Pathophysiology and Treatment Options in Trigeminal Meningoceles

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Abstract

Trigeminal meningoceles, lateral to the maxillary nerve (V2), have seldom been reported as underlying pathology for spontaneous rhinoliquorrhea. In contrast to sphenoid meningoceles arising from a persistent lateral craniofaryngeal canal (Sternberg–Cruveilhier, medial to V2), their occurrence seems to be generated by addition of erosive processes to the constitutively thin bony shell underneath the semilunar ganglion, lateral to the round foramen (and V2).

The developmental and anatomical relationships of trigeminal meningoceles to the sphenoid bone are depicted, and in a review of the literature we present the different surgical approaches employed for sealing the dura leak. In view of these techniques we discuss an unusual case of therapy-resistant rhinoliquorrhea with left-sided trigeminal meningocele involving the Meckel cave at the lateral sphenoid and reaching the superior orbital fissure and the medial orbital space.

In contrast to patients who have lateral sphenoidal meningoceles with a persistent lateral craniofaryngeal canal (Sternberg–Cruveilhier), who can be treated successfully using an endoscopic transsphenoidal approach (recurrence rate 13.7%), the recurrence rate of cerebrospinal fluid (CSF) efflux for trigeminal meningoceles lies much higher (endoscopically 66%, open craniotomy 33%). The surgical strategy thus has to be chosen individually, taking into account specific anatomical situations and eventually preceding operations.

Keywords
- cerebrospinal fluid (CSF)
- trigeminal meningocele
- spontaneous rhinoliquorrhea
- Sternberg–Cruveilhier canal
- lateral craniofaryngeal canal

Introduction

Among the different entities causing rhinoliquorrhea in closest proximity to the sphenoid bone, trigeminal meningoceles (TMs), lateral sphenoidal meningoceles (LSMs), and the persistence of the lateral craniofaryngeal canal (Sternberg–Cruveilhier canal, SC) have been described.

TMs have only been reported several times in the literature, and nomenclature is still heterogeneous. In general, reports cover various mass lesions in the Meckel cave, ranging from lipoma and meningioma to schwannoma. Only a few deal with meningoceles in the pterygopalatine region, such as in patients with typical stigmata of neurofibromatosis Type 1 (NF 1).1,2

TMs are localized in the lateral sphenoid wing lateral to V2 and the foramen rotundum underneath the semilunar ganglion, reaching to the superior orbital fissure, pterygopalatine fossa, and medial orbital space, mostly emerging from an

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enlarged Meckel cave and giving rise to osseous erosion of the sphenoid wing.

As a different and more frequent reason for rhinoliquorrhea, LSMs have been described. They lie medially to V2 near the base of the sphenoidal bone and can be distinguished from TMs by the absence of orbital or pterygopalatine fossa involvement.

The third dura leak–associated pathology of this region is the developmental anomaly of a persistent SC embryonal canal (lateral craniopharyngeal canal). Still controversially discussed in literature, it has been found in some cases as an underlying condition for LSMs. The canal lies medially to the foramen rotundum and extends from the maxillary nerve root (V2) to a recess of the lateral sphenoid wall, opening into the sphenoid sinus.

**Anatomical Considerations**

From an anatomical point of view, there are several “weak spots” in the embryonic development of the sphenoidal bone, which can explain the occurrence of meningoceles or spontaneous rhinoliquorrhea, especially in the area of the trigeminal ganglion and the carotid artery.

The cartilage precursor of the skull base has to form around the cranial nerves, arteries, and veins while respecting their lumen. This can lead to thinned cranial base structures that might be eroded during a lifetime by inflammatory processes or increased intracranial pressure.

The bone formation itself is inhomogeneous. The collision zone between endochondral ossification (lesser wing) and intramembranous ossification (greater wing) lies laterally to the foramen rotundum, extending to the region of the foramen ovale. In a computed tomographic (CT) study, 25 patients with lateral sphenoidal cerebrospinal fluid (CSF) leaks were all shown to have the bony defects in this area, lateral to the foramen rotundum.

The sutures between the ossification centers can leave small basal clefts, such as the lateral craniofaryngeal canal (SC). The canal represents a remnant of the fusion zone between the alisphenoid (greater wing) and the basisphenoid (SC). The canal appears to be imprecise, because LSMs also arise from it. The terms transalar sphenoid meningocele and transsphenoidal and transethmoidal meningoceles have been used synonymously in literature.

We suggest the term lateral sphenoidal meningocele to describe pathologies medial to V2 and the term trigeminal meningoceles as anatomically more precise for lesions lateral to the foramen rotundum and V2.

