Case Report

Intrasphenoidal Meningo-encephalocele: Report of two rare cases and review of literature

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Abstract

Background: Transsphenoidal encephaloceles represent a rare clinical entity with wide range of symptoms in adult. Such lesions require accurate diagnosis and surgical treatment. The incidence of congenital encephalocele is very low occurring in approximately 1 in 3000-5000 live births. Even though 63 similar cases have been reported in the literature not all of them have been discussed completely. Due to the rare occurrence of these lesions, we will focus on the main clues in the diagnosis and management of such lesions, which are challenging.

Case Description: We intend to present our experience with two cases of trans-sphenoidal meningoencephaloceles, one located medially and the other herniating through the Sternberg’s canal. The younger was 17 and the elder was 47 years old and both of them presented with cerebrospinal fluid (CSF) leakage. Both patients were treated successfully using pure endoscopic endonasal approach.

Conclusion: Ideal surgical approach for such patients is still not clear due to lack of adequate experience in the literature, it is suggested that full preoperative imaging studies might lead the surgeons to undertake minimally invasive skull base approaches in similar patients.

Key Words: Basal encephalocele, cerebrospinal fluid rhinorrhea, intra-sphenoidal encephalocele, lateral cranio-pharyngeal canal, trans-sellar meningo-encephalocele

INTRODUCTION

Encephalocele may be of congenital, spontaneous, and traumatic origin. The encephaloceles are classified according to the site of origin to: Occipital, sincipital, naso-frontal, naso-ethmoidal, naso-orbital and basal, spheno-orbital, sphenomaxillary, trans-ethmoidal, spheno-ethmoidal, trans-sphenoidal, and basio-occipital cephaloceles.

Trans-sphenoidal encephalocele can occur through a defect in the sella, planum sphenoidal, or posterior ethmoid sinuses. However, the trans-sellar trans-sphenoidal type is a rare variety and our search of the EBSCO, Pubmed, and Google revealed only 61 results, which are summarized in Table 1. When there is sufficient pneumatization of the sphenoid sinus, the lateral craniopharyngeal canal (Sterngberg’s canal) may act as the site of origin of congenital meningocele or cerebrospinal fluid (CSF) fistula. The trans-sphenoidal type represent less than 5% of all basal meningoencephaloceles and has an estimated incidence of one in 700000 live births. Abiko, et al. believed that there are two types of
| Case | Author/year | Endocrine | Visual | Rhinorrhea | Transnasal | Transcranial | Transethmoid | Transpterigoid | Conservation | Transpalatal | Complication |
|------|-------------|-----------|--------|------------|------------|-------------|--------------|---------------|--------------|-------------|--------------|
| 1    | McCoy G/1963[20] | 1         | 1      | 1          |            |             |              |               |              |             |              |
| 2    | Danoff D/1966[7]  | 1         |        | 1          |            |             |              |               |              |             |              |
| 3    | DeBartolo HM/1977[28] | 1       | 1      |            |            |             |              |               |              |             |              |
| 4    | Ellyin F/1980[16] | 2         |        |            |            |             |              |               |              | 2           |              |
| 5    | Vaquero J/1981[30] | 1         | 1      | 1          |            |             |              |               |              |             |              |
| 6    | Smith DE/1983[27] | 1         |        | 1          |            |             |              |               |              | 1           |              |
| 7    | Ahmad J/1985[2]  | 1         |        | 1          |            |             |              |               |              |             |              |
| 8    | Amagasa M/1985[2] | 1         |        | 1          |            |             |              |               |              |             |              |
| 9    | Buchfelder M/1987[25] | 1    | 1      |            | 1          |             |              |               |              |             |              |
| 10   | Deasy NP/1999[3]  | 1         |        | 1          |            |             |              |               |              | 2           |              |
| 11   | Schick B/2000[20] | 1         |        | 1          |            |             |              |               |              |             |              |
| 12   | Abe T/2000[11] | 1         | 1      | 2          | 2           |             |              |               |              |             |              |
| 13   | Koral K/2000[18] | 1         |        | 1          |            |             |              |               |              |             |              |
| 14   | Jabre A/2000[19] | 1         |        | 1          |            |             |              |               |              | 1           |               |
| 15   | Fraioli B/2003[12] | 1     | 1      |            |            |             |              |               |              |             |              |
| 16   | Formica F/2002[11] | 1   | 1      |            |            |             |              |               |              |             |              |
| 17   | Shamma R/2002[22] | 1         |        | 1          |            |             |              |               |              | 1           |               |
| 18   | Chen CS/2004[46] | 1         | 1      | 1          |            |             |              |               |              |             |              |
| 19   | Kahyaoglu/2007[17] | 1        |        |            |            | 1           | 1            |               |              |             |              |
| 20   | Hasegawa S/2007[14] | 1     | 1      |            |            |             |              |               |              |             |              |
| 21   | Spacca. B/2007[20] | 5       | 4      | 1          | 2           | 1           | 2            | 3             |               |             |              |
| 22   | Peltonen E/2008[21] | 1     |        |            |            |             |              |               |              |             |              |
| 23   | Sare GM/2009[26] | 1         | 1      | 1          |            |             |              |               |              |             |              |
| 24   | Franco D/2009[33] | 2         | 1      | 1          |            |             |              |               |              | 5           |               |
| 25   | Tabaei A/2010[29] | No reported | 11  | 5          | 5           | 3           |              |               |              |             | Two patient had CSF leak and one required endoscopic revision. one case experienced meningitis post operative |
| 26   | Maric. A/2010[19] | 1         | 1      | 1          |            |             |              |               |              | 1           |               |
| 27   | HashemiB/2010[15] | 1         | 1      |            |            |             |              |               |              |             |               |
| 28   | Rathore YS/2010[23] | 1     | 1      |            |            |             |              |               |              |             |               |
| 29   | Rathore YS/2011[24] | 2       | 4      | 4          |            |             |              |               |              |             | Recurrence of rhinorrhea in one case |
trans-sphenoidal meningoencephaloceles based on the integrity of the floor of the sphenoid sinus, that is, the intra-sphenoidal and the true trans-sphenoidal types.[5] The former describes those extending into the sphenoid sinus but confined by its floor. The latter describes those traversing the floor of the sphenoid sinus and protruding into the nasal cavity or naso-pharynx. In trans-sphenoidal encephalocele, the age of manifestation of disease ranges from newborn to 64 years, while the intra-sphenoidal encephalocele seem to manifest in the fifth or sixth decade of life.[5]

