Intracranial hypertension management in spontaneous skull base meningoencephaloceles

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Abstract

Spontaneous meningoencephaloceles are often associated with cerebrospinal fluid leak and intracranial hypertension. There is a data scarcity concerning the need for insertion of a cerebrospinal diversion device in these situations. Therefore, we provided an analysis of our patients with spontaneous skull base meningoencephaloceles (MEC) whether their definitive surgical treatment requires management of elevated intracranial pressure (ICP) or not. Retrospective evaluation and data collection of 10 subjects with spontaneous MEC was held. Among others, attention was paid to measuring of ICP, prior interventions, treatment with acetazolamide, characteristics for long term elevated ICP, etc. Our own indications for cerebrospinal fluid (CSF) diversion and use of postoperative external lumbar drain (ELD) underwent analysis as well. The sphenoid region was the most common location of MEC. CSF leak was diagnosed in all subjects. The most common graphical signs of elevated ICP were empty sella and arachnoid pits, both of which were presented in 90% of cases. A lumbar puncture with opening pressure measurement was performed in 7 patients. Ventriculoperitoneal shunt insertion was indicated in 4 cases prior to and in two cases after skull base repair. Two postoperative CSF leaks were managed with ELD and subsequent shunt installation. Spontaneous MECs are often associated with CSF leak. The ICP assessment should be a standard of care to ensure MEC operative repair success. Insertion of a CSF diversion device must be considered where direct or indirect signs of ICH are presented.

Introduction

By extension, a cephalocele is defined as a herniation of cranial contents through a defect in the skull. Depending on the content of the prolapsed mass the nomenclature distinguishes terms such us meningocele and meningoencephalocele (or encephalocele). In the narrow sense, meningoencephaloceles (MEC) present a condition when the cranial content prolapses through a defect in the skull base, from congenital or acquired origin. Acquired MEC may be classified as nontraumatic (e.g., caused by tumors or infections), traumatic (e.g., following accidental or iatrogenic trauma), or spontaneous, when the cause in not identified. The occurrence is closely related to the presence of spontaneous cerebrospinal fluid (CSF) fistulas, though a leakage may not always be the first symptom of MEC [1, 2].

Their clinical manifestation, especially during adulthood, depends on the anatomical location and associated pathological conditions. Apart from the above-mentioned CSF leak (e.g., rhinorrhea, otorrhea, occult fistulas), a large spectrum of intracranial infections (e.g., meningitis, ventriculitis, cerebritis/brain abscesses, subdural/epidural empyema), seizures, headache, chronic sinusitis, nasal fullness, middle ear effusions, otitis, conductive hearing loss etc. may lead to the diagnosis of MEC. Sepsis due to spontaneous MEC is an uncommon presentation, the most common associated bacteria being streptococcus, haemophilus and staphylococcus [3].

According to the anatomical location there are numerous proposed classifications for skull base MEC [4–8]. The most used, summarized by Connor in 2010, classifies three types: syncipital (anterior fossa), basal (anterior or middle fossa) and posterior. Basal MECs, the type most familiar to neurosurgeons, do not present with an external mass [9].

The pathogenesis of cephaloceles has been poorly understood but is very broad. For example, any process causing increased intracranial pressure (ICP), usually in the longer term, could lead to the development of MEC [10, 11]. In the field of spontaneous skull base MEC, this theory alone is the most adopted. Prior studies have demonstrated a strong association with idiopathic intracranial hypertension (IIH) [1, 12]. Unknown or untreated intracranial hypertension (ICH) leading to the formation of MEC and resulting in CSF fistula (or vice versa) present a risk of subsequent meningitis with possible devastating and life-threatening consequences.

Surgical repair (endonasal endoscopic or transcranial) of spontaneous MEC is recommended as the first line, to prevent principally infectious complications [4, 8, 13, 14]. Concomitant direct (ICP > 20 cm H_2O) or indirect (clinical and imaging findings) signs of ICH should suggest the need for medical or surgical treatment. Last year's experiences at our department demonstrated the necessity of surgical management of ICH in a short sequence following the skull base repair. The ideal was to perform both surgeries during one general anesthesia once there were obvious indices. Hence, we offer our perspective for handling spontaneous skull base MEC with elevated ICP.

