Polish Annals of Medicine

Journal homepage: https://www.paom.pl

Case Report

Congenital transsphenoidal meningocele in a newborn: A case report

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Article info

Article history
Received: November 28, 2023
Accepted: December 20, 2023
Available online: February 26, 2024

Keywords
Meningocele
Cleft palate
Congenital
Hypertelorism
Nasal mass
Trans-sphenoidal

Doi
https://doi.org/10.29089/paom/177338

Abstract

Introduction: Congenital transsphenoidal meningocele is extremely rare.

Aim: We present a newborn who was found to have a large midline cystic mass protruding from a cleft palate.

Case study: Radiological imaging showed a large cerebrospinal fluid mass herniating through a large anterior cranial fossa defect into the sphenoid sinus with extension to the nasal cavity through a cleft palate into the oral cavity.

Results and discussion: A bifrontal craniotomy and decompression of meningocele with dura repair was performed.

Conclusions: The child was discharged healthy postoperatively and on monthly follow-up and subsequently thriving well with improving neurological recovery.

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1. INTRODUCTION

The incidence of cephaloceles which includes meningocele and encephaloceles is assumed to range between 1 in 40,000 live births.[1] Cephaloceles are generally classified based on their site of origin such as occipital, sincipital or basal.[2] Transsphenoidal meningocele is a subset of the basal type of cephalocele in which a fluid-filled sac of meninges protrudes through the bony sphenoid sinus into the nasal cavity without any neural tissue herniation. In general, the aetiology of meningocele is either congenital or acquired, the latter being more common and often due to trauma of the skull base. Congenital basal cephalocele on the other hand is often clinically occult until they result in life-threatening complications.[3] The sphenoid sinus is rarely involved in the site of cephaloceles and to our knowledge true transsphenoidal meningocele with extension into the nasal cavity detected at birth has not been previously reported.

2. AIM

We present a newborn who was found to have a large midline cystic mass protruding from a cleft palate.

3. CASE STUDY

A term newborn was delivered healthy by spontaneous delivery. Antenatally, the mother had mild anaemia in pregnancy. Upon examination, there was a palatal cleft with a soft smooth, cystic mass protruding into the oral cavity (Figure 1). This was associated with other mid-facial dysmorphism features including hypertelorism, low-set ears and wide nasal bridge. Cold spatula test revealed adequate misting bilaterally. Neurologically, the child had a weak oral suctioning reflex at birth but no evidence of limb hypotonia. There was also no evidence of proptosis or squint suggestive of orbital involvement. Magnetic resonance imaging (MRI) was performed showing a large transsphenoidal meningocele with

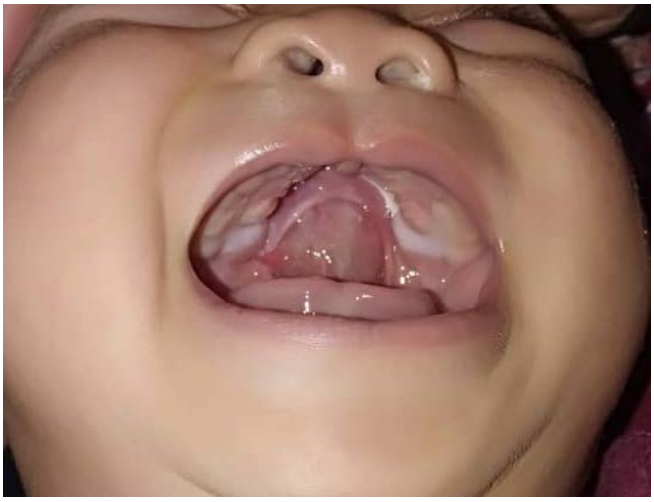

Figure 1. Intraoral examination reveled a U-shaped cleft palate with visible a large midline cystic mass (meningocele) protruding through the cleft palate.

hypoplasia of the corpus callosum (Figure 2). Otherwise, there was no sign of respiratory distress nor evidence of neurological deficit. The child was discharged well with a feeding tube. Other supportive examinations included an echocardiogram which revealed a patent foramen ovale with trivial tricuspid regurgitation and interestingly an auditory brainstem reflex revealed bilateral profound hearing loss which was. There were no signs of cerebrospinal fluid (CSF) leak and the child was monitored up to 1 year old.

