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 pathogenesis, clinical presentation, pathology, and therapeutic implications of basal encephaloceles are discussed. *Pediatrics* 1999;103(1). URL: http://www.pediatrics.org/cgi/content/full/103/1/e12; *basal encephalocele, respiratory distress, inspiratory stridor, meningitis.*

ABBREVIATIONS. MRI, magnetic resonance imaging; CT, computed tomography; CSF, cerebrospinal fluid.

This article reports a pharyngeal mass consisting of mature neuroglial tissue causing life-threatening respiratory distress and meningitis in a full-term newborn infant. A combination of clinical, radiologic, and pathologic findings led to the diagnosis and transcranial and transoral surgical treatment of a transalar sphenoidal encephalocele. A multidisciplinary approach was essential in the successful management of the patient.

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 oc-

curred. 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 para- and retrosellar position 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 with a diameter of 10 mm at the site of the foramen ovale. The pterygoid process was deformed and displaced in frontal direction (Fig 2).

On the third day of life, an examination was performed under general anesthesia. 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-transferrine. 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
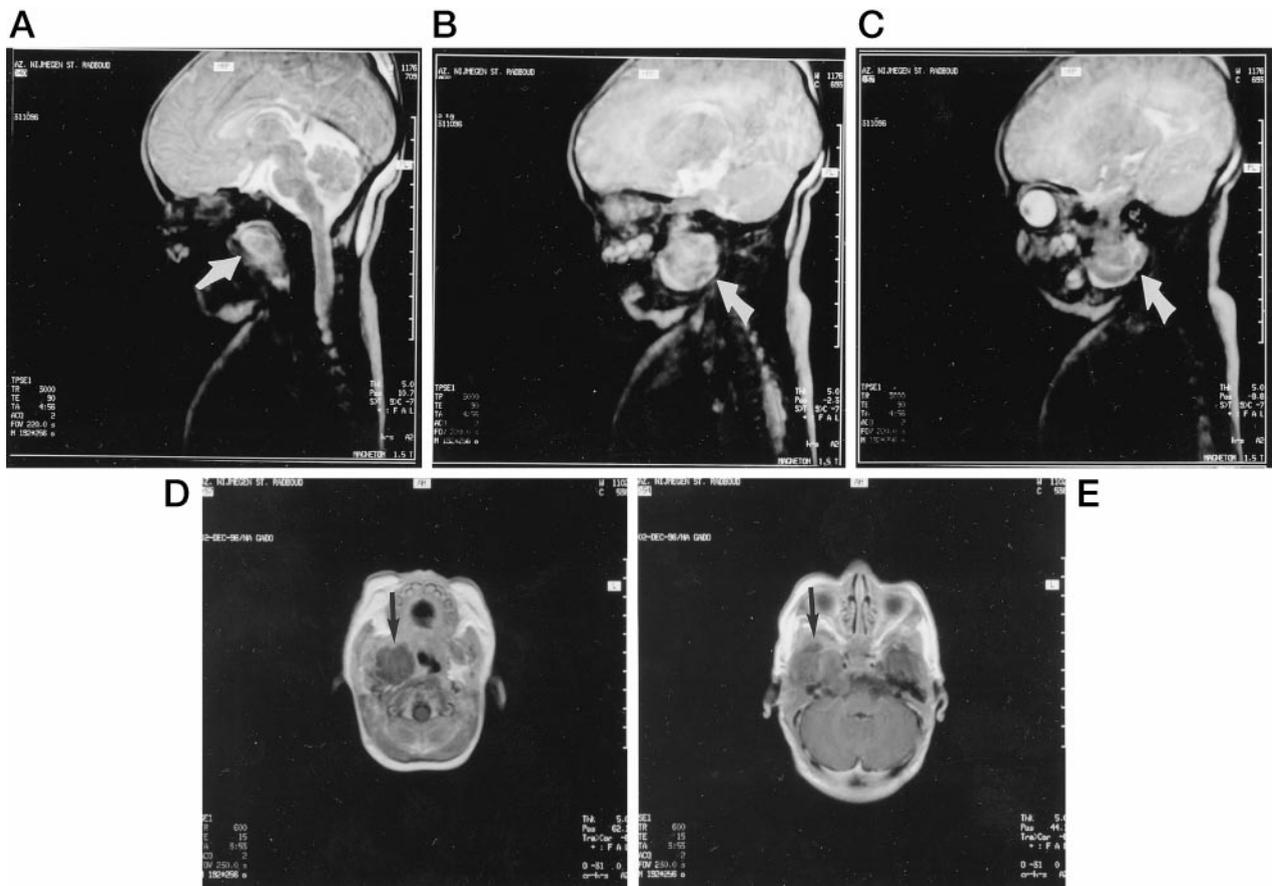
Fig 1. A–E. Sagittal (T2W) and transverse (T1W) MRI. A, pharyngeal tumor (arrow), normal intracranial midline structures; B, right parapharyngeal tumor (arrow) extending into the middle and posterior cranial fossa through a bony defect in the floor of the middle fossa; C, right parapharyngeal tumor (arrow) extending to the parasellar region; D, right parapharyngeal tumor (arrow); E, parasellar tumor (arrow) extending to the posterior fossa.

