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Intrasphenoidal encephalocele and spontaneous CSF rhinorrhoea*

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SUMMARY

Intrasphenoidal encephalocele is a rare clinical entity. In the international literature only 16 cases have been reported up today, with female predominance. Clinically they manifest at middle and advanced ages (40-67 years), when spontaneous CSF rhinorrhoea or recurrent meningitis occurs. We present our case, a 46 years old female, who had CSF rhinorrhoea from the right vestibule for 10 months. The diagnosis was based on the history and the high-resolution brain and skull base CT-scanning in conjunction with opaque fluid injection in the sub-arachnoidal space through a lumbar puncture. She was successfully treated with an operation, through an endonasal trans-ethmoid microendoscopic approach, using the Draf and Stammberger technique. We discuss the pathogenesis of the intrasphenoidal encephalocele, the existence of small occult defects in the skull base, which cause, at the middle and advanced ages, CSF fistula with spontaneous CSF rhinorrhoea and/or recurrent meningitis. Finally we emphasize the advantages of the endonasal surgical approach for the treatment of this condition.

Key words: intrasphenoidal encephalocele, microsurgical endonasal trans-ethmoid technique, obliteration of the sinus sphenoidalis, spontaneous CSF rhinorrhoea

INTRODUCTION

A combination of spontaneous cerebro-spinal fluid (CSF) rhinorrhoea and intrasphenoid sinus encephalocoele, without obvious causes such as trauma, tumour, inflammation, or surgery in the region of the nose and the paranasal sinuses, is an extremely rare occurrence (Buchfelder et al., 1987; Myssionec et al., 1987). In the literature, sporadic cases of spontaneous CSF rhinorrhoea are mentioned, and female predominance as well as localization in the rhino-ethmoid region seem to occur the most frequently, followed by the sphenoid sinus region (Benedict et al., 1991; Schasfer et al., 1976). The occurrence of intrasphenoid sinus encephalocoele seems clinically detectable at middle and advanced ages, when complications appear such as recurrent meningitis or CSF rhinorrhoea.

OUR CASE REPORT

A 46 year-old female was admitted to the ENT department of Thessaloniki University Hospital, presenting with clear, aqueous discharge from the right nasal vestibule, for 10 months, which was aggravated when bending forward. She also complained of rare headaches. Her personal and family history was clear. She only mentioned the presence of spasms in her childhood as a result of high fever due to the measles, and a similar episode 7 years ago. She did not report traumas or bouts of meningitis. ENT examination was normal. Rhinendoscopy revealed normal findings. On bending the head and trunk forward, the patient presented rhinorrhoea and thus preferred to sleep with her head in an upraised position. Ophthalmology and neurology check-ups were similarly normal. Blood tests and simple sinus X-rays were all clear. Examination of the aqueous discharge with a glucotest as well as biochemical analysis proved the fluid to be CSF. Following these, the localization of the leak was determined with high-resolution brain and base of skull CT-scanning in conjunction with opaque fluid injection in the sub-arachnoidal space through a lumbar puncture. The CT-scans axial and coronal images revealed a bone defect in the lower part of the lateral wall of the right sphenoid sinus, inside which a polyp-like mass in continuity with the endocranium was present. These findings were compatible with a intrasphenoid sinus encephalocoele (Figure.1).

Following these findings in the right sphenoid sinus, the patient underwent surgical reparation. In order to reach the sphenoid sinus, a microendoscopic endonasal trans-ethmoid approach was selected. The posterior ethmoid cells were removed, and