**Nomenclature**

In literature, many different terms have been employed to describe the pathology of a TM. *Arachnoid cyst of Meckel’s cavity* appears to be imprecise, because LSMs also arise from it. The terms transalar sphenoid meningocele and transsphenoidal and transethmoidal meningoceles have been used synonymously in literature.

**Illustrative Case**

A 29-year-old woman presented to the outpatient clinic for rhinoliquorrhea. She also reported a left-sided orbital swelling, especially when bending forward or in prone position. The patient experienced headaches, intermittent rhinoliquorrhea, vomiting, and chronic conjunctivitis. Her first meningitis was at the age of 17 years, a second occurred a few years later, and both had been treated successfully with antibiotics. A former traumatic head injury was denied and basal cranial fractures had been ruled out with multiple imaging techniques.

Extended neuroradiological imaging included cisternographic magnetic resonance (MR) scan after intrathecal gadolinium-application in prone position. It revealed a left-sided CSF leak along a CSF-containing enlargement of the temporal fossa that extended into the pterygopalatine fossa. CSF was seen in the periorbital fatty tissue and the temporal muscle. Origin of the fistula was suspected in the temporal muscle (Fig. 2a). Origin of the fistula was suspected in the temporal muscle (Fig. 2a). Origin of the fistula was suspected in the temporal muscle (Fig. 2a). Origin of the fistula was suspected in the temporal muscle (Fig. 2a). Origin of the fistula was suspected in the temporal muscle (Fig. 2a). Origin of the fistula was suspected in the temporal muscle (Fig. 2a).

The surgical strategy was discussed in the multidisciplinary skull-base board and under suspicion of a temporomandibular meningocele of the maxillary nerve, a pterional craniotomy with transsylvian approach and closure of the parasellar...
entry point of the meningocele was advised. In a first operation, the suspected entry point of the fistula was closed using multiple layers of subgaleal tissue. The efflux of CSF was stopped but reappeared after several months. The patient then decided to be treated in another neurosurgical department, where a second pterional operation was performed without relieving the symptoms.

About 2 years after first surgery, the patient decided to restart treatment in our institution. Intrathecal contrast-enhanced CT revealed the refilled fistula and an enlarged, CSF-containing space in the paraclival region, close to the maxillary nerve and the Meckel cave. A third pterional exploration was proposed, but the patient opted for conservative therapy. Only after increasing orbital swelling and reappearance of rhinoliquorrhea did the patient agree to another intervention.

After this intervention, an oculomotor palsy prompted surgical revision. Fusion of the CT dataset with neuronavigation pictures allowed identification of the entrance of the Meckel cavity. This was covered by dura mater and could be punctured for aspiration of CSF. The wall of the cavity showed a cisternlike arachnoid covering in which the nerve fibers crossing the cavity were partly adhering to the basal arachnoid layer of the cyst and were spread apart. Two walls of the cavity were found, corresponding to arachnoid cystic structures and dural tissue, thus displaying typical criteria for meningoceles. To close the fistula, abdominal fat and muscle tissue were harvested and the periarachnoid space of the Meckel cave was filled in proximal and distal direction. Hereafter, fat tissue was positioned in the remaining arachnoid space rostrally. Between fat and muscle tissue, liquid dura glue (DuraSeal Xact, Covidien, Mansfield, Massachusetts, USA) was injected into the cavity along the maxillary nerve. Additional muscle was placed on top and covered with fibrin glue. The opening of the cavity was sealed with TachoSil (Takeda Pharmaceuticals; Zurich, Switzerland) and Surgicel (Ethicon, Somerville, New Jersey, USA).

After this intervention, rhinoliquorrhea and orbital swelling disappeared and the third nerve palsy nearly completely recovered within 3 months.

