Ethmoid Meningoencephalocele in a Patient with Cerebrofacial Arteriovenous Metameric Syndrome

Rodrigo Fernández-Gajardo[1], Joao Paulo Almeida[2], Suganth Suppiah[2], Ian Witterick[3], Gelareh Zadeh[2]

Key words
- Arteriovenous malformation
- Cerebrofacial arteriovenous metameric syndrome
- Extended endoscopic endonasal approach
- Meningoencephalocele

Abbreviations and Acronyms
AVM: Arteriovenous malformation
CAMS: Cerebrofacial arteriovenous metameric syndrome
CSF: Cerebrospinal fluid
ICP: Intracranial pressure

From the [1]Department of Neurological Sciences, University of Chile, Santiago, Chile; [2]Division of Neurosurgery, University of Toronto, Toronto, Ontario, Canada; and [3]Department of Otolaryngology - Head & Neck Surgery, University of Toronto, Toronto, Ontario, Canada

To whom correspondence should be addressed: Rodrigo Fernández-Gajardo, M.D., Ph.D.(c). [E-mail: rod.fernandez.g@gmail.com]

Citation: World Neurosurg. (2018) 114:1-3. https://doi.org/10.1016/j.wneu.2018.02.133

Journal homepage: www.WORLDNEUROSURGERY.org

Available online: www.sciencedirect.com

BACKGROUND: **Skull base meningoencephaloceles are a rare condition, frequently secondary to traumatic or iatrogenic causes. Cerebrofacial arteriovenous metameric syndrome (CAMS) is characterized by the presence of retinal, facial, and cerebral arteriovenous malformations (AVMs) with metameric distribution. To our knowledge, this is the first reported case associating these 2 conditions.**

CASE DESCRIPTION: **A 45-year-old woman previously diagnosed with CAMS type 2 presented with a long history of cerebrospinal fluid (CSF) rhinorrhea. Magnetic resonance imaging and digital subtraction angiography demonstrated a right-sided facial and orbital AVM extending posteriorly along the optic tract into the suprasellar cistern, and a right-sided meningoencephalocele protruding into the olfactory recess and ethmoid sinus. An extended endoscopic endonasal approach was performed to resect the meningoencephalocele and to repair the CSF leak without complications.**

CONCLUSIONS: **We report the unusual association between the development of a meningoencephalocele and a metameric syndrome, and comment on clinical implications in the management of this patient.**

INTRODUCTION

Encephaloceles most commonly occur as a sequela of trauma and have been classified as being either occipital, sincipital, or basal.[1] Sincipital encephaloceles are the anterior herniations presenting with an external facial mass, whereas basal encephaloceles, including transethmoidal subdivision, are located at the level of the anterior and middle skull base. Common presenting symptoms of basal encephaloceles include cerebrospinal fluid (CSF) rhinorrhea, meningitis, seizures, and headaches.[2]

Occasionally, vascular lesions, such as dural arteriovenous fistula and less frequently brain arteriovenous malformations (AVMs), have been related to the development of encephaloceles, CSF leaks, and/or intracranial hypertension.[3-5] Herein, we report the endoscopic repair of an ethmoid meningoencephalocele and CSF leak in a patient with cerebrofacial arteriovenous metameric syndrome (CAMS) type 2, who undertook previous embolization of the facial component of a complex AVM.

CASE REPORT

A 45-year-old woman presented to our clinic with a history of several months of CSF rhinorrhea. She had a previous diagnosis of CAMS type 2 that was treated with partial embolization of a complex facial AVM 6 years ago and followed-up without major complications. Neurologic examination demonstrated severe visual loss of the right eye with no other abnormalities. Magnetic resonance imaging revealed a right-sided orbital AVM extending posteriorly along the optic tract into the suprasellar cistern and multiple prominent vessels through a transosseous defect in the frontal bone to the superior sagittal sinus, likely representing a sinus pericranii, and a right-sided meningoencephalocele protruding into the olfactory recess and ethmoid sinus (Figure 1). Digital subtraction angiography was performed to assess the angioarchitecture of the AVM, demonstrating a right chiasmatic AVM with a classical extension along the optic nerve and the retina, associated with a large right-sided facial AVM arising from branches of the internal maxillary and facial arteries (Figure 2). The AVM was not considered amenable to endovascular therapy.

