Encephaloceles and meningoencephaloceles of the base of the skull: imaging, differential diagnosis and pictorial essay.

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Learning objectives

Objective of this study is to evaluate the meningoencephaloceles of the skull base, define the classification, the causes of the onset and the imaging characteristics.
Background

*Meningoencephaloceles* are a congenital or acquired malformation characterized by protrusion of the meninges and brain tissue through a defect of the skeleton of the skull.

**CLASSIFICATION**

According to their location are classified as: occipital, sincipital (naso-frontal, naso-ethmoidal, nasal-orbital), of the cranial vault, of the posterior fossa and of the skull base.

Have an *incidence* of between 0.1 and 0.5 in every 1000 live births with no significant gender differences. Has not been demonstrated familiarity.

**MENINGOENCEPHALOCELES OF THE BASE**

Meningoencephaloceles of the base of the skull represent about 1.5 - 10% of all encephaloceles with an incidence of approximately about 1 in every 35,000 live births.

In accordance with the classification of Suwanela and Suwanela of 1972, as subsequently modified by Gerhardt in 1979, are divided into *four sub-types: trans-ethmoidal, spheno-ethmoidal, trans-sphenoidal and spheno-orbital.*

Abiko et al further distinguished two types of transsphenoidal meningoencephaloceles: the *intrasphenoidal and the true transsphenoidal.* The first one describes meningoencephaloceles extending into the sphenoid sinus but restricted by its floor. The latter describes encephaloceles traversing the floor of the sphenoid sinus and protruding into the nasal cavity or nasopharynx.
Imaging findings OR Procedure details

**TRANS-SPHENOIDAL MENINGOENCEPHALOCELES**

The trans-sphenoidal variant is the most common among encephaloceles of the base with an *annual incidence of about 1 in every 700,000 live births.*

Pollock has proposed the following *etiological theories* to explain its occurrence: lack of fusion of the ossification centers, increased intraventricular pressure with progressive erosion of the underlying bone or postnatal enlargement of small bony dehiscences, *persistence of the cranio-pharyngeal canal.*

**THE CRANIO-PHARINGEAL CHANNEL OR STERNBERG’S CHANNEL**

The persistence of this channel is a rare entity described for the first time from Landzert in 1868. It was reported in 5-10% of live births and approximately 0.42% of the asymptomatic population.

The channel has an *oval or circular shape*, through the body of the sphenoid from the midline of the floor of the sella turcica down until the nasopharynx at the junction between the vomer and rostrum of the sphenoid.

*Currarino* *et al* have divided this anomaly into two different subtypes in relation to their size: small channel or pituitary channel with a maximum width of about 15 mm and larger channel, most frequently related to encephaloceles and other abnormalities of the midline.

**The cause of the persistence of the channel is quite controversial:** some authors support the hypothesis that the channel is the *remnant of the ascending path followed by Rathke pouch* through the sphenoid from the nasopharynx to the sella turcica. This hypothesis is supported by histological studies have found that the presence of normal and abnormal pituitary tissue within the channel, from case reports describing the presence of sub-sellar or nasopharinx craniopharyngiomas associated with this anomaly or case reports describing the presence of intra-sphenoidal pituitary tissue. Other authors simply assert that the cranio-pharyngeal canal is the *remnant of a vascular channel* containing a small artery and veins embedded in a stromal connective tissue.

**SPHENOID BONE EMBRIOLOGY**

However, to correctly understand the embryological origin of the cranio-pharyngeal canal it is useful to recall the sphenoid bone embryology.
During embryological development the anterior sphenoid bone (presphenoid), the lesser wings, the posterior sphenoid bone (basisphenoid), the greater wings and the lateral pterygoid processes are built up first as independent cartilaginous precursors. Ossification starts at the third month of foetal life. While the presphenoid, lesser wings and basisphenoid, as well as the greater wings and pterygoid processes, fuse at the time of birth, only a weak cartilaginous union between the greater wings and the presphenoid/basisphenoid has been found in neonates. Bony fusion of the greater wings with the presphenoid/basisphenoid starts in its anterior portion. Posteriorly fusion can be incomplete, creating a lateral craniopharyngeal canal. This canal was mentioned by Sternberg in 1888. While Sternberg consistently found this canal in children at the age of 3 to 4 years, he described an incidence of only 4% in adults. A congenital bony defect resulting from incomplete fusion of the different sphenoid bone components can communicate with the sphenoid sinus after the necessary pneumatization has taken place.

SYMPTOMS

Symptoms are unilateral rhinorrhea, malfunction of the gland associated with its compression, unexplained hypophysectomy after adenoidectomy, recurrent meningitis after removal of nasopharyngeal mass.

IMAGING

Diagnostic imaging makes use of CT scan with window to show the bone defect and MRI expecially in differential diagnosis and to highlight the type of herniated tissue and its relationship to adjacent structures. CT cisternography, 3D reconstructed CT scan an MRI provide excellent three-dimensional definition of the lesion useful for diagnosis and surgical planning.

SURGICAL TREATMENT

Surgical treatment consists of two approaches: transcranial and trans-palatal. The first method allows a good repositioning of the herniated tissue with closure of the defect with a periosteal flap with or without bone fragment. The transpalatal approach is characterized by reduced invasiveness, often also the coexistence of a cleft palate makes this approach much more easier. Most frequent complications of this technique are infections.

TRANS-ETHMOIDAL MENINGOENCEPHALOCELES
The trans-ethmoid variant is due to a defect of the cribriform lamina, usually small and unilateral. This defect may have a congenital, traumatic, post-surgical or spontaneous origin.