However, about 4 months postoperatively the patient experienced an intermittent CSF leak. Shortly thereafter, orbital swelling and conjunctivitis reoccurred. T2- and CISS 3D T2-scans revealed a partly occluded, yet CSF-containing, meningocele. The patient refused any further interventions, especially implantation of a ventriculoperitoneal (VP) shunt to reduce the intracranial CSF pressure, and she was lost to follow-up.
<table>
<thead>
<tr>
<th>Patient</th>
<th>Age/ Gender</th>
<th>Symptoms</th>
<th>Diagnosis</th>
<th>Radiological findings</th>
<th>Therapy</th>
<th>Complications</th>
<th>f/u</th>
</tr>
</thead>
<tbody>
<tr>
<td>Present case</td>
<td>30/f</td>
<td>Orbital swelling, rhinoliquorrhea, cephalgia</td>
<td>TM, SC</td>
<td>Dysplasia of left sphenoid</td>
<td>Periortal craniotomy (3×)</td>
<td>Recurrent leakage; temporary third nerve palsy</td>
<td>24 months</td>
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<tr>
<td>Chapman et al (2000)</td>
<td>8/m</td>
<td>Meningitis, rhinosinusitis</td>
<td>TM</td>
<td>Enlargement of Meckel cave, enlargement of SOF</td>
<td>Middle fossa approach</td>
<td>CSF collection temporal left</td>
<td>18 months</td>
</tr>
<tr>
<td>Blaivie et al (2006)</td>
<td>73/f</td>
<td>Rhinoliquorrhea, meningitis</td>
<td>TM</td>
<td>Paramedial left lacuna in sphenoidal sinus</td>
<td>Transsphenoidal endoscopic surgery</td>
<td>Died during follow-up from cardiac failure</td>
<td>4 months</td>
</tr>
<tr>
<td>Castelnuovo et al (2007) 15 cases</td>
<td>60.3 ± 6.3, 9 female, 6 male</td>
<td>Rhinoliquorrhea (x = 15), headache and meningitis</td>
<td>SC</td>
<td>Sphenoidal dysplasia</td>
<td>Paraseptal direct transsphenoidal approach (×7), transeptal-pterygoid-sphenoidal approach</td>
<td>No major complications, no recurrences</td>
<td>37.6 ± 21.7 months</td>
</tr>
<tr>
<td>Tomazic et al (2009) 5 cases</td>
<td>44 ± 2, 4 female, 1 male</td>
<td>Rhinoliquorrhea (x = 5), cephalgia (x = 2)</td>
<td>SC</td>
<td>3×right-, 2×left-sided dysplasia of sphenoid with SC</td>
<td>3× transeptal-pterygoid-sphenoidal approach</td>
<td>1×maxillary nerve irritation, 2× recurring liquor, 1×bacterial meningitis</td>
<td>Up to 4 months</td>
</tr>
<tr>
<td>Tabaee et al (2010) case 1</td>
<td>61/f</td>
<td>Rhinoliquorrhea, cephalgia</td>
<td>LSM, SC</td>
<td>Sphenoidal dysplasia</td>
<td>Transnasal endoscopic surgery</td>
<td>None</td>
<td>11.3 years</td>
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<tr>
<td>Tabaee et al (2010) case 2</td>
<td>37/f</td>
<td>Rhinoliquorrhea, cephalgia</td>
<td>LSM, SC</td>
<td>Sphenoidal dysplasia</td>
<td>Transnasal endoscopic surgery</td>
<td>Perioperative meningitis</td>
<td>12.1 years</td>
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<tr>
<td>Tabaee et al (2010) case 3</td>
<td>78/f</td>
<td>Cephalgia, meningitis</td>
<td>LSM, SC</td>
<td>Sphenoidal dysplasia</td>
<td>Transnasal endoscopic surgery</td>
<td>None</td>
<td>0.7 years</td>
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<tr>
<td>Tabaee et al (2010) case 4</td>
<td>44/f</td>
<td>Rhinoliquorrhea, cephalgia</td>
<td>LSM, SC</td>
<td>Sphenoidal dysplasia</td>
<td>Frontotemporal craniotomy</td>
<td>None</td>
<td>5.0 years</td>
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<td>Tabaee et al (2010) case 5</td>
<td>36/f</td>
<td>Rhinoliquorrhea, cephalgia</td>
<td>LSM, SC</td>
<td>Sphenoidal dysplasia</td>
<td>Frontotemporal craniotomy</td>
<td>None</td>
<td>5.7 years</td>
</tr>
<tr>
<td>Tabaee et al (2010) case 7</td>
<td>73/f</td>
<td>Rhinoliquorrhea, cephalgia, meningitis</td>
<td>LSM, SC</td>
<td>Sphenoidal dysplasia</td>
<td>Transsphenoidal endoscopic surgery</td>
<td>None</td>
<td>3.2 years</td>
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</tbody>
</table>
# Table 1 (Continued)

