Transalar Sphenoidal Encephalocele and Respiratory Distress in a Neonate: A Case Report

ABSTRACT. We present a full-term newborn infant who suffered from immediate postpartum severe respiratory distress. The infant had an inspiratory stridor as a result of a swelling of the soft palate, extending from the roof of the nasopharynx. Transoral endotracheal intubation resulted in normal saturation levels. Histologic examination after an open biopsy showed mature neuroglial tissue. Radiology demonstrated the presence of a right parapharyngeal process obstructing the nasopharynx and oropharynx and extending to the right middle and posterior fossa, via the foramen ovale. After transoral debulking, the infant was extubated successfully. After an uneventful period of 5 months, the patient was readmitted at our hospital for treatment of meningitis. Subsequently, the inspiratory stridor recurred, and staged surgery was performed. First, a transcranial approach was used to remove a large intradural part of the process and close the defect at Meckel’s cave. Two weeks later the retro- and parapharyngeal part of the process were removed transorally. Given the site of the defect of the skull base and the intradural location of the process, the diagnosis is a transalar sphenoidal encephalocele. This is a rare type of basal encephalocele, and has never been reported in an infant nor known to present with respiratory distress. The infant had an inspiratory stridor as a result of a swelling of the soft palate, extending from the roof of the nasopharynx to the level of the base of the tongue, and into the soft palate and the tonsillar region. An open biopsy was performed. Histologic examination showed mature neuroglial tissue. Thus a malignancy was excluded, and a debulking was planned. On the third day of life, an examination was performed under general anaesthesia. The swelling extended from the roof of nasopharynx to the level of the base of the tongue, and into the soft palate and the tonsillar region. An open biopsy was performed. Histologic examination showed mature neuroglial tissue. Thus a malignancy was excluded, and a debulking was planned. On the 29th day of life, an examination was performed under general anaesthesia. The swelling extended from the roof of nasopharynx to the level of the base of the tongue, and into the soft palate and the tonsillar region. An open biopsy was performed. Histologic examination showed mature neuroglial tissue. Thus a malignancy was excluded, and a debulking was planned. On the 37th day of life. Because no additional episodes of respiratory distress occurred, it was decided to follow an expectant course as long as possible to allow normal craniofacial development. The patient was discharged from our hospital. During the next few months, no problems occurred. Intraoral inspection at the outpatient clinic at 4 and 5 months did not show growth of the lesion. At 5 months, however, the patient failed to thrive as a result of pathologic reflux, coughing spells, and subsequent vomiting. The reflux was treated medically with ranitidine and cisapride, and a nasogastric feeding tube was installed. At 6 months, the patient developed fever, convulsions, and a lower level of consciousness and was admitted to a regional hospital. Artificial ventilation was needed because of apneas. A culture of the CSF and a swab of the pharynx revealed Streptococcus pneumoniae. The meningitis was treated successfully with penicillin IV. At 7 months, the patient was readmitted to our hospital because of the reappearance of an inspiratory stridor, progressive snoring, and serious episodic oxygen desaturations during sleep. On examination of the pharynx, the swelling of the soft palate and right lateral pharyngeal wall was found to have progressed mildly. A nasal Mayo tube was installed, securing the airway. Multidisciplinary discussion resulted in the following staged surgical strategy.

CASE DESCRIPTION

After an uneventful first pregnancy of 42 ± 1 weeks, a boy weighing 3350 g was born to nonconsanguineous parents. Delivery had been spontaneous, with Apgar scores of 8 and 9 after 1 and 5 minutes, respectively. The infant had a normal appearance except for a slightly asymmetrical face. However, immediately postpartum, the infant developed an inspiratory stridor. On the first day of life, multiple episodes of oxygen desaturation occurred. Examination of the oral cavity and oropharynx revealed a swelling of the soft palate, blocking direct vision of the posterior wall of the oropharynx. The infant was transferred to our hospital and admitted to the neonatal intensive care unit. Transoral endotracheal intubation was performed, resulting in normal saturation levels without the need for artificial ventilation or oxygen administration. Physical examination did not reveal any other congenital malformations. Extensive endocrinologic screening for hypothalamic-pituitary dysfunction showed no abnormalities. In the first week of life, at the neonatal intensive care unit, central apneas occurred on several occasions. Results of electroencephalography and BAEP; thorax and cervical spine x-ray examination; and brain, heart, and kidneys ultrasound examination were normal.