After discussing treatment options, the patient agreed to proceed with an extended endoscopic endonasal approach to resect the right anterior skull base meningoencephalocele and repair the CSF leak. The surgery was carefully performed in conjunction with the otolaryngology team. A right-sided septal flap was raised based on the posterior septal branch, the suspected meningoencephalocele was identified emanating from the anterior skull base, and the CSF leak was successfully identified in the posterior portion. The dura was incised to reveal the frontal lobe region and the pia and arachnoid that were herniating through, and this was bipolar cauterized and resected. For closure of the skull base defect, dural

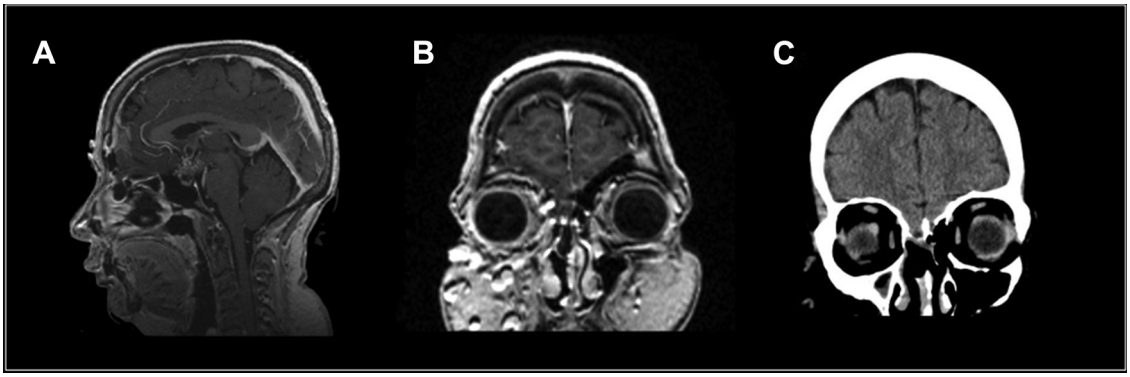

Figure 1. Preoperative gadolinium-enhanced (**A**) sagittal and (**B**) coronal brain magnetic resonance imaging and (**C**) coronal computed tomography scan demonstrating a chiasmatic arteriovenous malformation and a right-sided meningoencephalocele protruding into the olfactory recess and ethmoid sinus, likely being the site of the cerebrospinal fluid leakage.

substitutes were inserted inlay followed by a vascularized nasoseptal flap onlay. There were no complications throughout the procedure. Postoperatively, the patient presented with no signs of CSF leak or any other complications and was discharged 4 days after surgery.

DISCUSSION

Skull base encephaloceles are unusual entities frequently secondary to traumatic or iatrogenic causes in adults.[6] Spontaneous encephaloceles are a rare condition, accounting for only 3%–5% of all CSF leaks.[7-9] Previously reported case series suggest that idiopathic intracranial hypertension may be a prevalent occurrence among these patients, and it is thought to be a predisposing factor for development of encephaloceles.[2,10] Case reports have demonstrated some unusual associations with spontaneous encephaloceles, such as dural arteriovenous fistula and intracranial hypertension in pregnancy.[3,4,11]

CAMS is a rare, nonhereditary disease characterized by the association of retinal, facial, and cerebral AVMs with metameric distribution, including a spectrum of different phenotypic expressions. A somatic mutation developing in the region of the neural crest or adjacent cephalic mesoderm prior to migration is thought to produce the vascular lesions seen in CAMS.[12,13] Particularly, CAMS type 2 has been proposed to be related to a disorder in the lateral prosencephalic area with involvement of the occipital lobe, chiasm, optic nerve/retina, and maxilla, with visual loss being the most common presenting symptom.[12] Intracranial AVMs in CAMS are often clinically silent and rarely present with a neurologic deficit, intracranial hemorrhage, or seizure.[12] However, these AVMs are considered

