontal and maxillary bone, probably at the frontal-maxillary suture (Fig 3). As there was no decrease in quality of life for the patient, the decision was made to treat conservatively. The patient was advised to sleep on her right side. The parents were educated to consult us in case of deteriorating clinical picture. A follow-up of four years did not show any complications. Exceptional swelling still does occur in the morning and disappears spontaneously after one hour.

DISCUSSION

Anterior encephaloceles are characterised by a defect in the skull and dura with an extension of intracranial contents (1). In spite of various hypotheses, the exact etiopathogenesis is unknown: a multi-factorial cause seems to be most likely (1,2,3). Anterior encephaloceles are uncommon. Reports quote an American and European incidence of 1:35000 live births, but they are more common in South-east Asia (1/

5000 live births) (1,2). Our patient is half African, half Caucasian. Anterior encephaloceles are classified as nasofrontal, nasoethmoidal or nasoorbital and are visible in the newborn or infant. They are characterised by a frontonasal mass, broadening of the nasal bridge and hypertelorism (3). The combined use of CT-scanning, MRI and cisternography establishes the diagnosis (2,4,5). The treatment of an encephalocele is surgical and aims to create a watertight closure of the dural defect, to repair the encephalocele and to reconstruct the skull defect (1,2,4). In case of nasoethmoidal encephaloceles, post-operative CSF leakage through the nose (rhinorrhea) is not uncommon. Mahapatra and all found rhinorrhea in 22 of 103 cases after encephalocele surgery (4).

To our knowledge, post-operative CSF leakage into the periorbital soft tissues has not yet been described in the literature. Palpebral CSF leakage is a rare finding and may appear when there is a communication between periorbital structures and subarachnoidal space (6,7,8,9). Similar cases have been reported in the literature after trauma or associated with a tumour (7,10,11). The management of all CSF fistulas remains controversial. Some authors recommend surgical treatment for all cases to prevent infection of the central nervous system, while others have advocated surgical repair only if conservative treatment fails (10,12). Our patient was treated conservatively as the patient did not have a decrease in quality of life.

CONCLUSION

A CSF leakage into the orbital or periorbital region is quite rare. In our department, we encountered a patient with a CSF leakage in the periorbital soft tissues, originating from a bony defect in the medial orbital wall. It presented as an intermittent palpebral swelling after surgery for an encephalocele. To our knowledge, this is the first case of such CSF leakage after encephalocele surgery reported in the literature.

REFERENCES

- (1) Hedlund G – Review. Congenital frontonasal masses: developmental anatomy, malforma-

- tions, and MR imaging. *Pediatr Radiol* 2006; 36: 647-662.
- (2) Hoving EW – Nasal encephaloceles. *Childs Nerv Syst* 2000; 16: 702-706.
 - (3) Holmes AD, Meara JG, Kolker AR, Rosenfeld JV, Klug GL – Frontoethmoidal encephaloceles: reconstruction and refinement. *J Craniofac Surg* 2001; 12(1): 6-18.
 - (4) Mahapatra AK, Agrawal D – Anterior Encephaloceles-a series of 103 Cases over 32 Years. *J Clin Neurosci* 13: 536-539.
 - (5) Schütt SM, Kehler U, Gliemroth J, Petersen D, Sperner J – Frontoethmoidal encephalocele with ocular leakage of cerebrospinal fluid. *J Neurosurg (Pediatrics 2)*, 2004; 101: 218-221.
 - (6) Harjinder Singh Bhatoe – Blepharocoele after head injury. *Skull base*, volume 12, number 2, 2002.
 - (7) Harjinder Singh Bhatoe – Blepharocoele following head injury in a child. *Indian journal of neurotrauma* 2005, Vol.2, No.1, pp 51-53.
 - (8) Arslantas A, Vural M, Atasoy MA, Ozsandik A, Topbas S, Tel E – Posttraumatic cerebrospinal fluid accumulation within the eyelid: a case report and review of the literature. *Childs Nerv Syst* 2003; 19: 54-56.
 - (9) Sibony PA, Anand AK, Keuskamp PA, Zippen AG – A posttraumatic cerebrospinal cyst of the orbit. *J Neurosurg* 1985; 62: 922-924.
 - (10) Salame K, Segev Y, Fliss DM, Ouaknine GE – Diagnosis and management of posttraumatic oculorrhea. *Neurosurg Focus* 2000; 9(1): 3.
 - (11) Dryden RM, Wulc AE – Pseudoepiphora from cerebrospinal fluid leak: case report. *British Journal of ophthalmology* 1986; 70: 570-574.
 - (12) Caldicott WJH, North JB, Simpson DA – Traumatic cerebrospinal fluid fistulas in children. *J Neurosurg* 1973; 38(1): 21-9.
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