Endoscopic endonasal resection of congenital trans-sphenoidal meningoencephalocele with extension to the epipharynx in early childhood: a case report

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Abstract
Background: The clinical presentation of sphenoid sinus meningoencephaloceles (MEC) may have insidious onset and evolution. Contemporary treatment incorporates endoscopic resection via the endonasal route.
Case description: We present a case of 3 year old girl who had been complaining of permanent nasal discharge, impeded nasal breathing and difficulty with feeding since she was 5 months old. There was no history of rhinoliquorrhea. Pre-operative magnetic resonance imaging demonstrated MEC that extended from the sellar region through the non-pneumatized sphenoid sinus to the nasopharynx. The lesion was resected via endoscopic endonasal approach. Follow-up rhinoscopy confirmed the absence of post-operative cerebrospinal fluid leak.
Conclusion: Endoscopic endonasal approach can be an effective and a safe treatment option for resection of congenital trans-sphenoidal MEC in early childhood.
Keywords: Sphenoid sinus; endoscopic endonasal approach; meningoencephalocele; CSF leak; congenital.
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Introduction
Meningoencephalocele (MEC) is a congenital malformation that is almost evenly distributed between males and females with slight preponderance to the female gender. MEC is a saccular herniation of meningeal and/or brain tissue through a circumscribed cranial defect.[1] Skull base MEC with expansion to the paranasal sinuses represent only 10% of all cases and its prevalence worldwide approximates 1 per 35 000 – 40000 neonates.[2] Usually, they present with spontaneous rhinoliquorrhea or other complications including obstruction of nasal airways, persistent nasal discharge, meningitis, etc. More than 50%of basal MEC have ethmoidal expansion while those protruding to the sphenoid sinus are rare.[3]
The surgical treatment of these lesions can be challenging and includes a variety of approaches – transcranial, lateral rhinotomy, sublabial, trans-septal and endonasal. Recently, the endonasal approach has become a standard treatment option in adult patients with similar lesions of the anterior cranial fossa.[4] This surgical route can be extremely difficult in pediatric patients due to anatomic and physiological restrictions such as the narrow nasal cavity

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and variable degree of aeration of the paranasal sinuses.[4,5]

Case report

We present a case of 3 year old girl who had been complaining of permanent nasal discharge, impeded nasal breathing and difficulty with feeding since she was 5 month old. There was no history and clinical data of rhinoliquorrhea. Her somatic and neurological status were normal.

The preoperative magnetic resonance imaging (MRI) showed MEC that extended from the sellar region through the non-pneumatized sphenoid sinus to the nasopharynx (Fig. 1).

The lesion was accessed via endoscopic endonasal approach. The sac was removed to the site of its entrance into the non-pneumatized sphenoid sinus. We performed plastic closure of the dural defect with fibrin glue and nasal mucosa harvested from the inferior nasal concha (Fig. 2A-F). The histological examination was consistent with MEC (Fig. 2G).

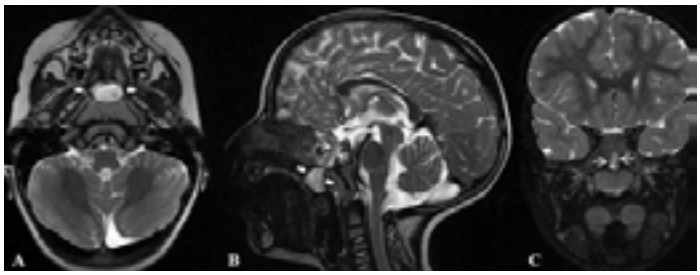

Fig. 1: Preoperative T2-weighted MRI: A) Axial view - saccular formation protruding to the epipharynx (thick arrows); B) Sagittal view – trunk (thin arrows) and sac (thick arrows) of the malformation; C) Coronal view – trunk of the MEC (thin arrows).

The postoperative follow-up period at 6 months was uneventful. Rhinoscopy confirmed proper epithelialization of the plastic closure of the skull base defect with no evidence of CSF leak. Follow-up MRI demonstrated total removal of the MEC sac (Fig. 2H).