Materials And Methods

Retrospective data collection and evaluation of subjects with spontaneous skull base MEC who underwent surgical repair at the Department of Neurosurgery of the University Hospital Hradec Králové between December 2013 and January 2021 were carried out. Institutional approval from the Ethics Committee of the University Hospital Hradec Králové was obtained prior to the study. Meningoencephaloceles were considered spontaneous when there was no previous history of trauma or tumor in the location of the skull base defect. Assessed variables included demographics, BMI, type/location of the encephalocele, prior predisposing pathologies, surgical techniques for skull base reparations, and outcomes. Also studied were relevant data concerning increased ICP, such as clinical signs, ICP measuring via lumbar puncture (LP), prior interventions (e.g., for CSF leak, ventriculoperitoneal or lumboperitoneal shunt), treatment with acetazolamide, and graphical characteristics for long term elevated ICP. Finally, our own indications for CSF diversion insertion were analysed. The first author (P.P.) and a radiologist (J.J.) reviewed the accessible imaging of the patient, and the presence of the following radiographic signs of increased ICP were evaluated: empty sella, optic nerve (CN II) vertical tortuosity, distended CN II sheaths, and aberrant arachnoid granulations (so called arachnoid pits).

Results

Patient characteristics

We identified 10 patients (6 women, 4 men, mean age 56 years) with spontaneous skull base MEC who underwent neurosurgical intervention in the specified period. Table 1 offers their basic characteristics. Six participants qualified as obese (BMI>30 kg/m^2), with an average BMI of 32.4 kg/m^2. Two patients

admitted with meningitis declared a CSF leak prior to the infectious complication. Three other patients treated primarily for meningitis presented a rhinorrhea prior to skull base repair during the hospitalization (Table 1). Cerebrospinal fluid leakage was diagnosed (clinically, by detection of beta-trace protein or beta-2 transferrin) in all subjects. Two patients had a known history of elevated ICP (IIH and Torkildsen shunt, patients 7 and 5 respectively), one presented a cerebellar gangliocytoma (incidentally diagnosed during CSF leakage investigation, patient 6), and one patient had suffered from meningitis in childhood (patient 8). A lumbar puncture with opening pressure measurement was performed preoperatively in 6 patients (one unsuccessful), and once after the skull base repair (patient 4).

Table 1

Group characteristics

Pt.	Age (y)	Sex	BMI (kg/m^2)	MEC location	1. diagnosed sign	LP	Op. press. (cm H_2O)	Surgery	VP shunt	ELD	Empty sella	CN II distension	CN II tortuosity	Arac pits
1	83	m	26,2	SS (left lateral rec.)	Meningitis	Y	NA	Cranio	N	N	Y	Bilat.	Bilat.	Y
2	73	w	42,9	Cribriform plate (bilat.)	CSF leak	N	NA	Cranio	N	N	Y	Left	Left	Y
3	62	w	34,3	SS (left lateral rec.)	CSF leak	Y	NA	Cranio	N	N	Y	N	N	Y
4	44	w	30,9	Cribriform plate (bilat.)	Meningitis*	Y#	40	Cranio	Y‡	Y	Y	Bilat.	N	Y
5	51	m	14,7	FS (right dorsal wall)	Meningitis*	Y	20	Cranio	N	N	Y	NA	NA	N
6	32	m	29,4	SS (bilat. lateral rec.)	CSF leak	Y	25	EEA	Y‡	Y	N	Bilat.	Bilat.	Y
7	53	w	39,0	Cribriform plate (MC)	CSF leak	Y	25	Cranio	Y	N	Y	Left	Left	Y
8	43	w	30,1	SS (right lateral rec.)	CSF leak	Y	30	Cranio	Y	N	Y	N	Bilat.	Y
9	59	w	24,3	Mpl. (ant./middle fossa)	Meningitis	Y	22	EEA	Y	N	Y	Bilat.	N	Y
10	63	m	51,9	SS (left lateral rec.)	Meningitis	Y	NA	Cranio	N[Ref]	N	Y	Bilat.	Bilat.	Y