On the second day of life, magnetic resonance imaging (MRI; Siemens Magnetom 1.5T system, Erlangen, Germany) (spin-echo, T1-weighted sagittal sections and spin-echo proton-density, T2-weighted transverse sections) revealed a large right parapharyngeal process (Fig 1). The process obstructed the nasopharynx and oropharynx almost completely and extended to the right middle and posterior cranial fossa, most probably via the foramen ovale (Fig 1). Coronal, transverse, and sagittal images showed a right posterior retrosellar location of the intracranial part of the process, primarily in an extradural location. MR angiography depicted the parasellar internal carotid artery in normal position, medial to the process. The cerebrum had a normal appearance. In all sequences, computed tomography (CT) demonstrated a bony defect of a diameter of 10 mm at the site of the foramen ovale. The pterygoid process was deformed and placed in frontal direction (Fig 2).

On the third day of life, an examination was performed under general anaesthesia. The swelling extended from the roof of nasopharynx to the level of the base of the tongue, and into the soft palate and the tonsillar region. An open biopsy was performed. Histologic examination showed mature neuroglial tissue. Thus a malignancy was excluded, and a debulking was planned. On the 29th day of life, after a midline incision of the soft palate, a partial resection of a cystic lesion was performed. Analysis of aspirated fluid did not reveal cerebrospinal fluid (CSF)-specific β2-transfetin. Postoperative inspection of the pharynx demonstrated enough space, and the patient was extubated successfully on the 37th day of life. Because no additional episodes of respiratory distress occurred, it was decided to follow an expectant course as long as possible to allow normal craniofacial development. The patient was discharged from our hospital. During the next few months, no problems occurred. Intraoral inspection at the outpatient clinic at 4 and 5 months did not show growth of the lesion. At 5 months, however, the patient failed to thrive as a result of pathologic reflux, coughing spells, and subsequent vomiting. The reflux was treated medically with ranitidine and cisapride, and a nasogastric feeding tube was installed. At 6 months, the patient developed fever, convulsions, and a lower level of consciousness and was admitted to a regional hospital. Artificial ventilation was needed because of apneas. A culture of the CSF and a swab of the pharynx revealed Streptococcus pneumoniae. The meningitis was treated successfully with penicillin IV. At 7 months, the patient was readmitted to our hospital because of the reappearance of an inspiratory stridor, progressive snoring, and serious episodic oxygen desaturations during sleep. On examination of the pharynx, the swelling of the soft palate and right lateral pharyngeal wall was found to have progressed mildly. A nasal Mayo tube was installed, securing the airway. Multidisciplinary discussion resulted in the following staged surgical strategy.

At the age of 7 months, surgery was performed by the neurosurgeon. After a right frontotemporal trepanation with inspection of the subtemporal dura, the cisternal section of the trigeminal nerve was found to be stretched as a result of an intradural...
beige-colored process in the posterior cranial fossa, just caudal to
the trigeminal nerve. After incision of the tentorium and limited
debulking of the relatively soft tissue, it was decided to discon-
tinue the operation and approach the process via the suboccipital
route for optimal exposure. Intraoperative histopathologic exam-
ination had shown the presence of glial tissue, similar to that
obtained by the earlier transoral route.

Two weeks after the frontotemporal trepanation, a suboccipital
trepanation was performed. Subsequently, the process of
2 by 3-cm situated anterior and caudal to the trigeminal nerve was
removed. The process was adherent to both the trigeminal and the
abducens nerve. Some of the branches of the trigeminal nerve
appeared to transverse the process. The lesion was clearly located
intradurally, in the subarachnoidal space adjacent to the brain-
stem. The arachnoidea and pia of the brainstem were intact. The
process looped around the internal carotid artery. A large dural
defect was found at Meckel’s cave, through which the process
continued. After removal of the process the defect was closed with
a free galea peristium flap and fibrin glue (Tissucol, Immuno
A.G. Vienna, Austria). A right abducens paresis was present post-
operatively despite the intraoperative anatomic preservation of
the nerve.

After 2 weeks, the infant underwent surgery again, and this
time the process was approached transorally to remove the retro-
and parapharyngeal part. A midline incision of the mucosa at the
soft palate was extended to the right lateral pharyngeal wall. The
process was identified, mobilized, and followed into the naso-
pharynx. Sharp dissection high in the nasopharynx resulted in
some residual tissue at the skull base. There were no postoperative
complications.