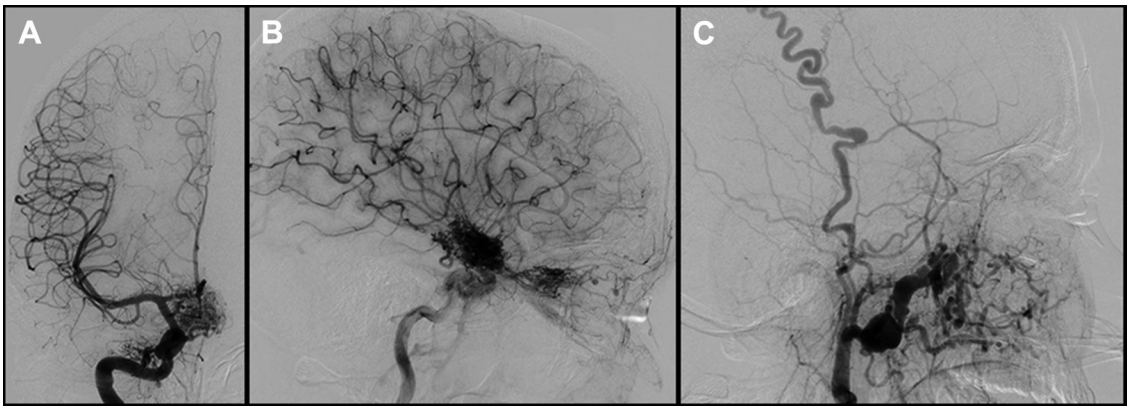

Figure 2. (**A**) Frontal and (**B**) lateral projections of the internal carotid digital subtraction angiography demonstrating a diffuse right chiasmatic arteriovenous malformation (AVM) nidus, fed by numerous, smoke-like branches of the right anterior choroidal, lenticulostriate, and ophthalmic arteries with a classical extension along the optic nerve and the retina. (**C**) Lateral projection of the external carotid digital subtraction angiography demonstrating a large facial AVM arising from branches of the right internal maxillary and facial arteries. There are several flow-related aneurysms along the branches of the right internal maxillary, and an enlarged tortuous superficial temporal artery.

dynamic and progressive, and most of them are deemed to be incurable.[14]

To our knowledge, this is the first reported case of the development of a skull base meningoencephalocele and CSF leak in a patient with CAMS. Two potential mechanisms may be involved in the association of these conditions. Embryologically, formation of the skull base and facial skeleton is a complex process involving migration of neural crest cells through ectodermal- and mesodermal-derived structures. Neural crest—derived cells participating in the pathophysiology of CAMS also contribute to the development of the prechordal chondrocranium (ie, sphenoid, ethmoid bones),[15,16] and alterations of this process may lead to the herniation of intracranial content.[17] On the other hand, elevated intracranial pressure (ICP) and papilledema have been reported in patients with unruptured brain AVMs,[18-21] most likely associated with venous hypertension and hydrodynamic alterations of CSF.[22] Moreover, a recent case report showed for the first time the association of intracranial hypertension and a CSF leak in a patient with a posterior temporal AVM, supporting the association of these events.[5]

We propose that in our case, the combination of a congenital osseous defect and the elevated ICP secondary to the AVM is responsible for the unusual clinical presentation of this patient. From a clinical perspective, the complex nature of the vascular malformations makes it difficult to achieve a definitive treatment of the AVMs, and the appropriate surgical repair of the skull base defect becomes a priority. Additionally, the fact that CAMS may be related to both sustained elevated ICP and a congenital anatomic defect makes a surgical treatment even more challenging than in idiopathic cases and warrants a strict follow-up.

CONCLUSIONS

The pathophysiology of ethmoid meningoencephaloceles is not well understood, and their treatment remains a challenge despite surgical advances. We report the case of a patient with CAMS that developed an ethmoidal meningoencephalocele potentially secondary to intracranial hypertension. Endoscopic endonasal resection of the meningoencephalocele was performed, and repair of the skull base defect was successfully achieved. CAMS is a rare disease that had not been previously related to the development of meningoencephaloceles or CSF leaks. A multidisciplinary approach is recommended for successful management of this complex clinical scenario.