The child underwent surgical intervention at 1 year old. Endoscopic examination (Figure 3) revealed this mass was located at the midline at the posterior nasal septum extending posteriorly into the nasopharynx, superiorly to a skull base defect, and anteroinferior into the oral cavity via a palatal defect. We had to abandon our initial choice of endoscopic repair and this was quickly converted to a bifrontal craniotomy approach, followed by a durotomy and retraction of both frontal lobes allowing visualization of the anterior skull base (Figure 4). The pituitary stalk was identified and the meningocele wall was seen beneath the stalk. The meningocele sac protruded inferiorly through a large skull

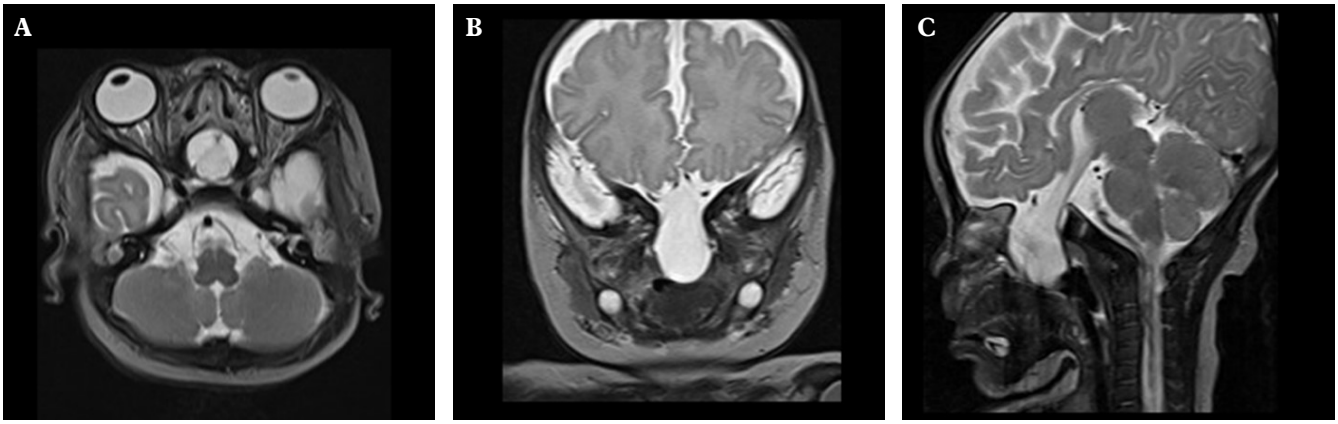

Figure 2. T2 Weighted MRI BRAIN: (A) axial cut showing hyperintense mass anterior to optic chiasm; (B) coronal cut showing hyperintense mass extending inferiorly into the nasal cavity; (C) sagittal cut showing homogenous hyperintensity extending up to the sellar floor.