Histologic examination of both the extracranial and the intra-
cranial parts of the lesion revealed mature glial tissue with focal
neurons and dispersed calcifications. Immunohistochemically, the
glial tissue showed strong positivity for a glial marker (mono-
clonal antibody glial fibrillary acidic protein, gift of Dr Van Muyen,
Nijmegen, The Netherlands, dilution 1:20), whereas the neural
perikarya and neurites were variably but often strongly positive
for neuronal markers (monoclonal neurofilament, gift of Dr Van
Muyen, dilution 1:10; polyclonal antibody synaptophysin, Dako-
patt, Kopenhagen, Denmark, dilution 1:100). Focally, the neuro-
glial tissue showed a somewhat laminar architecture of gray and
white matter. The extracranial part was intermingled with fibrous
tissue and partly covered with pharyngeal mucosa (Fig 3).
Ependymal lining, choroid plexus, and meningeal layers were
absent.

The patient was discharged from our hospital 4 weeks after the
operation. In the 3-month follow-up period, there was a gradual
and complete disappearance of the abducens paresis. There were
no more respiratory problems. The patient has resumed eating
and has gained weight according to his age, and his cognitive and
motor development is normal.

DISCUSSION

Basal Encephaloceles

Mature neural tissue in the nasopharynx or
pharynx of a neonate may be encountered in the
context of an encephalocele, a cerebral heterotopia,
or a teratoma. An encephalocele is a developmen-
tal anomaly defined as a herniation of neural tissue
through a bony defect in the skull. Cerebral hetero-
topia is mature neural tissue encountered out-
side of the subarachnoidal space.1 In our case, a
teratoma was unlikely because teratomas consist of
at least two germ layers and intracranial commu-
nication is rare.2 The most common site of an en-
cephalocele is in the midline at the occiput. In the
Western world, ~75% of the encephaloceles are occipital; 15% sincipital, presenting at the dorsum of the nose, orbit, or forehead; and 10% basal. A basal encephalocele is a rare finding, occurring in 1 in 40,000 live births. The more posterior the basal encephalocele, the less frequent it is. According to the site of herniation, the basal encephalocele is located in the nasal cavity (transethmoidal and sphenoethmoidal type), in the orbit (sphenoorbital type), in the sphenoid sinus or nasopharynx (sphenoethmoidal and transsphenoidal type), or in the sphenomaxillary fossa (sphenomaxillary type). In the literature, only four cases with a transalar sphenoidal encephalocele have been described in which the lesion protruded through the greater wing of the sphenoid bone, and two cases with lesions at the petrous apex. All cases with transalar lesions involved adults, with large defects and protruding meningoencephaloceles including the foramen ovale, in at least two cases.

Pathogenesis
In the pathogenesis of encephaloceles, a teratogen, as well as a genetic factor, is thought to be present.

Clinical Presentation
Symptoms as a result of a basal encephalocele vary according to the site and size of the lesion and develop in the neonatal and early infantile period in the majority of patients. The common presentation is that of a mass, usually in the midline. Airway obstruction may result from lesions in the nose, nasopharynx, and oropharynx. Consequently, respiratory distress, episodes of apnea, difficulty with feeding, and failure to thrive are seen. Severe respiratory distress such as that seen in our patient is associated with pharyngeal airway obstruction. CSF rhinorrhea and recurrent meningitis may occur. Late symptoms may be visual disturbance and pituitary-hypothalamic dysfunction. Basal encephaloceles, especially transsphenoidal encephaloceles, are associated with other congenital malformations such as hypertelorism, cleft lip and/or palate, and optic and brain anomalies. In our patient, no other malformations were present. The documented cases of transalar sphenoidal encephaloceles were associated with nonlocalizing symptoms such as seizures and headache. Two patients with lesions at the petrous apex suffered recurrent meningitis. One patient...
with a transalar sphenoidal lesion had symptoms of trigeminal neuralgia.8

Pathology

Microscopically, encephaloceles consist of nonneoplastic mature neuroglial tissue, often with reactive changes. A varying degree of organization is found, and meninges may be present. Differentiation between a true encephalocele and cerebral heterotopia may be difficult. In the absence of meninges, it is not possible to differentiate based on histopathology alone. To diagnose an encephalocele, CT with or without contrast in the CSF and, particularly, MRI or surgery are necessary to prove cerebral connection.5,11,12 In our patient, CT unequivocally demonstrated the bony skull base defect. An intradural continuation of the process was suspected on MRI and confirmed by surgery. Knowledge about the presence of an intradural communication is of importance.