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age/ Gender</th>
<th>Symptoms</th>
<th>Diagnosis</th>
<th>Radiological findings</th>
<th>Therapy</th>
<th>Complications</th>
<th>f/u</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tabaee et al (2010) case 8</td>
<td>44/f</td>
<td>Rhinoliquorrhea, cephalgia</td>
<td>LSM, SC</td>
<td>Sphenoidal dysplasia</td>
<td>Transethmoidal endoscopic surgery</td>
<td>Persistent leak requiring revision, endoscopic closure</td>
<td>3.6 years</td>
</tr>
<tr>
<td>Tabaee et al (2010) case 9</td>
<td>59/m</td>
<td>Rhinoliquorrhea</td>
<td>LSM, SC</td>
<td>Sphenoidal dysplasia</td>
<td>Transethmoidal endoscopic surgery</td>
<td>None</td>
<td>2.9 years</td>
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<tr>
<td>Tabaee et al (2010) case 10</td>
<td>56/m</td>
<td>Rhinoliquorrhea</td>
<td>LSM, SC</td>
<td>Sphenoidal dysplasia</td>
<td>Transethmoidal endoscopic surgery</td>
<td>Persistent leak that closed spontaneously</td>
<td>3.2 years</td>
</tr>
<tr>
<td>Tabaee et al (2010) case 11</td>
<td>73/m</td>
<td>Rhinoliquorrhea</td>
<td>LSM, SC</td>
<td>Sphenoidal dysplasia</td>
<td>Transethmoidal endoscopic surgery</td>
<td>None</td>
<td>3.3 years</td>
</tr>
<tr>
<td>Tabaee et al (2010) case 12</td>
<td>47/m</td>
<td>Rhinoliquorrhea, cephalgia</td>
<td>LSM, SC</td>
<td>Sphenoidal dysplasia</td>
<td>Transnasal endoscopic surgery</td>
<td>None</td>
<td>4.3 years</td>
</tr>
<tr>
<td>Tabaee et al (2010) case 13</td>
<td>68/m</td>
<td>Rhinoliquorrhea, cephalgia</td>
<td>LSM, SC</td>
<td>Sphenoidal dysplasia</td>
<td>Transnasal endoscopic surgery</td>
<td>None</td>
<td>3.2 years</td>
</tr>
<tr>
<td>Bendersky et al (2011)</td>
<td>46/f</td>
<td>Rhinoliquorrhea, meningitis</td>
<td>TM, SC</td>
<td>Herniated temporal lobe into sinus sphenoidal, bony defect related to SC</td>
<td>Transsphenoidal surgery, fronto-temporal craniotomy</td>
<td>Recurrence of CSF leak 2 months after transsphenoidal approach</td>
<td>3 years</td>
</tr>
</tbody>
</table>

Abbreviations: CSF, cerebrospinal fluid; LSM, lateral sphenoid meningocele; SC, Stemberg–Cruveilhier canal; SOF, superior orbital fissure; TM, trigeminal meningocele.
Review of the Literature

Review of the literature was performed in the Medline database using the search terms trigeminal, encephalocele, Meckel’s cave, sphenoid, pterygopalatine, CSF fistula, and Sternberg in all combinations of two keywords.

Search resulted in 38 relevant cases from 23 reports, ranging from 1888 to 2011 (Table 1). Within these 38 reports, female predominance was found (gender ratio 1.92:1 f:m). Mean age was 52.9 years (age range 8 to 78 years). Patients presented with rhinoliquorrhea in 92.1% (35 patients), headaches in 34.2% (13 patients), and meningitis in 15.7% (6 patients).

Out of 38 patients, 6 had a TM, whereas a persistent SC or LSM was found in 32 patients.

One patient did not receive surgery, leading to persistence of symptoms. A total of 29 patients underwent endoscopic surgery for LSMs, whereas in 3 cases the transcranial approach was preferred. In two cases surgery for LSMs, whereas in 3 cases the transcranial approach was preferred. In two cases surgery for LSMs, whereas in 3 cases the transcranial approach was preferred.

A third case died perioperatively due to cardiac failure during follow-up. One of three patients undergoing craniotomy and open repair experienced recurrent therapy-resistant CSF leakage (present case, 33%).

From the six TMs, three were treated by an endoscopic transsphenoidal approach, with recurrence of CSF leaks in two of them (66%). A third case died perioperatively due to cardiac failure during follow-up. One of three patients undergoing craniotomy and open repair experienced recurrent therapy-resistant CSF leakage (present case, 33%).

Apart from persistent rhinoliquorrhea, the most common complications in the endoscopic group were meningitis (6.3%) and maxillary nerve irritation (3.1%). In the craniotomy group, facial paresthesia (3.1%) and transient diplopia (3.1%) were encountered.