REFERENCES

1. Ingraham FD, Matson DD. Spina bifida and cranium bifidum. *N Engl J Med*. 1943;228:815-820.
2. Schlosser RJ, Bolger WE. Management of multiple spontaneous nasal meningoencephaloceles. *Laryngoscope*. 2002;112:980-985.
3. Willems PW, Willinsky RA, Segev Y, Agid R. Aggressive intracranial dural arteriovenous fistula presenting with cerebrospinal fluid rhinorrhea: case report. *Neurosurgery*. 2009;65:E1208-E1209 [discussion: E1209].
4. Aggarwal V, Nair P, Shivhare P, Jayadevan ER, Felix V, Abraham M, et al. A case of evolving bilateral sphenoidal meningoencephaloceles: case report and review of the literature. *World Neurosurg*. 2017;100:708.e11-708.e17.
5. Beaudoin OX, Lévêque M, Bélair M, Saliba I, Bojanowski MW. Cerebral arteriovenous malformations causing cerebrospinal fluid fistula. *Can J Neurol Sci*. 2010;37:881-884.
6. David DJ. Cephaloceles classification, pathology, and management - a review. *J Craniofac Surg*. 1993;4:192-202.
7. Mattox DE, Kennedy DW. Endoscopic management of cerebrospinal fluid leaks and cephaloceles. *Laryngoscope*. 1990;100:857-862.
8. Burns JA, Dodson EE, Gross CW. Transnasal endoscopic repair of cranionasal fistulae: a refined technique with long-term follow-up. *Laryngoscope*. 1996;106:1080-1083.
9. Zlab MK, Moore GF, Daly DT, Yonkers AJ. Cerebrospinal fluid rhinorrhea: a review of the literature. *Ear Nose Throat J*. 1992;71:314-317.
10. Nyquist GG, Anand VK, Mehra S, Kacker A, Schwartz TH. Endoscopic endonasal repair of anterior skull base non-traumatic cerebrospinal fluid leaks, meningoceles, and encephaloceles. *J Neurosurg*. 2010;113:961-966.
11. Yee LM, Kacmar RM, Bolden JR. Basal transethmoidal encephalocele and malignant hypertension in a parturient with a seizure disorder. A case report. *J Reprod Med*. 2015;60:55-57.
12. Bhattacharya JJ, Luo CB, Suh DC, Alvarez H, Rodesch G, Lasjaunias P. Wyburn-Mason or Bonnet-Dechaume-Blanc as cerebrofacial arteriovenous metameric syndromes (CAMS). A new concept and a new classification. *Interv Neuroradiol*. 2001;7:5-17.
13. Lasjaunias P. A revised concept of the congenital nature of cerebral arteriovenous malformations. *Interv Neuroradiol*. 1997;3:275-281.
14. Jiarakongmun P, Alvarez A, Rodesch G, Lasjaunias P. Clinical course and angioarchitecture of cerebrofacial arteriovenous metameric syndromes. Three demonstrative cases and literature review. *Interv Neuroradiol*. 2002;8:251-264.
15. Balczerski B, Zakaria S, Tucker AS, Borycki AG, Koyama E, Pacifici M, et al. Distinct spatiotemporal roles of hedgehog signalling during chick and mouse cranial base and axial skeleton development. *Dev Biol*. 2012;371:203-214.
16. McBratney-Owen B, Iseki S, Bamforth SD, Olsen BR, Morriss-Kay GM. Development and tissue origins of the mammalian cranial base. *Dev Biol*. 2008;322:121-132.
17. Ieva AD, Bruner E, Haider T, Rodella LF, Lee JM, Cusimano MD, et al. Skull base embryology: a multidisciplinary review. *Childs Nerv Syst*. 2014;30:991-1000.
18. Barrow DL. Unruptured cerebral arteriovenous malformations presenting with intracranial hypertension. *Neurosurgery*. 1988;23:484-490.
19. Chimowitz MI, Little JR, Awad IA, Sila CA, Kosmorsky G, Furlan AJ. Intracranial hypertension associated with unruptured cerebral arteriovenous malformations. *Ann Neurol*. 1990;27:474-479.
20. Vorstman EB, Niemann DB, Molyneux AJ, Pike MG. Benign intracranial hypertension associated with arteriovenous malformation. *Dev Med Child Neurol*. 2002;44:133-135.
21. David CA, Peerless SJ. Pseudotumor syndrome resulting from a cerebral arteriovenous malformation: case report. *Neurosurgery*. 1995;36:588-590.
22. Rossitti S. Pathophysiology of increased cerebrospinal fluid pressure associated to brain arteriovenous malformations: the hydraulic hypothesis. *Surg Neurol Int*. 2013;4:42.

Conflict of interest statement: The authors declare that the article content was composed in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

Received 27 November 2017; accepted 22 February 2018

Citation: World Neurosurg. (2018) 114:1-3.
https://doi.org/10.1016/j.wneu.2018.02.133

*Journal homepage: **www.WORLDNEUROSURGERY.org***

*Available online: **www.sciencedirect.com***

1878-8750/$ - see front matter