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 discontinue the operation and approach the process via the suboccipital route for optimal exposure. Intraoperative histopathologic examination 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 brainstem. 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 periosteum flap and fibrin glue (Tissucol, Immuno A.G. Vienna, Austria). A right abducens paresis was present postoperatively 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 nasopharynx. 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 intracranial 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 (monoclonal 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 developmental anomaly defined as a herniation of neural tissue through a bony defect in the skull. Cerebral heterotopia is mature neural tissue encountered outside of the subarachnoidal space.[1] In our case, a teratoma was unlikely because teratomas consist of at least two germ layers and intracranial communication is rare.[2] The most common site of an encephalocele is in the midline at the occiput. In the
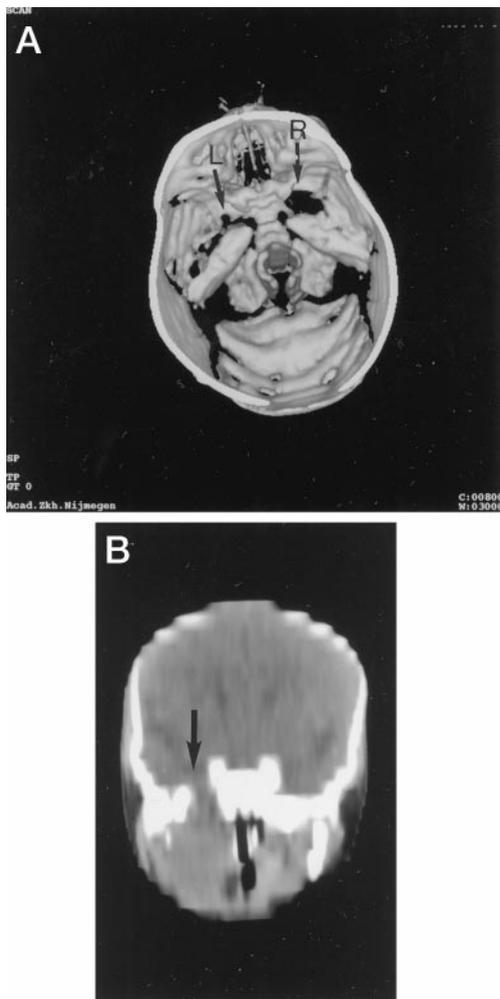
Fig 2. CT. A, three-dimensional reconstruction. Bony defect with a diameter of 10 mm at the location of the right foramen ovale (R). Compare with the left foramen ovale (L). B, coronal view of bony defect at right foramen ovale (arrow).
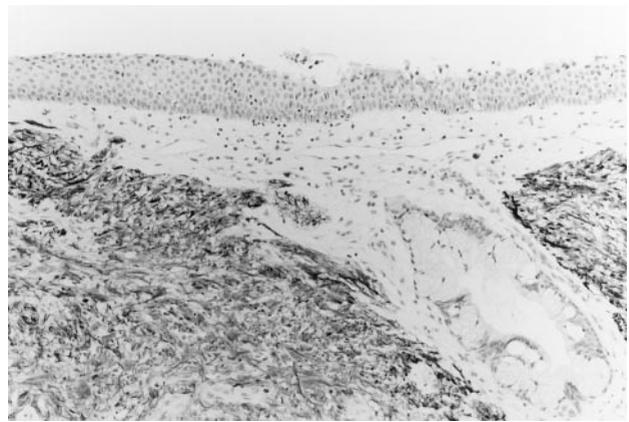

Fig 3. Microscopy of the pharyngeal mass; note the large, circumscribed areas in the submucosal connective tissue that are positive for glial fibrillary acidic protein, confirming the (neuro)glial nature of these areas (immunohistochemical glial fibrillary acidic protein staining; hematoxylin counterstaining; original magnification × 100).

Two mechanisms may be responsible for its origin. The existence of a primary bony defect as a result of failure in chondrification and ossification of the skull may lead to secondary herniation of intracranial structures. A second possibility is a failure of the anterior neuropore to close at the end of the fourth week of gestation, as a result of which the neuroectoderm remains attached to surface ectoderm.[4] In the latter case, a partial or complete secondary closure of the bony defect may lead to a sequestered encephalocele or cerebral heterotopia, no longer in contact with the subarachnoidal space.[1,10] Although in our patient, MRI showed mostly an extradural localization, surgery proved the presence of a substantial intradural component of the process. Thus, by definition the lesion found in this patient involves an encephalocele, but without a clear-cut connection with brain tissue.

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.[3] The common presentation is that of a mass, usually in the midline. Airway obstruction may result from lesions in the nose, nasopharynx, and oropharynx.[5,6] 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.[5] Late symptoms may be visual disturbance and pituitary-hypothalamic dysfunction.[5] 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.[7,9] Two patients with lesions at the petrous apex suffered recurrent meningitis.[9] One patient

Western world, ~75% of the encephaloceles are occipital; 15% sincipital, presenting at the dorsum of the nose, orbit, or forehead; and 10% basal.[1,3,4] 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.[5] 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).[4,6] 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,[7–9] and two cases with a lesion at the petrous apex.[9] 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.[4]

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.

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