[3] Han JJ, Massagli TL, Jaffe KM. Fibrocartilaginous embolism – an uncommon cause of spinal cord infarction a case report and review of the literature. Arch Phys Med Rehabil 2004;85:153–7.
[4] Moorhouse DF, Burke M, Keohane C, et al. Spinal cord infarction caused by cartilage embolus to the anterior spinal artery. Surg Neurol 1992;37:448–52.
[5] AbdelRazek MA, Mowla A, Farooq S and al. Fibrocartilaginous embolism: a comprehensive review of an under-studied cause of spinal cord infarction and proposed diagnostic criteria. J Spinal Cord Med 2016;39:146-154.
[6] Bots GT, Wattendorf AR, Buruma OJ, et al. Acute myelopathy caused by fibrocartilaginous emboli. Neurology 1981;31:1250–6.
[7] Hayes MA, Creighton SR, Boysen BG, et al. Acute necrotizing myelopathy from nucleus pulposus embolism in dogs with intervertebral disc degeneration. J Am Vet Med Assoc 1978;173:289–95.
[8] Novy J, Carruzzo A, Maeder P, et al. Spinal cord ischemia. Clinical and imaging patterns, pathogenesis, and outcomes in 27 patients. Arch Neurol 2006;63:1113–20.
[9] Masson C, Pruvo J, Meder J, et al. Spinal cord infarction: clinical and magnetic resonance imaging findings and short term outcome. J Neurol Neurosurg Psychiatry 2004;75:1431–5.
[10] Meng YY, Dou L, Wang CM, et al. Spinal cord infarction presenting as Brown-Séquard syndrome from spontaneous vertebral artery dissection: a case report and literature review. BMC Neurol 2019;19:321.

https://doi.org/10.1016/j.jocn.2020.05.021

# Spontaneous symptomatic orbital meningoencephalocele in an adult patient. Case report and review of the literature


Marco Zurita [a,†], Carlos Candanedo [a,*,†], Samuel Moscovici [a], Andrew H. Kaye [a,b], Sergey Spektor [a]

[a] Department of Neurosurgery. Hadassah-Hebrew University Medical Center, Jerusalem, Israel
[b] Department of Surgery, The University of Melbourne, Parkville, VIC, Australia

ARTICLE INFO

*Article history:*
Received 28 March 2020
Accepted 3 May 2020

*Keywords:*
Adult
Blepharocele
CSF leak
Meningoencephalocele
Orbital roof

ABSTRACT

Symptomatic spontaneous meningoencephalocele (MEC) is a very rare entity in adults and there have been no reported cases of spontaneous MEC through the orbital roof in an adult. We report a 41-year-old woman who presented with a left eyelid swelling for several weeks without any history of trauma. Brain magnetic resonance imaging (MRI) showed a MEC through the orbital roof causing a significant blepharocele in this young patient. Supraorbital craniotomy was performed to repair the bone defect. The symptoms resolved immediately after surgery. Even though blepharocele is a rare manifestation of spontaneous orbital MEC it should be considered in the differential diagnosis for appropriate surgical management.



## 1. Introduction

Encephalocele is a herniation of cranial contents beyond the normal confines of the skull base or calvaria [1], and spontaneous MEC in an adult is rare, with an overall incidence of approximately 1 in 35,000 persons [2]. The herniated sac may contain meninges (meningocele [MC]), or both brain matter and meninges (meningoencephalocele [MECs]) or may communicate with the ventricles (meningoencephalocystocele) [1]. The extension of the orbital MEC and cerebrospinal fluid (CSF) into the eyelid (known as blepharocele) is a rare clinical manifestation [3] usually secondary to trauma [3–5]. Most cases are reported in children and only one spontaneous adult case was reported with a meningocele [6] without brain tissue inside the bone defect. We report a very unusual case of an adult patient with a spontaneous symptomatic orbital MEC.

## 2. Case report

### 2.1. Clinical presentation

A 41-year-old female patient, with a history of asthma, but otherwise healthy, was referred to an ophthalmologist after she complained of left eyelid swelling for several weeks, more intense during night-morning, and improving during day course. (Fig. 1A) Patient denied previous abnormal nasal or ear discharge, chronic headache or any visual disturbance. There was no history of head injury, infection of the central nervous system or other neurological pathologies.

Her neurological examination was otherwise unremarkable with normal body mass index, and without signs of congenital craniofacial deformation.

Neuroradiological studies (computed tomography [CT] (Fig. 2C) and magnetic resonance imaging [MRI]) demonstrated a bone defect in the left orbital roof, 8 × 13 mm in size with MEC within the superior orbit. Per MRI, the brain filled the cranial defect with slight extension into the orbit. Most of the blepharocele was caused by the CSF accumulation inside the orbit (Fig. 2A-B).