Therapeutic Implications

Reviewing the literature of the posteriorly located basal encephalocele, Yokota and colleagues state that operative indications and approaches are controversial.5 Surgery is even considered contraindicated by some because in transsphenoidal encephaloceles, vital structures such as the hypothalamic-pituitary system and anterior cerebral arteries may be included in the herniated brain tissue. Progressively symptomatic basal encephaloceles should be operated at an early stage, either by a transcranial or a combined approach.3,5,6 The goal of surgery is to resect the encephalocele with minimal damage to vital brain tissue and to achieve closure of the defect in the dura and large bony defects. A solely transpalate approach is rarely sufficient.4 In the reported cases of transalar sphenoidal encephaloceles, two patients were treated surgically using a transcranial approach alone. As the pharyngeal component of the lesion was so extensive, in our patient the combined approach was the only option. Closure of the dural defect, imperative as a preventive measure against recurrent meningitis, would have been impossible using the transoral approach alone.

In conclusion, this case is unique because it concerns an infant with severe respiratory problems from the time of birth as a result of a transalar sphenoidal encephalocele. Only four cases of transalar sphenoidal encephalocele have been reported in the literature. On the preoperative MRI in our patient, a primarily extradural process was seen and an intradural component was suspected. The histopathologic findings could fit an encephalocele as well as a cerebral heterotopia. The occurrence of meningitis supposed the presence of an encephalocele, yet the final diagnosis was made only after surgery. The multidisciplinary approach to the management of this patient, including pediatrics; oral and maxillofacial surgery; neurosurgery; pathology; radiology; and otorhinolaryngology, was essential. The combined-approach surgery is appropriate for these extensive lesions and, in this patient, proved successful.

Emmanuel A.M. Mylanus, MD, PhD
Henry A.M. Marres, MD, PhD
Department of Otorhinolaryngology

José Vlietman, MD
Louis A.A. Kollée, MD, PhD
Department of Pediatrics

Hans Peter M. Freihofer, MD, DMD, PhD
Department of Oral and Maxillofacial Surgery

Henk O.M. Thijssen, MD, PhD
Department of Neuroradiology

Joost de Vries, MD, PhD
Department of Neurosurgery

Pieter Wesseling, MD, PhD
Departments of Pathology and Neurology
University Hospital Nijmegen
6500 HB Nijmegen, The Netherlands

REFERENCES
# Transalar Sphenoidal Encephalocele and Respiratory Distress in a Neonate: A Case Report

Emmanuel A.M. Mylanus, Henry A.M. Marres, José Vlietman, Louis A.A. Kollée, Hans Peter M. Freihofer, Henk O.M. Thijssen, Joost de Vries and Pieter Wesseling

*Pediatrics* 1999;103:e12

<table>
<thead>
<tr>
<th>Updated Information &amp; Services</th>
<th>including high resolution figures, can be found at: /content/103/1/e12.full.html</th>
</tr>
</thead>
<tbody>
<tr>
<td>Subspecialty Collections</td>
<td>This article, along with others on similar topics, appears in the following collection(s): <strong>Fetus/Newborn Infant</strong> /cgi/collection/fetus:newborn_infant_sub <strong>Pulmonology</strong> /cgi/collection/pulmonology_sub</td>
</tr>
<tr>
<td>Permissions &amp; Licensing</td>
<td>Information about reproducing this article in parts (figures, tables) or in its entirety can be found online at: /site/misc/Permissions.xhtml</td>
</tr>
<tr>
<td>Reprints</td>
<td>Information about ordering reprints can be found online: /site/misc/reprints.xhtml</td>
</tr>
</tbody>
</table>

PEDIATRICS is the official journal of the American Academy of Pediatrics. A monthly publication, it has been published continuously since 1948. PEDIATRICS is owned, published, and trademarked by the American Academy of Pediatrics, 141 Northwest Point Boulevard, Elk Grove Village, Illinois, 60007. Copyright © 1999 by the American Academy of Pediatrics. All rights reserved. Print ISSN: 0031-4005. Online ISSN: 1098-4275.
Transalar Sphenoidal Encephalocele and Respiratory Distress in a Neonate: A Case Report
Emmanuel A.M. Mylanus, Henry A.M. Marres, José Vlietman, Louis A.A. Kollée, Hans Peter M. Freihofer, Henk O.M. Thijssen, Joost de Vries and Pieter Wesseling

The online version of this article, along with updated information and services, is located on the World Wide Web at:
/content/103/1/e12.full.html