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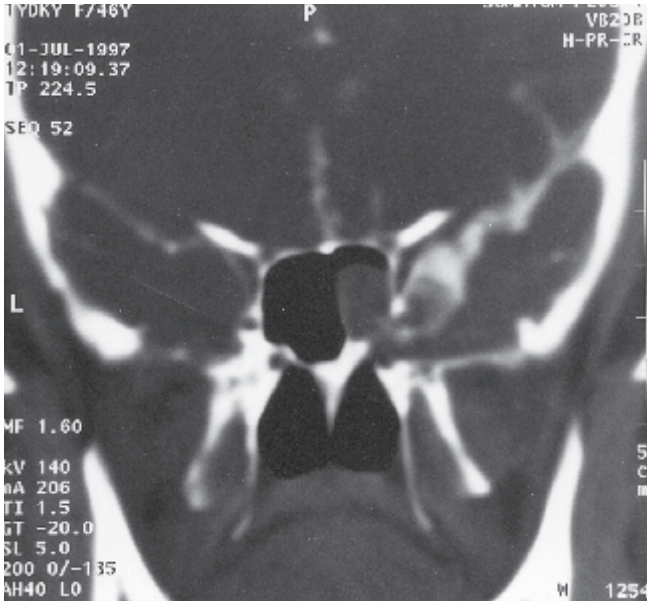

Figure 1: HRCT : Coronal view of the sphenoid sinuses at mid-level, following enhancement of the sub-arachnoid spaces with radio-opaque fluid. Bone defect may be observed in the inferior third of the lateral wall of the right sphenoid sinus, inside which a polypoid mass in continuity with the endocranium is imaged.

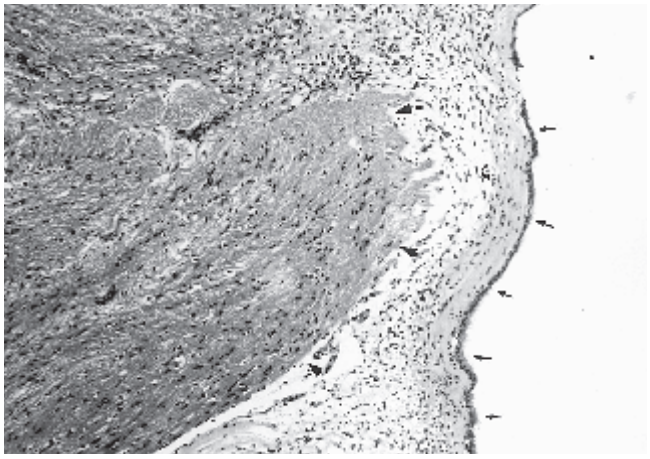

Figure 2: Respiratory-type mucosa (small arrows). Brain tissue (heads of arrows). H.E., 20×.

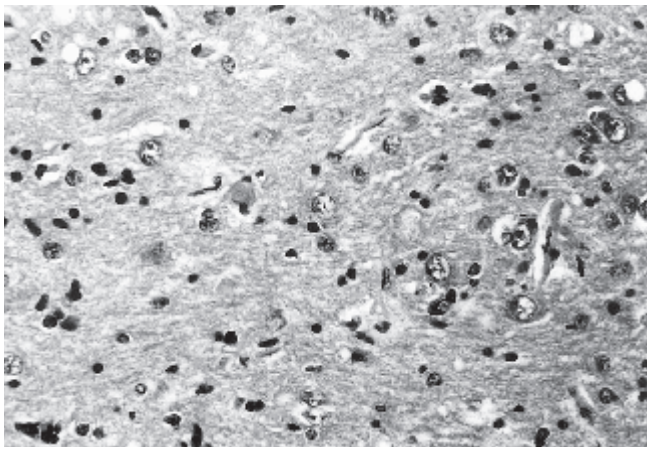

Figure 3: Enlargement of the previous photograph. Neuroglia with scattered neurons. H.E., 100×.

the anterior sphenoid wall was opened. The lateral sphenoid sinus wall was found covered by oedematous, thickened mucosa, which extended towards the sinus cavity. With the aid of a microscope, the mucosa was removed completely, and a pulsating leak of clear fluid became visible from a small bone defect in the area. This was patched with the application of lyodura on top of the defect in such a manner as to apply the edges underneath the defects corresponding edges. Fibrin glue was then applied, a larger piece of lyodura was superimposed, and the sinus cavity was obliterated with fragments of muscle tissue and surgicell. Nasal tamponnade followed, initially in the posterior ethmoid area with sponge inserted in a surgical glove finger, followed by insertion of merocel sponge in the nasal cavity. The nasal packing remained inside for 6 days under intravenous antibiotic cover. During this period, the patient remained in a semi-sitting position and avoided abrupt movements. Post-surgical recovery was uneventful.