### 2.2. Surgical procedure

A left curvilinear skin incision was made behind the hair line without shaving, interfascial dissection was done to preserve the periosteum and temporal muscle aponeurosis. A supraorbital frontal craniotomy (Fig. 2D) was performed, providing extradural subfrontal approach. The bone defect was found in the anterior part of the orbital roof with a dural sac inside it. The sac contained gliotic brain descending into the orbit with brisk CSF flow along the herniation. It was excised completely. The dural defect was covered with a patch of temporal aponeurosis and was sutured,

* Corresponding author.
*E-mail address:* ccandanedomd@hotmail.com (C. Candanedo).
† Dr. Zurita and Dr. Candanedo made equal contributions to this manuscript.

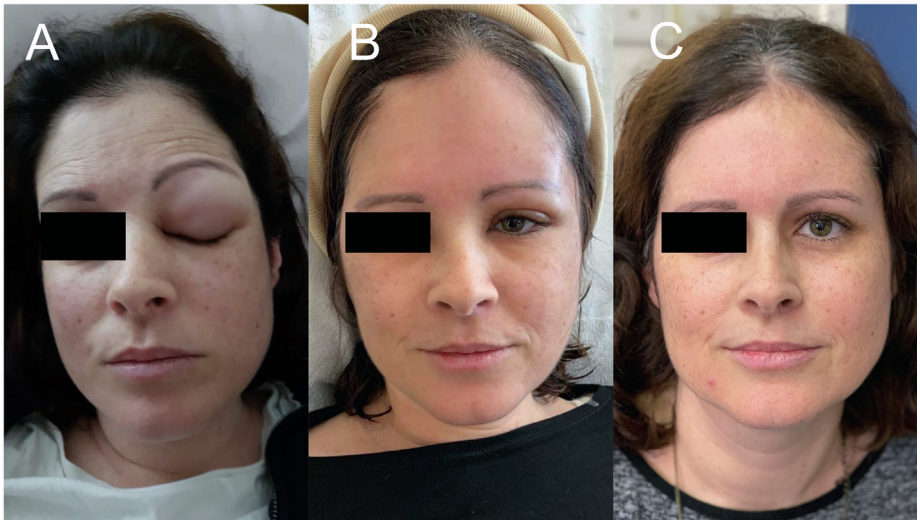


**Fig. 1.** A) 41-year-old female with spontaneous blepharocele before the surgery. Two postoperative pictures were taken B) 2 weeks after the surgery, and C) at the outpatient clinic visit 3 months later.

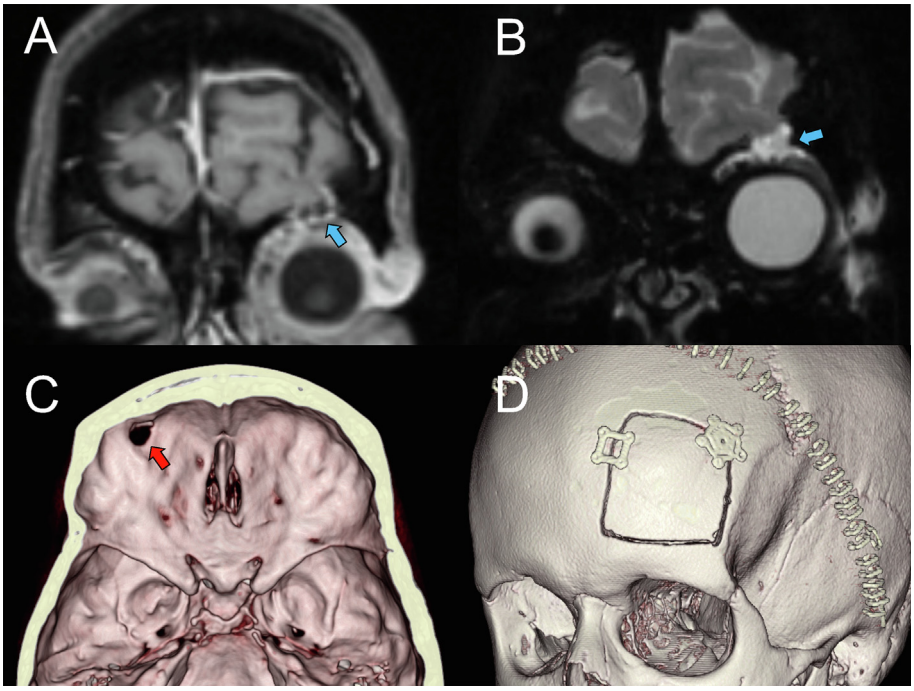


**Fig. 2.** Preoperative coronal views of brain A) T1 gadolinium-enhanced weighted magnetic resonance imaging (MRI) and B) T2 weighted MRI showing the 8x13mm bone defect with a meningoencephalocele (MEC) (blue arrows) in the left orbital roof. 3D reconstruction head CT showing the C) preoperative bone defect (red arrow) in the left orbital roof and D) a supra-orbital craniotomy performed.