We intend to report cases of a 15-year-old boy with large intra-sphenoidal meningocele and a 45-year-old female with laterally located sphenoidal meningomyelocoele both presenting with CSF leakage and treated by endoscopic endonasal approach. Discussing the management of these cases might be informative for young neurosurgeons interested in the field of pediatric skull base surgery.

CASE REPORTS

Case 1
A 17-year-old boy was referred complaining from headache and intermittent CSF rhinorrhea of 2 years duration. In physical examination, hypertelorism, cleft lip, high arch palate, and see saw nystagmus were detected. Endocrinological examinations showed deficient growth hormone (GH), adrenocorticotropic hormone (ACTH), and luteinizing hormone (LH). Brain magnetic resonance imaging (MRI) revealed intrasphenoidal meningocystocele with infundibular recess descending into the meningocele and no other brain anomaly [Figure 1]. Brain angiography, showed hypoplasia of the left A1 segment. The patient was operated by endoscopic transnasal transsphenoidal approach. Preoperatively, it was planned to pack the epidural space of the floor of the sella. At surgery, epidural packing was impossible because of tight adhesion of the fibrous tissues around the base of the meningocele. The epidural space could not be teased off the surrounding bone structures, so the cavity of the cyst and the sphenoid sinus were packed with autogenous abdominal fat supported by dural glue. There was no rhinorrhea in 3-and 6-month follow up.

Case 2
A 45-year-old female was admitted complaining of refractory headache and recent CSF rhinorrhea. She had no history of head trauma, meningitis or previous surgical operation. In physical exam, cranial nerves were intact and laboratory tests including hormonal studies were all normal. In MRI, the sella was enlarged and filled with CSF compatible with the pattern of empty sella [Figure 2a-d, f, and g]. There was another mass with signal intensity compatible with mixed CSF and glial tissue, protruding into the sella from the lateral recess of the sphenoid sinus. This mass contained tissue resembling extension of glial tissue of the temporal lobe extending into the cyst [Figure 2a-c and e]. Metrizamide CT cisternography identified an intrasphenoidal meningocele located in the left side of the sinus resembling persistence of open Sternberg’ canal connecting the subarachnoid cistern of the medial temporal region into the cyst [Figure 2h]. Endoscopic endonasal approach to the sella revealed the CSF containing cyst hanging into the sphenoid sinus and fibr-oglial tissue remnants within it. The overlying fibrous tissue, resembling the dura was coagulated and removed little by little along with the neurovascular glial tissue within it. At the end, there was an orifice located in the left lateral side of the sphenoid cavity. There was no communication between the cyst cavity and the sella tursica. The orifice could be packed with autogenous fat and covered by a pediculate mucosal flap. In the postoperative course, she was suffering from dry left eye. Rhinorrhea recurred a week after surgery. Lumbar drainage was established for 5 days and rhinorrhea stopped. She was asymptomatic during the first 6 months after surgery but CSF leakage recurred after 1 year. A lumbo-peritoneal shunt was placed and there has been no CSF leakage during the next 10 months follow up.

