ONGENITAL meningoencephaloceles of the cranial base are commonly classified according to the site of the bone defect through which the herniation occurs. This type of classification is predicated on the observation that most of these lesions are located at junctions between the bones of the skull base. The nomenclature used reflects this. That convention may be difficult to apply in cases in which the bone defect is extensive, obscuring its precise anatomical localization. Another circumstance in which such a classification is of limited use occurs when a meningoencephalocele results from some other identifiable abnormality of bone formation. Perhaps the best example of this is the sphenoid wing dysplasia associated with NF1. We recently encountered a child with recurrent meningitis related to an unusual meningocele of the lateral wall of the cavernous sinus, which extended extracranially through an enlarged superior orbital fissure into the pterygopalatine fossa adjacent to the nasal cavity. It was successfully obliterated, via an intradural middle fossa approach, with fat packing and fenestration into the subarachnoid space. This meningocele most likely represents a variant of cranial nerve dural ectasia occasionally seen in individuals with NF1. It has as its basis the same mesodermal defect responsible for the more common sphenoid wing dysplasia and spinal dural ectasias identified with this condition. Involvement of the trigeminal nerve with expansion of the lateral wall of cavernous sinus has not been reported previously. The authors surmise, however, that it may be present in some cases of orbital meningocele associated with sphenoid wing dysplasia.

KEY WORDS • meningocele • Meckel’s cave • neurofibromatosis • meningitis • cavernous sinus • pterygopalatine fossa

Case Report

History. This 8-year-old boy presented in January 1998 with a history of recurrent meningitis. His first bout of meningitis had occurred when he was 4 years of age. It was preceded by a mild case of rhinosinusitis. The causative organism was Streptococcus pneumoniae. The patient was successfully treated with penicillin without neurological sequelae. Six weeks before his present evaluation, a second bout of presumed bacterial meningitis was diagnosed. There were no respiratory symptoms. The patient’s peripheral white blood cell count was 27,700 cells/mm³. His CSF contained 17,950 polymorphonuclear leukocytes with a glucose level of 2 mg/ml. There were no organisms present and the results of cultures were negative. The patient was successfully treated with intravenously administered penicillin and metronidazole followed by orally administered penicillin. At the time of his initial evaluation at our institution, a review of his symptoms was notable for chronic mild nasal congestion with frequent snifing behavior. An apparently unrelated problem was episodic vomiting, particularly with bowel movements, which had been attributed to gastroesophageal reflux.

Examination. The patient’s family history was noncontributory. Otolaryngological and neurological examinations yielded normal findings at the time.
Computerized tomography scanning (Fig. 1A–E) revealed an apparent enlargement of Meckel’s cave. The SOF was enlarged, with protrusion of fluid density into the upper pterygopalatine fossa. The small collection in the pterygopalatine fossa abutted the mucosa close to the sphenopalatine foramen and the sphenoethmoid recess. There was relative hypoplasia of the left lateral body of the sphenoid bone. A foramen rotundum was present. The lateral margin of the abnormality took the apparent shape of the lateral wall of the cavernous sinus. Also noted was an enlargement of the labyrinthine segment of the facial nerve canal and a small expansion in the region of the geniculate turn of the facial nerve.

Axial and coronal MR imaging (Fig. 2) revealed that the abnormality followed the CSF flow on all sequences. The carotid artery extended through the presumed meningocele associated with NF1.
gocele and small septations within it were visualized. The fluid signal on the T2-weighted image could be followed through the internal auditory canal to the slight dilation at the geniculate turn.

**Clinical Course.** Until the relationship between these unusual radiographic findings and the patient’s clinical problem could be resolved, we elected to treat the child with prophylactic antibiotic agents at the time he experienced upper respiratory symptoms. Eight months later, the child’s mother reported that he had been having a recent increase in sniffling and throat clearing associated with clear nasal discharge, especially from the left nostril. He also had experienced a severe bout of streptococcal pharyngotonsillitis, which responded to a regimen of cefadroxil. An endoscopic nasal and nasopharyngeal examination was performed while the patient was in a state of general anesthesia following lumbar puncture with fluorescein instillation. Fluorescein-stained CSF was identified as it arrose from the left posterolateral nasal wall in the region of the sphenoid recess. This corresponded to the pterygopalatine fossa extension of the small meningocele that had been suggested on imaging studies. There was a palpable defect in the nasal wall and the mucosa in this region had a pulsatile character. Otomicroscopic examination performed at the time documented a dry left middle ear with no fluorescein staining.

**Operation.** The lateral wall of the meningocele was exposed intradurally by an anterior subtemporal craniotomy. Except for being expanded, it had the appearance of a normal lateral wall of the cavernous sinus. Opening the substantial dural layer, we entered the meningocele sac. Traversing the cavity posteroanteriorly was a large nerve that we provisionally identified as the second division of the fifth nerve. As the lateral wall of the meningocele was opened more posteriorly, we encountered a substantial nerve invested within this structure running toward the skull base. This had the appearance of the first division of the fifth nerve. The deep aspect of the sac consisted of prominent venous channels of the cavernous sinus itself. The accessible anterior portion of the meningocele cavity was obliterated with fat and the lateral wall was left open.

**Postoperative Course.** An MR image obtained several days postoperatively revealed that the meningocele sac was decompressed. There was, however, an alarmingly large CSF collection inferior and lateral to the temporal lobe, causing substantial mass effect. This was associated with ballooning of the basal portion of the third ventricle. Mild tonsillar herniation was again seen. The ballooning of the inferior third ventricle had been noted transiently on one occasion before surgery. In spite of these findings, the child was without symptoms and the MR imaging changes subsequently resolved. The cause of transient middle fossa CSF collection was unclear. We speculated that it might relate to redirection of CSF from the meningocele via the fenestration in the presence of the mild underlying abnormality of CSF circulation. The patient has continued to do well 18 months postoperatively with resolution of his previous postnasal discharge. Pediatric neurological evaluation subsequent to surgery documented the presence of multiple café-au-lait spots. Genetics evaluation identified a single Lisch nodule, supporting the diagnosis of NF1.