meticulously in a watertight fashion, with Prolene 5–0, TF needle (Ethicon, Cornelia, GA, USA).

The bone defect was packed with a fat graft wrapped tightly into the Surgicel sheet (Ethicon, Somerville, N.J., USA) and it was forcefully plugged into the orbital bony defect. The pedicle periosteal flap was placed in between the dura and the orbital roof for reinforcement.

The surgery was uneventful and there was no need for postoperative spinal drainage. The patient showed immediately improvement of the blepharocele (Fig. 1B), without relapse at a 3-months (Fig. 1C) and 1-year follow-up. The histology was consistent with meningoencephalocele.

## 3. Discussion

We report a case of a patient with a symptomatic spontaneous orbital MEC without history of trauma. We did not find other reports of spontaneous MEC through the orbital roof in the literature. Interestingly, the patient did not have any sign of rhinorrhea or external CSF leak; the fluid accumulated in the orbital fat, contained with periorbital tissue, causing significant eyelid swelling.

The etiology of non-acquired MEC remains unclear since the terms congenital and spontaneous are used quite loosely [7] and it is difficult to define whether the bone defects are truly

**Table 1**
Published cases of spontaneous anterior skull base encephalocele in literature.

| Location | N° of patients | Type of herniation | sCSF Rhinorrhea | Meningitis | Blepharocele | References |
|---|---|---|---|---|---|---|
| Nasofrontal | 1 | MEC | – | – | – | [17] |
| Transsphenoidal | 4 | MEC | + | – | – | [13–15,18–20] |
| Transethmoidal | 7 | MC, MEC | + | †+† | – | [8–12,15,21,22] |
| Spheno-ethmoidal | 2 | MEC | + | + | – | [15,23,24] |
| Orbital roof | 1 | MC | – | – | + | [6] |
| | 1 | MEC | – | – | + | Present case |

Abbreviations: sCSF, spontaneous cerebrospinal fluid; MEC, meningoencephalocele; MC, meningocele.
†Not presented in all of the patients.

congenital or formed by bone remodeling or reabsorption during the patient's life.

MEC has been reported usually secondary to trauma [3–5], and only few spontaneous symptomatic anterior skull base encephalocele cases have been reported in the literature [8–17], mostly through the sphenoid or ethmoid sinuses (Table 1). One author reported a case of a spontaneous orbital roof meningocele [6] in an adult patient, but no brain tissue was identified herniated through the bone defect.

The dilemma is how to repair this type of defect. The orbital surgeons were involved into the discussion and they suggested the transorbital route, with or without endoscopic assistance. Our concerns with transorbital route were: 1) The ability of a watertight dural defect suturing, 2) the ability of tight plugging of the cranial defect and 3) the risk of complicating with an external CSF leak through the surgical wound in case of a failure of a watertight dural closure and cranial defect obliteration. For these reasons we preferred a regular craniotomy and microscopic repair, taking care of aesthetic aspects as well. The same strategy was used by Germano et al. [6], a frontal craniotomy with subfrontal access to the orbital roof for repair of a MC.

Blepharocele, presenting as a painless and unilateral upper eyelid edema [5], is a very rare clinical manifestation of an anterior skull base encephalocele, usually observed after head injury [3]. The absence of a craniofacial deformation and the fact the bone defect is not located in a cranial suture (as in ethmoid or sphenoid encephaloceles) do not support a congenital etiology in this 41-year-old female. To our knowledge this is the first case of a spontaneous symptomatic orbital MEC in an adult patient reported in the literature.