Pt, patient; *y*, year; *m*, man; *w*, woman; *MEC*, meningoencephalocele; *SS*, sphenoid sinus; *rec.*, recess; *bilat.*, bilateral; *FS*, frontal sinus; *MC*, meningocele; *Mpl.*, multiple localization; *ant.*, anterior; *CSF*, cerebrospinal fluid; *LP*, lumbar puncture; *N*, no; *Y*, yes; *Op. press.*, opening pressure at lumbar puncture; *NA*, not applicable; *Cranio*, craniotomy; *EEA*, endoscopic endonasal approach; *VP*, ventriculoperitoneal; *Ref*, refused by the patient; *ELD*, external lumbar drainage; *CN II*, second cranial nerve/optic nerve

*patient not being aware of CSF leak before meningitis #LP after the skull base repair ‡VP shunt insertion under a separate general anesthesia

Imaging analysis

All sphenoid sinus MECs extended through the lateral recess of the sinus (Fig. 1). Figures 2 and 3 offer other examples of MEC localization. There was only one case of meningocele. In 90% of cases both CT and MRI scans showed an empty sella (Fig. 4 and 5) and arachnoid pits. The CN II tortuosity and sheaths distention were analyzed on MRI on a sample of 9 subjects (patient 5 had no MRI). Sheaths distention and tortuosity, at least on one side, were present in 77.8% and 66.7%, respectively (Table 1).

Surgical skull base repair and IH management

Among 8 patients having skull base repair through a craniotomy, two underwent a VP shunt insertion during the same anesthesia. In one case the VP shunt was placed a few days following the skull base repair. The endoscopic endonasal approach (EEA) was performed in two patients. One subject received a VP shunt in a second step. The other one, presenting multiple small MECs, first had VP shunt installation, and the following EEA did not reveal any ongoing CSF leakage (also with the use of fluorescein). Two postoperative CSF leaks, in patients 4 and 6, were first managed with external lumbar drainage (ELD). Later, in the same hospitalization they underwent VP shunt installation (Table 1).

Discussion

Most cases of spontaneous MEC and spontaneous CFS leakage go hand in hand. The natural history of their formation is now better understood.

Spontaneous CSF leaks have a high rate (50–100%) of encephalocele formation [15–17]. Our cohort of MECs presented a 100% occurrence of CSF leak prior to the surgical skull base repair. Both entities are associated with elevated ICP or there is underlying IIH. Chronically elevated ICP may lead to erosion and

thinning of the skull, including the skull base [18–21]. For a long time, there was no consensus on whether active ICP management should be standard in the treatment of spontaneous CSF leaks/MEC. In the last decade several authors demonstrated not only the association of spontaneous CSF fistulas and increased ICP, but also emphasized the need for ICH management, mainly surgical [21–24]. Likewise a few authors advocated acetazolamide administration to control the leakage [23, 25, 26]. Recently Kreatsoulas and his colleagues created a precise management protocol for dealing with spontaneous CSF rhinorrhea. They introduced a standardized postoperative (24–48 hour) LP to measure the opening pressure. Patients with mild ICP elevation (20–25 cm H_2O) received acetazolamide, and those with ICP > 30 cm H_2O received CSF diversion. Those with ICP between 25 and 30 cm H_2O were indicated to shunt insertion if significant other factors were present (i.e., imaging findings, large defect, concern for patient compliance, very high BMI, known IIH) [17].

All forms of information about ICP conditions are helpful for a surgeon to decide about ICH management. For many years we have been following the idea of opening pressure measurement, but mostly prior to MEC surgery. Having only a few cases of spontaneous MEC/CSF leaks in the past and ambiguous literary data, we took the view of minimal invasiveness in term of CSF diversion. Of note, unsystematic acetazolamide use was applied only perioperatively. Ventriculoperitoneal shunt insertion was indicated in 6 patients – in 4 cases prior to skull base repair (based on the opening pressure at LP, and once based on the indirect graphical signs of ICH and high BMI); and in two cases after skull base surgery (based on elevated opening pressure and postoperative CSF leak) (Table 1). Our experiences show the need for intracranial pressure assessment. We share the opinion of previous studies showing that immediately after closure of the skull base defect, ICP significantly increases during the postoperative period [17, 23].