Discussion

The case presented here illustrates a rare TM extending from an enlarged Meckel cave into the medial cranial and pterygopalatine fossa, causing therapy-resistant CSF leakage and orbital affection with an unusual collection of CSF in the temporal muscle (Fig. 2). Although the predominant symptoms of CSF leaks such as rhinoliquorrhea, headache, and meningitis do not help in localizing the dural defect, clinical appearance may vary with the localization of the specific arachnoidal cyst, meningocele, or encephalocele. For example, arachnoid cysts of the region of the Meckel cavity often become symptomatic with facial numbness or pain due to their relation to the trigeminal nerve.

The therapeutic difficulties of that unusual entity have not been solved. In contrast to the far lateral TMs, LSMs and persistent SC can be treated successfully with a transsphenoidal approach. This may be caused by their more medial localization, medially to the foramen rotundum and V2 at the sphenoid base.

Patients with TMs are likely to have a greater benefit from a transcranial approach, which provides better access to the lateral aspects of the Meckel cave and the semilunar ganglion. This is underlined by the higher recurrence rate when using the transsphenoidal approach (Table 1). Alternatively, transmaxillary transpterygoid approaches may be employed for TMs with a limited pterygoid fossa leakage.

CSF leaks associated with TMs can be assumed to be of idiopathic origin with an elevated intracranial pressure (ICP) as an additional factor. Thus, ICP recording and ICP normalization should be taken into account, and routine use of lumbar drainage as a diagnostic and therapeutic measure has been proposed in lateral sphenoid CSF leaks.

In light of the reviewed literature, together with the presented therapy-resistant case, alternative strategies such as implantation of a VP shunt or temporary lumbar drainage to reduce CSF pressure have to be discussed. The occurrence of the spontaneous CSF leak as part of an idiopathic hydrocephalus syndrome should be considered, with acetazolamide or furosemide being a treatment option. However, the one case treated conservatively experienced persistence of symptoms, similar to our patient, who experienced recurrence after deciding for nonsurgical follow-up (Table 1).

Midfacial degloving has been suggested as approach to the pterygopalatine fossa meningoceles and remains an option for recurrence.

A staged treatment algorithm using lumbar CSF drainage, ICP recording, and invasive location of the skull base defect with intrathecal administration of contrast medium seems to be most promising. Depending on the location and anatomical shape of the TM, open craniotomy or endoscopic transpterygoid approach can be selected. Whereas for medial pathologies (SC, LSM) endoscopic transsphenoidal approaches appear to be advantageous, TMs should be primarily considered for an open craniotomy. Alternatively to transcranial approaches, transfacial or transmaxillary/transpterygoid approaches have to be considered as treatment for recurrent, extended TMs involving the pterygopalatine fossa, in which transsphenoidal endoscopic techniques have limited success rates.

In case of therapy-refractory CSF leaks and marked elevation of ICP, VP shunt placement and/or medical treatment should be considered before reintervention.

Conclusion

Compared with the spontaneous rhinoliquorrhea caused by a persistent SC or LSMs, the point of leakage in TMs lies laterally to V2 and cannot always clearly be visualized, even by sophisticated neuroradiological techniques. This can result in considerable technical difficulties and therefore therapeutic decisions should follow the advice of a multidisciplinary skull base team after taking into account the individual anatomic situation of a patient, preceding operations, and specific risk factors for elevated ICP.

References

2. de Vries J, Freihofer HPM, Menovsky T, Cruysberg JRM. Successful surgical repair of progressive exophthalmos caused by a

Table 1

<table>
<thead>
<tr>
<th>TM Localization</th>
<th>Treatment</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Medial Cranial</td>
<td>Transsphenoidal</td>
<td>Successful</td>
</tr>
<tr>
<td>Pterygopalatine</td>
<td>Transmaxillary</td>
<td>Successful</td>
</tr>
<tr>
<td>Sphenoid</td>
<td>Transcranial</td>
<td>Successful</td>
</tr>
<tr>
<td>Pterygoid</td>
<td>Transpterygoid</td>
<td>Successful</td>
</tr>
</tbody>
</table>

Table 1: Summary of TM Localization, Treatment, and Outcome.
8 Sternberg M. Ein bisher nicht beschriebener Kanal im Keilbein des Menschen. Anat Anz 1888;2:784–785
9 Radioievitch S, Jovanovitch S. [Cruevilier-Sternberg embryonal canal (lateral craniopharyngeal canal) and its vestiges in the adult human]. Rev Laryngol Otol Rhinol (Bord) 1956;77(3-4):223–232