## 4. Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

## Declaration of competing interest

The authors have no conflicts of interest to declare

## References

[1] Suwanwela C, Suwanwela N. A morphological classification of sincipital encephalomeningoceles. J Neurosurg 1972;36:201–11.
[2] Lai SY, Kennedy DW, Bolger WE. Sphenoid encephaloceles: disease management and identification of lesions within the lateral recess of the sphenoid sinus. Laryngoscope 2002;112:1800–5.
[3] Bagolini B. Leakage of spinal fluid into upper lid following trauma. AMA Arch Ophthalmol 1957;57:454–6.
[4] Bhatoe HS. Blepharocele after head injury. Skull Base 2002;12:73–6.
[5] Mathew JM, Haran RP, Chandy MJ. Post traumatic CSF blepherocele. Neurol India 1997;45:46–7.
[6] Germano RAS, Silva MV, Germano FAS, Brandão MM, Germano CS, Souza BLd, et al. Eyelid liquoric fistula secondary to orbital meningocele. Revista Brasileira de. Oftalmologia 2015;74:46–8.
[7] Papanikolaou V, Bibas A, Ferekidis E, Anagnostopoulou S, Xenellis J. Idiopathic temporal bone encephalocele. Skull Base 2007;17:311–6.
[8] Harada N, Nemoto M, Miyazaki C, Kondo K, Masuda H, Nomoto J, et al. Basal encephalocele in an adult patient presenting with minor anomalies: a case report. J Med Case Rep 2014;8:24.
[9] Bullard DE, Crockard HA, McDonald WI. Spontaneous cerebrospinal fluid rhinorrhea associated with dysplastic optic discs and a basal encephalocele: case report. J Neurosurg 1981;54:807–10.
[10] Yamashita H, Kurihara M, Kawano T, Mori K, Kunimura M. Basal encephalomeningocele in an adult–a case report and clinico-anatomical classification. No Shinkei Geka 1985;13:425–31.
[11] Kitahara Y, Takagi H, Ichikawa F, Yamada M, Ootsuka E. Basal encephalocele–a report of two cases and consideration of its pathogenetic classification. No Shinkei Geka 1988;16:983–8.
[12] Kubo A, Sakata K, Maegawa J, Yamamoto I. Transethmoidal meningoencephalocele in an elderly woman. Case report. Neurol Med Chir (Tokyo) 2005;45:322–6.
[13] Hasegawa S, Hayashi N, Kubo M, Hamada H, Kuwayama N, Shojaku H, et al. Basal encephalocele associated with hypoplasia of the internal carotid artery. Neurol Med Chir (Tokyo) 2007;47:572–5.
[14] Sasani M, Ozer AF, Aydin AL. Endoscopic treatment of trans-sellar trans-sphenoidal encephalocele associated with morning glory syndrome presenting with non-traumatic cerebrospinal fluid rhinorrhea. J Neurosurg Sci 2009;53:31–5.
[15] Hammer A, Baer I, Geletneky K, Steiner HH. Cerebrospinal fluid rhinorrhea and seizure caused by temporo-sphenoidal encephalocele. J Korean Neurosurg Soc 2015;57:298–302.
[16] Ommaya AK, Di Chiro G, Baldwin M, Pennybacker JB. Non-traumatic cerebrospinal fluid rhinorrhoea. J Neurol Neurosurg Psychiatry 1968;31:214–25.
[17] Agrawal A, Rao KS, Krishnamoorthy B, Shetty RB, Anand M, Jain H. Single stage craniofacial reconstruction for fronto-nasal encephalocele and hypertelorism in an adult. Singapore Med J 2007;48:e215–9.
[18] Kwon JE, Kim E. Middle fossa approach to a temporosphenoidal encephalocele -technical note. Neurol Med Chir (Tokyo) 2010;50:434–8.
[19] Rivierez M, Valsaint P. Spontaneous temporo-sphenoidal encephalocele. A case report. Neurochirurgie 2000;46:383–6.
[20] Soyer P, Dobbelaere P, Reizine D, Ferquel C. Transalar sphenoidal meningoencephalocele associated with buccal angiomatosis. One case. J Neuroradiol 1990;17:222–6.
[21] Matsumoto M, Akati K, Hashimoto T, Nakamura N. Basal encephalomeningocele occurring in an aged woman; a case report and the usefulness of MRI in diagnosis. No Shinkei Geka 1992;20:157–9.
[22] Mikami T, Saito K, Okuyama T, Sakamoto Y, Takahashi A, Shibata K. A case of transethmoidal meningocele showing increased activity of 99mTcHM-PAO at seizure attack. No To Shinkei 1998;50:63–7.
[23] Acherman DS, Bosman DK, van der Horst CM. Sphenoethmoidal encephalocele: a case report. Cleft Palate Craniofac J 2003;40:329–33.
[24] Lesavoy MA, Nguyen DT, Yospur G, Dickinson BP. Nasopharyngeal encephalocele: report of transcranial and transpalatal repair with a 25-year follow-up. J Craniofac Surg 2009;20:2251–6.

https://doi.org/10.1016/j.jocn.2020.05.020