The histological examination of the sphenoid tissues revealed the presence of brain tissue and fragments of mucosa covered by respiratory-type epithelium with sero-mucinous glands present (Figure 2,3).

Today, a year after the operation, the patient is symptom-free and a recent CT-scan shows total obliteration of the right sphenoid sinus where the bone defect is still visible (Figure 4a,b).

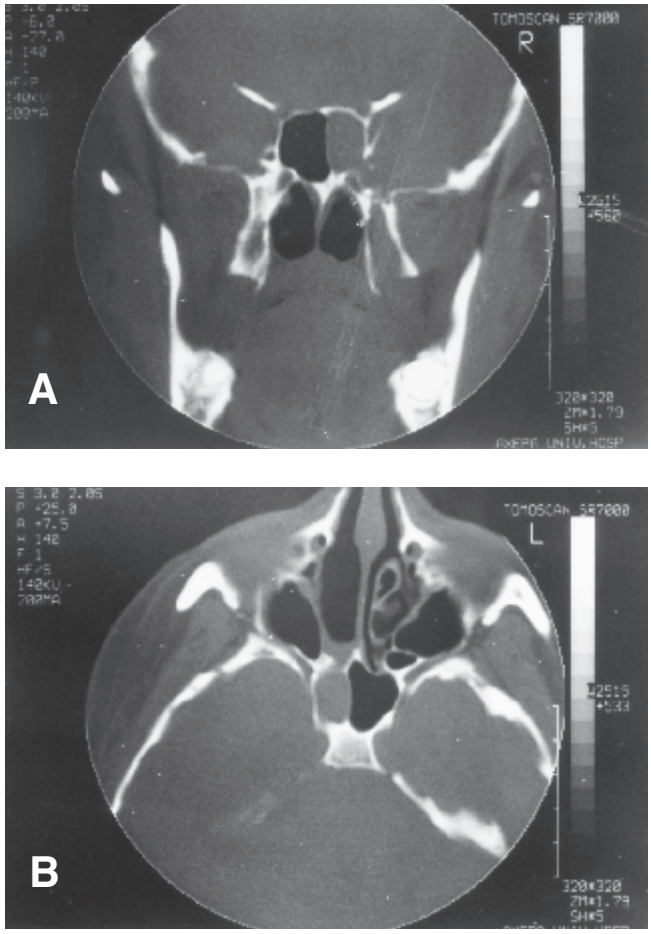

Figure 4 a,b: Coronal and axial cuts, one year following surgery, at the same level as pre-operative CT images. Complete obliteration of the right sphenoid sinus. The bone defect is visible.

DISCUSSION

Intrasphenoidal encephalocoeles are a very rare condition. They constitute a specific clinical entity of transsphenoidal encephalocoeles in that they become clinically evident at an advanced age and differ in the surgical methods of treatment (Buchfelder et al., 1987)

Their pathogenesis lies in the existence of small occult congenital dysplasias, in the form of defects or clefts, in the base of the skull in the middle cranial fossa. These bony defects constitute weak spots and, possibly under the pulsatile fluctuating pressure of the CSF, may favor the creation of meningocoeles or encephalocoeles which finally become clinically apparent as spontaneous CSF rhinorrhoea (Bernstein et al., 1997; Schick et al., 1997)

The existence of occult congenital malformations in the region of the sphenoid sinus along the scull base with the accompanying CSF leak may be attributed either to incomplete pneumatization of the sinuses or to osteogenetic disturbances of the sphenoid bone during the early stages of pregnancy, specifically at the junction between the basisphenoid and the presphenoid, or finally to embryogenetic anomalies (Benedict et al., 1991; Kaufman et al., 1979; Otto et al., 1994)

In contrast to the clinically manifest malformations of the base of the skull which cause aesthetic and functional deficits in the area of the face, occult malformations manifest themselves clinically when complications arise such as spontaneous CSF rhinorrhoea or recurring meningitis (Schick et al., 1997)