DISCUSSION

Incidence and review of literature
The incidence of congenital encephalocele is approximately 1 in 3000-5000 live births.[28] Trans-sphenoidal encephaloceles comprise less than 5% of all basal meningo-encephaloceles and has an estimated incidence of 1 in 70000 live birth.[16,17,19] We performed a comprehensive search in the available
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When there is sufficient pneumatization of the sphenoid sinus, the lateral craniopharyngeal canal of Sternberg may act as the site of origin of congenital meningocele or CSF fistula. Another alternative suggestion is that the defect is secondary to the cumulative effect of CSF pressure on naturally occurring defects in the floor of middle cranial fossa leading in a fistulous tract. Considering the presence of empty sella and recurrence of rhinorrhea after proper package of the sella, the last theory can explain development of the anomaly in our second case.

Clinical presentation

The clinical presentation of patients with trans-sphenoidal encephalocele is variable depending upon the age (i.e., the type described previously). Excluding the characteristic face of some of these patients with hypertelorism, the diagnosis can be delayed into adolescence or adulthood when an unexplained rhinorrhea, meningitis, endocrine dysfunction or progressive visual field deficit prompt evaluations leading in diagnosis. Both our cases presented with CSF leakage without history of trauma.

Paraclinical evaluations

MRI is essential in evaluation of trans-sphenoidal encephalocele to confirm the extent of lesion, the possible associated abnormalities and to plan for the safest approach. MR angiography is helpful to evaluate intracranial vasculature. MRI with contrast material injection can evaluate the content of cyst. CT scan can visualize the bone defect in the skull base.

We used almost all the mentioned evaluations in our cases, which were quite sensitive regarding the endoscopic approach undertaken in both cases [Figures 1 and 2].
Treatment

The indications for treatment and choice of surgical approach for trans-sphenoid encephaloceles remain controversial. The main indication for intervention are: Obstruction of respiratory pathway, repeated meningitis, rhinorrhea, and progressive visual deficit attributable to the lesion.[31]

The management of the trans-sphenoidal encephalocele require a multidisciplinary approach.[17,22,24] Trans-sphenoidal encephalocele have been treated by the trans-cranial, trans-palatal, and trans-sphenoidal approaches. Trans-cranial approach is associated with a high rate of postoperative hypothalamic pituitary dysfunction.[9,21] Endoscopic route has been effectively used in the treatment of encephaloceles, which did not contain neural tissue.[19,23,25] Determination of the ideal approach is based on various factors including the degree of lateral sphenoid pneumatization, location and size of the meningo-encephalocele and the ability to perform adequate skull base repair through a given exposure. We succeeded in treating both of our cases, which contained small amount of neural elements using endoscopic technique.

Repair of persistent CSF leakage is the major indication for surgery in intrasphenoidal encephalocele. Trans-sphenoidal or trans-ethmoidal approaches must be decided individually depending upon the exact location of the defect in skull base diagnosed by the precise preoperative imaging and the experience of the surgeon.[9]

Regarding the delicate points worthy to be considered in the technique of surgery, in median perisellar lesions such as our case 1, a trans-nasal, trans-oral route or trans-oral trans-palatal route is safe. In encephalocele within the lateral recess of the sphenoid sinus, that is, coming through the Sternebgs’s canal, the endoscopic trans-pterigoid approach can be a better passage than the traditional trans-nasal and trans-ethmoid avenue.

CONCLUSION

In asymptomatic patients with true trans-sphenoidal encephalocele, or in patients with endocrine disorders or stable visual deficits, indications for operative repair are less clear. In symptomatic patients, the surgical approach must be tailored according to the individual patient’s age, anatomical characteristics and associated anomalies.[21] The ideal surgical approach for trans-sphenoidal encephalocele is still not clear due to lack of adequate experience in the literature. It can be concluded from our review that adult patients with trans-sphenoidal encephalocele should be considered for trans-nasal trans-sphenoidal endoscopic approach.[30]
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