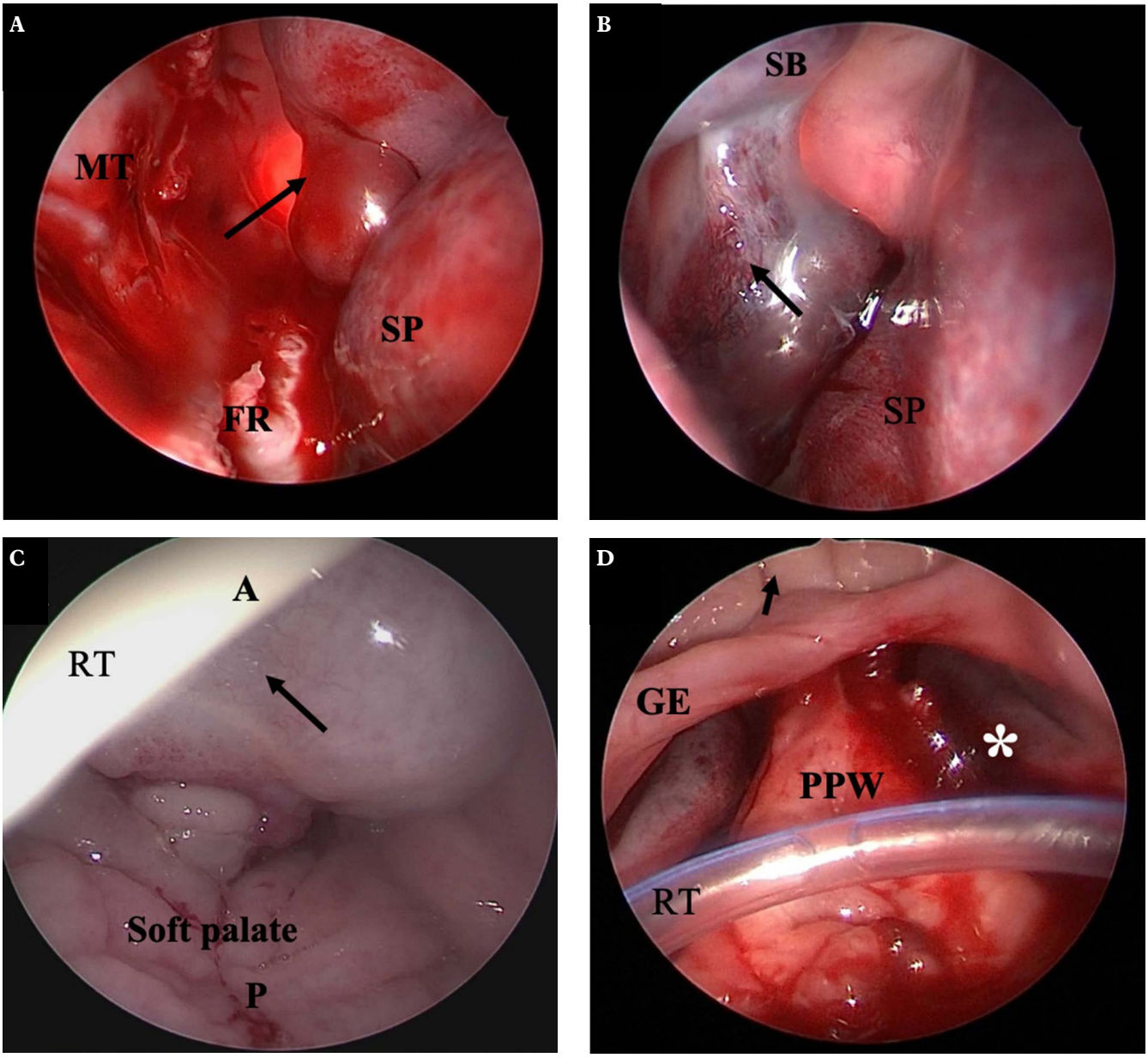

Figure 3. Endoscopic transnasal and per oral view: (A) Endoscopic transnasal view with middle turbinate (arrow indicates meningocele); (B) Endoscopic transnasal view with skull base; (C) Reverse endoscopic per oral view with scope viewing the palate superiorly (arrow indicates bulging of mass in the oral cavity in front of the soft palate); (D) Reverse endoscopic per oral view with scope viewing deeper to view the nasopharynx (* denotes the nasopharynx extending superiorly). Comments: MT – middle turbinate; FR – floor of nose, SP – anterior septal wall; SB – skull base superiorly, SP – anteroinferior septal wall; A – anterior; P – posterior; RT – feeding tube; GE – glossoepiglottic fold with absence of uvula in the middle; PPW – posterior phayrngeal wall.

base defect starting from the posterior planum sphenoidale expanding the entire width of the sellae turcica. The meningocele sac was decompressed by releasing the CSF which immediately resulted in a reduction of the size in the nasal and oral cavity. Following this, the sac wall was protracted through the defect cranially and proxied to the surrounding dura mater using Liga clip. A peri cranial fascial flap was harvested and reinforced using TISSEEL fibrin sealant above the clipped defect.

Postoperatively, the child was kept intubated and monitored in the intensive care unit for 3 days. He was allowed to feed on a feeding tube upon extubation, after which the child developed hyponatremia second to the salt-wasting syndrome. Apart from that, there was no evidence of a CSF leak. The child was discharged home 2 weeks after surgery. After 3 months of surgery and oral rehabilitation, the child was able to tolerate a soft diet with minimal nasal regurgitation. The child is further planned for palatal surgery and hearing rehabilitation.