However, we believe that a patient already suffering from ICH before the skull base repair (without other diseases that would clearly lead to ICH) will have an elevated ICP also after the surgery. We have support for this statement in only one case out of six (patient 6). There are no data either to confirm or disprove our theory. Nevertheless, a huge metanalysis from Teachey showed that evaluation and intervention for elevated ICP in spontaneous CSF leaks is associated with significantly improved success rates [21].

Graphical indicators for ICH are often apparent on preoperative imaging. They might not only draw attention to a more likely explication for encephaloceles or rhinorrheas [1, 2, 17, 21], but might also be helpful in the decision-making process for whether or not to insert a CSF diversion device. The same applies to high BMI (patient 10). The rise in obesity worldwide will continue to have a major impact on the incidence of spontaneous CSF leaks due to increased ICP [24, 27].

Concerning skull base repair in MEC, we advocate both approaches (craniotomy and EEA). Temporal craniotomy was performed in larger defects of the middle fossa with important lateral extension of the sphenoid sinus recess (patients 1, 3, 6, 8 and 10). Nevertheless, having gained more experience and observed the tendencies worldwide [28–32], we now tend to use endonasal endoscopic techniques.

The small cohort of patients and the absence in the past of a systematic protocol in the care of patients with spontaneous MEC do not allow us to offer results arising from statistical analysis. However, sharing our experiences and results from this retrospective study might be helpful when dealing with spontaneous MEC. Once diagnosed, this pathology demands an immediate solution with the aim, in particular, of preventing the development of infectious complications. It is also often related to increased ICP, which will be a more frequent phenomenon in clinical practice. So the ICP should be managed before or after the repair of the skull base defect.

Conclusion

Spontaneous MECs are strongly associated with CSF leak and in most cases, we should look at them through the same prism. One of their common denominators is the increased ICP. Its assessment should be an essential element in the standard of care to ensure operative repair success. Once the ICP is elevated prior to/after the skull base repair and there are obvious graphical signs of ICH, insertion of a CSF diversion device must be considered.

Declarations

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Author contribution Conceptualization: PP, KZ, TČ; Methodology: PP, MC, JA; Formal analysis and investigation: PP, MC, KZ, JA, PČ, RK; Writing - original draft preparation: PP, MC, KZ, PČ, RK; Writing - review and editing: PČ, JA, TČ; Supervision: TČ.

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Data availability Correspondence and requests for materials should be addressed to PP.

Code availability Not applicable.

Ethical approval This study was approved by the Ethics Committee of the University Hospital Hradec Králové.

Consent to participate Not applicable.

Consent for publication All authors have reviewed the manuscript and are in agreement with submission for publication in its current form.

Conflict of interest The authors declare no competing interests.

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Figures

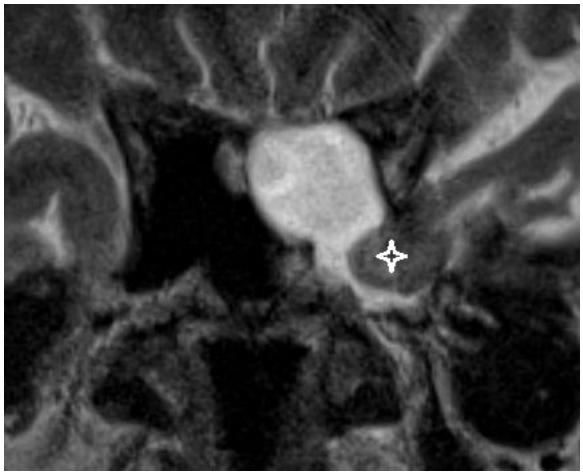

Figure 1

Left sided lateral sphenoid meningoencephalocele (asterisk). T2w MRI image in coronal plane