The congenital forms are more frequently associated with encephaloceles and other craniofacial anomalies (lacrimal duct obstruction, agenesis of the corpus callosum).

The herniated tissue includes portions of the frontal lobes and olfactory apparatus and simulates the presence of polyps.

Almost one half of basal meningoencephaloceles are identified during the first year of life due to the presence of an intranasal mass lesion with manifestations such as nasal obstruction, nasal discharge, mouth breathing and snoring, and/or associated anomalies such as hypertelorism, cleft lip and palate, bifid nose, and coloboma. The presence of an intranasal mass with pulsation synchronous with the pulse and/or respiration in a newborn or young child indicates occult basal encephalocele, since nasal polyps are most uncommon in this age group.

SYMPTOMS

Common symptoms include: intermittent unilateral rhinorrhea, headache, nasal obstruction, olfactory disorders, recurrent meningitis and seizures.

IMAGING

Diagnostic imaging makes use of CT scan with window to show the bone defect and MRI especially in differential diagnosis and to highlight the type of herniated tissue and its relationship to adjacent structures. CT cisternography, 3D reconstructed CT scan an MRI provide excellent three-dimensional definition of the lesion useful for diagnosis and surgical planning.

SURGICAL TREATMENT

Surgical treatment involves endoscopic endonasal approach. The bone defect is repaired with a layer of fat intradural, epidural layer of bone and a flap of vascularized nasal mucosa. Required is the subsequent balloon compression. This repair with multilayer graft is presently preferred for to block the loss of cerebrospinal fluid but also to reinforce the base of the skull and prevent recurring encephaloceles.
The sphen-o-ethmoidal variant consists of a defect in ethmoid and sphenoid bone with encephalocele in the posterior nasal fossa.

**SPHENO-ORBITAL MENINGOENCEPHALOCELES**

The sphen-maxillary variant consists of herniation of meninges and tissue through the superior orbital fissure.
Fig. 1: Embriology of sphenoid bone. Eur Arch Otorhinolaryngol (2000) 257:430-432 © Springer-Verlag 2000

Fig. 2: STERNBERG'S CHANNEL. Eur Arch Otorhinolaryngol (2000) 257 :430-432 © Springer-Verlag 2000

Fig. 3: STERNBERG’S CHANNEL. Eur Arch Otorhinolaryngol (2000) 257 :430-432 © Springer-Verlag 2000

Fig. 4: STERNBERG’S CHANNEL. Turkish Neurosurgery 2012, Vol: 22, No: 2, 242-245

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Fig. 5: CT CISTERNOGRAPHY IN INTRASPHENOIDAL MENINGOENCEPHALOCELE. Eur Arch Otorhinolaryngol (2000) 257:430-432 © Springer-Verlag 2000

Fig. 7: BONE DEFECT IN RIGHT SPHENOID SINUS. CORONAL CT

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Fig. 6: BONE DEFECT IN RIGHT SPHENOID SINUS. AXIAL CT

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Fig. 9: RIGHT INTRASPHENOIDAL MENINGOENCEPHALOCELE. CORONAL T2 MR

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Fig. 10: RIGHT INTRASPHENOIDAL MENINGOENCEPHALOCELE. AXIAL MR.

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Fig. 8: RIGHT INTRASPHENOIDAL MENINGOENCEPHALOCELE. T1 CORONAL MR

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Fig. 11: POST-SURGERY CORONAL CT.

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**Fig. 12**: POST SURGERY CORONAL MR

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Fig. 13: TRANSETHMOIDAL MENINGOENCEPHALOCELE. Neurol Med Chir (Tokyo) 45, 322-326, 2005

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Fig. 14: TRANSETHMOIDAL MENINGOENCEPHALOCELE. Turkish Neurosurgery 2008, Vol: 18, No: 3, 281-285

Fig. 15: GIANT TRANSETHMOIDAL MENINGOENCEPHALOCELE. Hindawi Publishing Corporation Case Reports in Medicine Volume 2012, Article ID 763259

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Fig. 16: GIANT TRANSETHMOIDAL MENINGOENCEPHALOCELE. Hindawi Publishing Corporation Case Reports in Medicine Volume 2012, Article ID 763259

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Conclusion

Meningoencephaloceles of the skull base are very rare pathological entity. Their recognition first makes use of the detection of the symptoms and then of CT scan and MRI also useful in post-operative evaluation.
References


Encephalomeningocele cases over 10 years in Thailand: a case series Sitthiporn Agthong*1 and Viroj Wiwanitkit - BMC Neurology 2002, 2:3

CSF Rhinorrhea and Recurrent Meningitis Caused by Transethmoidal Meningoencephaloceles. Parul Garg Vinita Rathi Satish K. Bhargava Anju Aggarwal*. INDIAN PEDIATRICS VOLUME 42__OCTOBER 17, 2005


Transethmoidal Meningoencephalocele in an Elderly Woman -Case Report- Atsuhiko KUBO, Katsumi SAKATA, Jiro MAEGAWA*, and Isao YAMAMOTO Neurol Med Chir (Tokyo) 45, 322¿326, 2005

Transcranial Approach for Spontaneous CSF Rhinorrhea due to Sternberg's Canal Intrasphenoidal Meningoencephalocele: Case Report and Review of the Literature. Turkish Neurosurgery 2012, Vol: 22, No: 2, 242-245

An intranasal mass K GOWDA, MD, M FARRUGIA, MD, FRCR and C PADMANATHAN, FRCR. The British Journal of Radiology, 79 (2006), 269-270
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