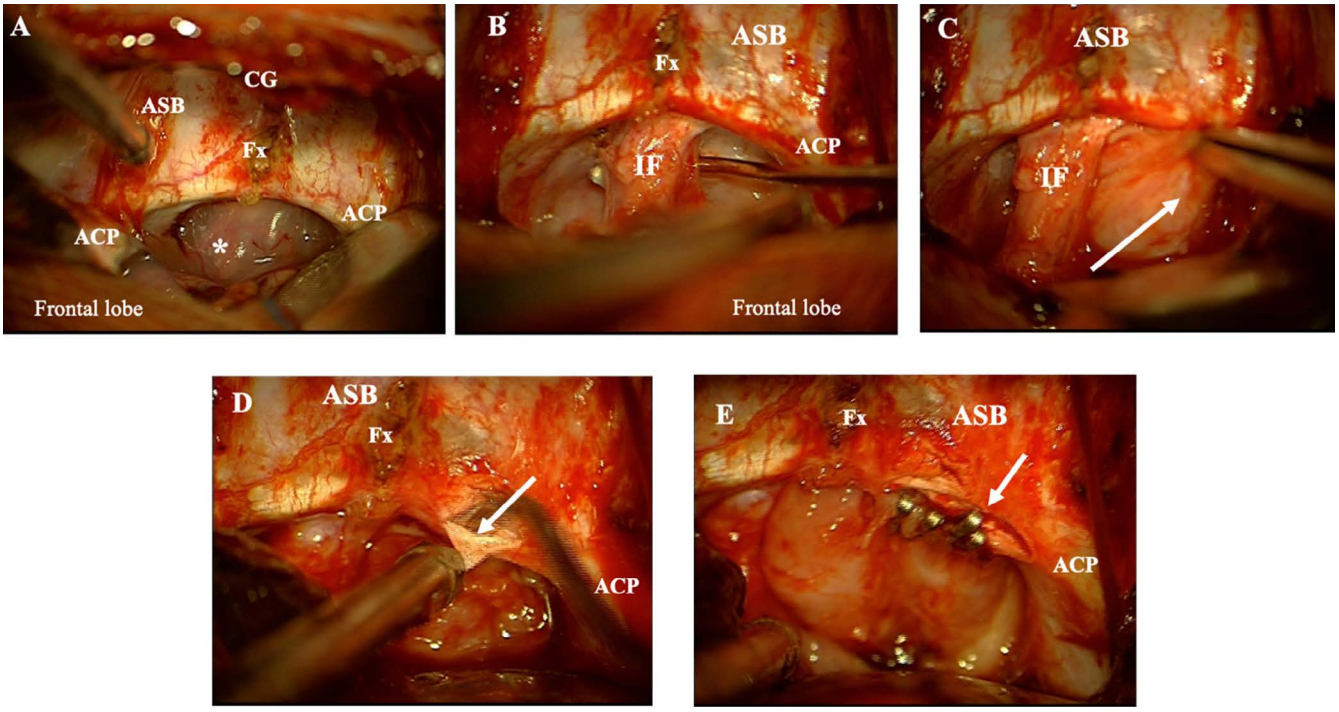

Figure 4. (A) View after bicoronal craniotomy and retraction of bilateral frontal lobe; (B) The infundibular stalk (IF) is located and preserved; C) Meningocele being lifted superiorly before being divided from the infundibular stalk (white arrow); D) Pericranial facsia harvested (white arrow); E) Sac wall protracted cranially and pexied with dural lining using Liga clip (white arrow). Comments: * skull defect spanning enterior width of sella; ASB – anterior skull base, CG – crista galli, Fx – sectioned falx cerebri, ACP – anterior clinoid process.