**Discussion**

Discussions of basal cranial meningoceles and meningoencephaloceles are typically concerned with describing the extracranial sac, its location and contents, and the bone defect through which it herniates.8,9,10 These characteristics provide the basis for all classifications of these lesions. Little or no attention is given to associated intracranial abnormalities, other than a description of cerebral dysplasias and/or hydrocephalus. In the present case, the most prominent feature was a CSF space within the dural layers of the lateral wall of the cavernous sinus, extending from Meckel’s cave through the skull base in the region of the SOF. This was associated with relative hypoplasia of the basisphenoid bone, widening of the SOF at the expense of the greater wing of the sphenoid bone, and enlargement of the foramen rotundum. The extracranial meningocele associated with these bone defects was relatively inapparent. It might not have been appreciated had we not been looking for a source for the recurrent meningitis. Finding that the child had NF1 ultimately allowed us to relate this unusual lesion to that condition and better understand its character.

Mesodermal defects of the craniospinal axis are recognized concomitants of NF1. In the spine, these typically involve varying degrees of ectasia of the meninges with associated bone anomalies such as foraminal enlargement and scalloping of the vertebral bodies. The thoracic spine is most commonly affected, with intrathoracic meningoceles presenting as asymptomatic paraspinal masses on x-ray films.6 In the lumbosacral spine, abnormalities vary from dural ectasia with an enlarged spinal canal to extensive bilateral meningoceles associated with enlarged neural foramina. Rarely one finds an anterior sacral meningocele.8

The most common cranial anomaly identified with NF1 is hypoplasia or absence of the greater wing of the sphenoid bone. There may be associated prolapse of the dura and anterior temporal lobe into the orbit, resulting in pulsating exophthalmos.12 Aside from this abnormality, which affects 5 to 10% of patients with NF1, there is only a single known case report of frontobasal encephalocele with NF.14 That patient presented with CSF rhinorrhea and was found to have multiple bone defects in the orbital roofs and the frontoethmoidal sinuses, with associated meningeal and brain herniations. Hydrocephalus due to aqueductal stenosis was also present in that patient and required shunt placement after repair of the CSF leak. The pathological changes described bear no resemblance to those of our patient. In that report, the bone erosions and herniations appeared to result from increased intracranial pressure associated with untreated hydrocephalus. Kurimoto, et al.,10 have recently described an enormous suboccipital meningocele in an elderly women with NF1. The meningocele arose in relation to an occipital bone defect adjacent to the foramen magnum and presented as a retropharyngeal mass.

Individuals with NF1 may have enlargement of one or more cranial nerve foramina associated with ectasia of the dural root sleeve(s). Although this phenomenon is considerably less common than its spinal counterpart, the pathological process appears to be similar. It has been described for optic and acoustic nerves and may be uni- or bilateral.4,11 In our case, a striking feature was the continuity of
Pterygopalatine meningocele associated with NF1

the CSF-containing space between Meckel’s cave posteriorly and the enlarged SOF and foramen rotundum anteriorly. Meckel’s cave represents an invagination of the arachnoid and meningeal layers of the dura. This creates a CSF space about the preganglionic segment of the fifth cranial nerve and the gasserian ganglion. Fusion of the meningeal dural layer with the postganglionic nerves defines the anterior limit of Meckel’s cave. The nerves are thereafter invested within the dural layers of the outer wall of the cavernous sinus as they approach their respective foramina.15 The imaging and operative findings in our case suggest that the meningocele represented an extreme form of dural ectasia involving the trigeminal nerve. This is consistent with prior observations of cranial nerve dural ectasias in patients with NF1. It is also relevant that our patient had enlargement of the ipsilateral facial nerve canal.

Although this appears to be an isolated case, it is possible that some instances of NF1-related sphenoid wing dysplasia and orbital meningocele may include trigeminal dural ectasia. For example, De Vries, et al.,3 recently described successful operative treatment in such a case. Their published computerized tomography scans suggest continuity between the large anterior middle fossa/orbital meningocele and an expanded CSF space, extending posteriorly along the cavernous sinus to Meckel’s cave. In our case, the basisphenoid bone and the medial greater wing of the sphenoid bone were relatively hypoplastic. The meningeal herniation most closely approximated a sphenomaxillary encephalocele, which is thought to arise between the lateral and medial ossification centers of the sphenoid bone. Depending on the extent of sphenoid dysplasia present in individual cases of NF1, it would not be surprising if trigeminal dural ectasia is occasionally associated with that condition.

Conclusions

We have described an unusual meningocele of the lateral wall of the cavernous sinus and the anterior skull base in a young patient with typical stigmata of NF1. This lesion was discovered during evaluation for recurrent meningitis. It represented an anterior continuation of Meckel’s cave into a large CSF space within the lateral wall of the cavernous sinus, extending extracranially through an enlarged SOF into the pterygopalatine fossa adjacent to the nasal cavity. It was successfully obliterated, via an intradural middle fossa approach, with fat packing and fenestration into the subarachnoid space. This meningocele most likely represents a variant of cranial nerve dural ectasia occasionally seen in individuals with NF1. It has as its basis the same mesodermal defect responsible for the more common sphenoid wing dysplasia and spinal dural ectasias identified with this condition. Involvement of the trigeminal nerve with expansion of the lateral wall of cavernous sinus has not been reported previously. The authors surmise, however, that it may be present in some cases of orbital meningocele associated with sphenoid wing dysplasia.

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