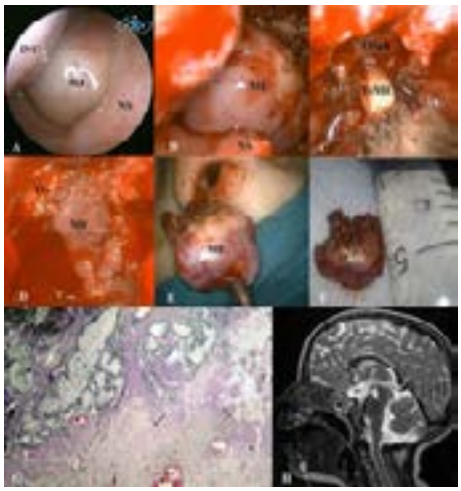

Fig. 2. A-F) Intraoperative images demonstrating different stages of the endoscopic resection of the MEC: INC – inferior nasal concha; – meningoencephalocele; NS – nasal septum; Osph – sphenoid bone; TrME – trunk of meningoencephalocele; TG – fibrin-tissue glue; - mucosal flap; G) Photomicrograph (H&E, x100) of the MEC specimen – submucosal epipharyngeal tissue with small mucous glands (black&white arrow), brain tissue (black arrow) and meningeal tissue (double black arrows); H) Postop sagittal T2-weighted MRI showing resection of the MEC sac with restoration of the normal passage of the nasal airways

Discussion

Congenital and acquired defects of the sphenoid bone (SB), including osteitis, excessive aeration, focal bone erosions and MEC are usually associated with intra-extracranial fistulas with low or normal intracranial pressure.[6] The knowledge of the ontogenetic development of the SB is important to understand the mechanism of formation of the non-traumatic congenital or acquired defects of the skull base. During embryological development, independent cartilaginous precursors are formed separately to become, after the ossification period, particular parts of the SB: presphenoid (anterior sphenoid bone), basisphenoid

(posterior sphenoid bone), orbitosphenoid (lesser wings), and alisphenoid (greater wings, lateral parts of the pterygoid process). Only the medial plate of the pterygoid process is built up by membranous ossification.[7] The SB is formed by multiple ossification centers that are initially separated by synchondroses. The fusion of these parts is a complex process and, if impaired, can lead to different bone defects.[8] Of major importance is the fusion of central with lateral parts that occurs after birth. Only a weak cartilaginous connection between central and lateral parts has been found in neonates' bones. Thus, the fusion plane creates a line of lesser resistance within the sphenoid bone. Ossification of this cartilaginous connection starts anteriorly and progresses posteriorly. Before final fusion, a small canal connecting the middle cranial fossa with the nasopharynx is created, called the craniopharyngeal canal (Sternberg's canal). This canal extends intracranially from the sphenoid bone body and the posterior roof of the lesser wing and goes downwards on the side of the sphenoid bone body into the nasopharynx, where it ends at the level of the vaginal process, near the vomerovaginal canal.[7] Persistent Sternberg's canal can be found in approximately 4% of adults.[2] This fact imposes the idea that the defects of the SB at the fusion sites are actually congenital.[9] The physiological dynamics of the intracranial pressure can cause enlargement of the dural and bone defects. Therefore, this can explain the expansion of these defects to the lateral recesses of the sphenoid sinus (16-27% of adults have well-developed lateral recesses through which brain herniation and CSF leak can take place).[10]

One study reported that there were 58 cases with sphenoid MEC with mixed etiologythat had been published until 2011. In most cases, the defect was localized in the lateral and posterolateral areas of the sphenoid sinus with preponderance to the female gender.[3] Blaivie et al. stated that their case of trans-sphenoid MEC was the second of congenital origin described in literature after those of Schick et al., as in both cases there was persistent Sternberg's canal.[2,11] Our case is also a clear example of congenital transsphenoidal MEC but here it was located strictly in the midline.We speculate that the midline location here is related to persistent transsphenoidal canal in combination with changes that probably occur concurrently with the formation of the pituitarygland and faulty subsidence ofthe Rathke'spouch.

Several authors share that the basic principles of endoscopic endonasal surgery in adults cannot be directly applied in the pediatric population.[4,5,12] The small dimensions of the nasal cavity in early childhood impede the free movements of the endoscope and the surgical instruments. This can make difficult the presentation of the bone defect area. In cases where the endonasal access is difficult, some authors recommend the utilization of the sublabial surgical corridor.[12] The absence of pneumatization of the sphenoid sinus can mislead the anatomical orientation of the surgeon. Therefore, the endoscopic endonasal surgery in early childhood can be extremely challenging.[4] Aggressive removal of bony and/or cartilaginous structures in children during endoscopic endonasal procedures may affect future development of this anatomical region, but compared to other approaches, this theoretical risk is relatively low.[13]

Conclusion

Sphenoid MEC and other congenital malformations of the skull base continue to be therapeutic challenge. Our case illustrates that endoscopic endonasal surgery can be effective and safe treatment option for such lesions in early childhood that has certain advantages over conventional extracranial or intracranial approaches.

Conflict of interest

None declared.

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