4. DISCUSSION

Knowledge of the ontogeny of the sphenoid bone is key to understanding the pathogenesis of a rare congenital form of skull base defect. In embryological development,[2] the sphenoid body is derived from precursors of the anterior sphenoid (presphenoid) and posterior sphenoid (basis sphenoid) centres. The bony fusion of the greater wings with the presphenoid and basisphenoid starts in its anterior portion while the posterior fusion can be incomplete creating a lateral craniopharyngeal canal.[2] This incomplete fusion was found consistently in children up to age 3–4 years old but only 4% in the adult.[3] Also known as the Sternberg's canal because it was first described by Maximilian Sternberg[4] it has been found either medial or lateral to the foramen rotundum. However, to date, no more than 36 cases have been reported of meningocele truly arising from this lateral canal.[5] Congenital transsphenoidal meningocele is thus even less described and to date, only 1 case was reported.[6] Our patient manifested with a huge meningocele in the midline extending into the oral cavity is a true transsphenoidal meningocele that could not be explained by Sternberg's canal alone. Some authors suggested that with the presence of this lateral Sternberg's canal, even normal intracranial pressure may cause an enlarged bony defect inducing brain herniation and CSF leak.[6] A cleft on the sphenoid bone or a partial absence of the dural lining of the skull base in the initial development has also been suggested to cause herniation of the meningeal sac into the nasopharynx.[7]

Trans-sphenoidal encephalocele occurs in about 1 in 700,000 live births.[8] True transsphenoidal meningocele is rarely detected early unless it results in life-threatening complications such as persistent rupture of meningocele during suctioning or meningitis.[9] In adults, they are reported to manifest as persistent CSF rhinorrhea[5] or persistent headaches.[10] In our case, a diagnosis of congenital meningocele is straightforward with the presence of midfacial anomalies including hypertelorism and broad nasal bridge. In our patient, embryological failure of palatal fusion is likely to be a secondary effect of such a large meningocele. Despite such a gross extension through the cleft palate, there were no incidences of CSF rhinorrhea nor any signs of meningitis before surgery. Despite a large tumour occupying the nasal cavity and oral cavity, our patient did not have any breathing difficulty which is a common feature of transsphenoidal meningocele.[11] Another feature worth noting is that most cases reported frequent absence of the posterior part of the septum. Similarly, in our case, we noticed that the meningeal sac extending posterior-inferiorly through the posterior septum was covered only by a thin layer of the nasal mucosa with the absence of the posterior bony septum.

Surgical treatment is the ultimate option for correction/repair of the meningocele. This is mandatory in order to prevent central nervous system infection with about 40%

long term risk of meningitis.[12] We initially planned for an endoscopic repair; however, this was abandoned for a few reasons. First, the superior aspect of the mass proved to be large and such a defect may not be adequately sealed by endoscopic technique alone. Second, there was very little room for manipulation of the sac as well as an instrument in the overly crowded nasal cavity. Third, despite our best effort to separate the meningocele sac from the nasal mucosa plane, we were not able to achieve a safe separating plane between them thus this gives rise to concern of iatrogenic perforation and risk of meningitis in an anticipated prolonged surgery. The endoscopic approach of anterior congenital cephalocele repair is currently preferred over open surgery due to its minimal invasiveness, safety and effectiveness as well as decreased postoperative complication.[13,14] Despite that, in the face of such a large defect, we believe that an open technique allowed more complete decompression of the meningocele, the superior advantage of ensuring a tight seal for the reconstruction of the anterior cranial defect as well as reduced operative time and risk of meningitis. The bi-coronal approach also provides a good aesthetic effect.[15] However, some studies report morbidity of up to 70% and mortality of 50%.[16]

In dealing with the relatively smaller lateral wall of the sphenoid meningocele, endoscopic endonasal surgery can be a method of choice because it provides direct, extracranial access to the lesion and does not require manipulation of the brain.[17] However, this method is very challenging in newborns and infants who have smaller nasal cavities and more difficult access.[18] Hoff et al. had reported the transpalatal approach which allows for access through oral cavity and allow for combined team approach surgery with neurosurgery and otolaryngology.

As for the reconstruction material, Zoli et al. published good outcomes of multilayer closure with facia lata, mucoperiosteum and bone or fat with the nasoseptal flap. None of the reported patients developed neurological sequelae and no recurrent issue of CSF leak or seizures.[17]

5. CONCLUSIONS

Large transsphenoidal meningocele appearing in the oral cavity has not been previously reported. Despite its intimidating size, our child has undergone open surgery uneventfully and postoperatively appeared to have improved neurological performance.

Conflict of interest

No conflict of interest declared.

Funding

None declared.

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