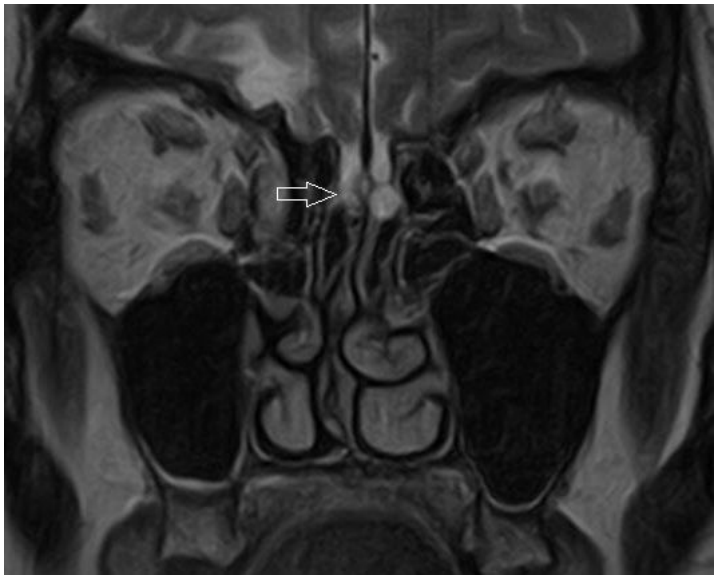

Figure 2

Meningoencephaloceles (arrow) pushing through the cribriform plate. T2w MRI image in coronal plane

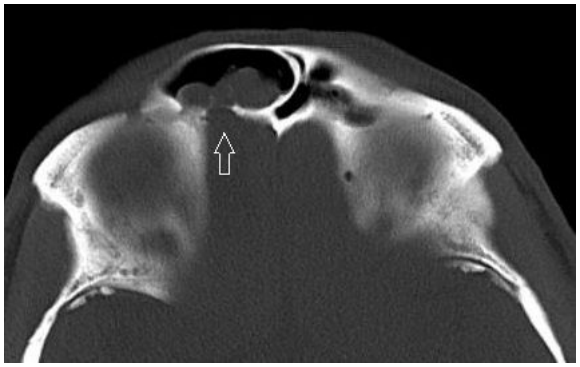

Figure 3

Right sided frontal sinus meningoencephalocele (arrow). Bone window CT scan in axial plane

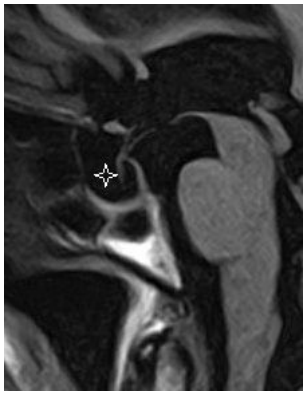

Figure 4

Empty sella (asterisk). Sagittal T1w MRI image

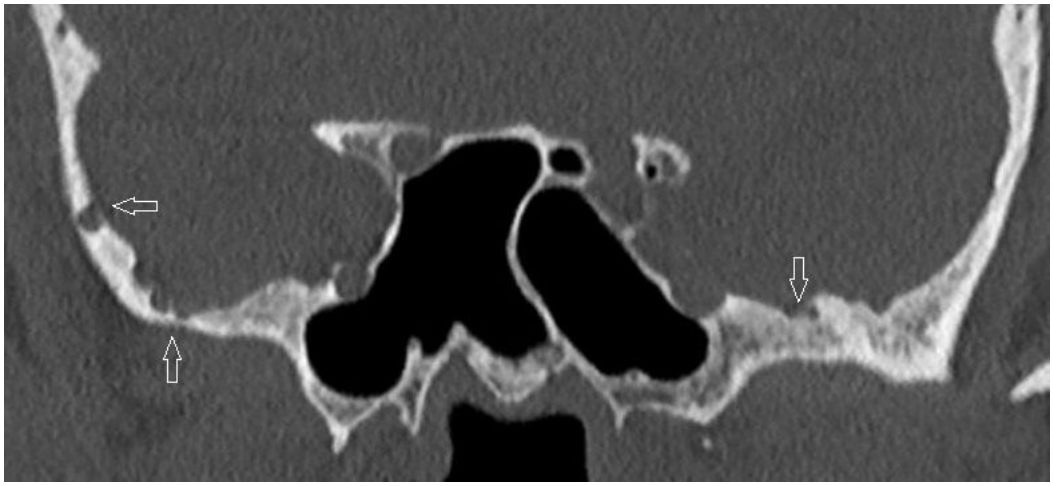

Figure 5

Intradiploic sphenoid arachnoid pits (arrows). Bone window CT scan in coronal plane