According to Buchfelder et al. (1987), until 1987 only 7 cases of intrasphenoidal encephalocoele had been mentioned in the international literature. Since then and after an overview of the literature accessible to us, we discovered 9 more cases including the present one. From this study it has become evident that in the majority of cases the bony defect is localized in the lateral or postero-lateral wall of the sphenoid sinus, involves mainly women, the patients age varies mostly between 40 to 67 years of age, and the clinical picture consists mainly of spontaneous CSF rhinorrhoea, recurring meningitis, and headaches. In two cases the bony defect was associated with an empty sella turcica, in one of which endocrinological disturbances such as hyperprolactinhaemia and hyperthyroidism were manifest (Ahmadi et al., 1985; Buchfelder et al., 1987). In our case the only symptom was rhinorrhoea for 10 months from the right nares, especially in specific body positions such as bending over forward or while during physical effort.

Bernstein et al. (1997) described 2 sisters aged 41 and 40 years old, of which the first presented with recurrent meningitis and spontaneous CSF rhinorrhoea for 20 years, while the other presented with spontaneous CSF rhinorrhoea for 2 months. In both cases a bone defect was discovered in the posterolateral wall of the left sphenoid sinus with an intrasphenoidal encephalocoele. According to the authors, the fact that the clinical symptoms were identical in both sisters as well as that rhinorrhoea appeared at an adult age in both, points towards the congenital base of skull occult bone malformation etiology and towards the fact that fluctuating endocranial CSF pressure predisposes to the formation of a CSF fistula.

Schick et al. (1997) reported 4 cases of base of skull occult malformations, two of which presented a bone defect of the lateral wall of the sphenoid sinus while the remaining two involved double bone defects in the ethmoid glabella and the apex of the petrous bone. Due to these double defects, the authors urged proper examination of the base of skull both in the frontal as well as the lateral areas. The dangers of recurrent meningitis arising from sinus bacteria were also stressed, especially those involving *Streptococcus pneumoniae.*

A spontaneous rhinorrhoea associated with an individualized tumor in the sphenoid sinus may be considered a diagnostic challenge. Careful history taking may distinguish spontaneous from traumatic CSF rhinorrhoea, however this may be difficult in cases where traumatic alterations of the meninges remain asymptomatic for long periods of time or when the patient forgets trauma (Ocada et al., 1991).

Base of skull examination with modern imaging techniques solves these problems. High-resolution computerized tomography via lumbar puncture and addition of radio-opaque solution in the sub-arachnoid space with axial and coronal images is now well accepted as being diagnostically a very efficient tool. In our case, the small bone defect of the lateral wall of the sphenoid sinus and the possible endoencephalic continuity of the sphenoid lesion were demonstrated. Surgical removal of the mass and histologic examination confirmed fully the tomographic findings. The addition of magnetic resonance imaging was judged superfluous, although MRI does add to the differential diagnosis between neoplastic and inflammatory alterations (Albernaz et al., 1991).

In dealing with spontaneous CSF rhinorrhoea and intrasphenoid encephalocoele, most authors agree to operate via an exocranial rhinosurgical approach (Albernaz et al., 1991; Benedict et al., 1991; Simmen et al., 1998; Stammberger, 1993; Willner et al., 1994). Rhinosurgery, as opposed to intracranial neurosurgical approaches, has the advantage of being more easily tolerated by the patient, has a low mortality rate, high success rates and preservation of the sense of smell (Bernstein et al., 1997; Draf et al., 1993).

In our case, we preferred an endonasal transethmoid approach using the Draf and Stammberger microendoscopic technique because the sphenoid sinus may thus be reached sooner and easier, causing as little damage as possible, and providing the possibility of cleaning out the inflammatory ethmoid cells with the aid of the surgical microscope. We prefer the transethmoidal approach instead of the approach through the space between the nasal septum and middle turbinate, because it offers a better view of the surgical field for the sphenoid sinus. With this technique the time required for the approach of the sphenoid sinus is a few minutes longer, but according to our experience it does not cause more bleeding, and the operating field is kept clear. The use of a microscope has the advantage of providing comfortable surgical maneuvers, comparing with the endoscopic method.

For the closure of the meningial fistula, lyodura was used with fibrin glue. With the same material as well as fragments of muscle tissue and surgicel, the right sphenoid cavity was obliterated

after complete removal of the mucosa. We consider as very important to the success of the operation proper packing of the posterior ethmoid cell region initially, followed by packing of the entire nasal cavity, which remains in place for 6 days under antibiotic cover. The patients compliance in avoiding sudden movements and remaining in a semi-sitting position for a week is important. In this manner, complete success was achieved. CT-scanning one year later confirmed this. The muscle packing that was used causes the haziness in the CT scan. No indication of secondary infection of the sphenoid sinus occurred.

REFERENCES

1. Ahmadi J, Weiss MH, Segall HD, Schultz DH, Zee CS, Gianotta SL (1985) Evaluation of cerebelospinal fluid rhinorrhea by metrizamide computed tomographic cisternography. Neurosurgery 16:54-60.
2. Albernaz SM, Horton DW, Adkins YW, Garen DP, (1991) Intrasphenoidal encephalocele. Otolaryngology-Head and Neck Surgery. 104:279-281.
3. Benedict M, Schultz-Coulon JH, (1991) Spontane Rhinoliquorrhae: Atiologie-Differentialdiagnose-Therapie. H.N.O. 39:1-7.
4. Bernstein MJ, Roland Thomas Jr. J, Persky SM (1997) Spenoid Cranial Base Defects in Siblings presenting with cerebelospinal fluid leak. Skull Base Surgery. 7:193-197.
5. Buchfelder M, Fahlbusch R, Huk JW, and Thierauf P (1987) Intrasphenoidal Encephaloceles, a Clinical Entity. Acta Neurochir (Wien) 89:10-15.
6. Draf W, Bergaus A (1993) Tumoren und Pseudomumoren (Tumorahnliche Lasionen) der frontalen Schudelbasis, augenhend von der Nase, der Nasennebenhohlen und dem Nasenrachenraum. Rhinochirurgishes Referat. European Archives of Oto-Rhino-Laryngology. Suppl. 1993/I. 105-186.
7. Kaufman B, Yonas H, Witt RJ, Miller CF (1979) Acquired midle cranial fossa fistulas: normal pressure and nontraumatic origin. Neurosurgery 5:466-472.
8. Myssionek D, Cohey NL (1987) Intrasphenoidal meningoencephalocele: a case report. Am. Journal of Otolaryngology. 8:391-394.
9. Ocada J, Tsuda T, Takasugi S, Nishida K,Toth Z, Matsumoto K (1991) Unusually late onset of cerebrospinal fluid rhinorrhea after head trauma. Surg. Neurol. 35:213-217.
10. Otto HD (1994) Teratogenetische und Klinische Aspecte bei Missbildungen des Kopf und Halsbereiches. Eur. Arch. Otolaryngology supp. 1:15-100.
11. Schasfer M, v Wild K, Grau H, Haacker H (1976) Zur Diagnostik und Therapie der Nasoethmoidalen Enzephalozellen. Laryng. Rhinal 55:581-585.
12. Schick B, Draf W, Kahle G, Weber R, Wallenfang T (1997) Occult Malformations of the Skull Base. Arch Otolaryngol Head Neck Surgery. 123:77-80.
13. Simmen D, Bischaff Th (1998) Rhinochirurgisches Konzept zur Versorgung von Frontobasis-defekten mit Rhinoliquorrhoe. Laryngol-Rhinol-Otol 77:264-271.
14. Stammberger H (1993) Komplikationen entzundlicher Nasennenbenhohlenerkrankungen einschliesslich iatrogen bedingter Komplikationen. European Archives of Oto-Rhino-Laryngology, suppl. 1993/I 62-102.
15. Willner A, Kantrowitz BA, Cohen FA (1994) Intrasphenoidal encephalocele: Diagnosis and management. Otolaryngology - Head and Neck Surgery. 111:834-837.

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