Developmental anterobasal temporal encephalocele and temporal lobe epilepsy

RICHARD LEBLANC, M.SC., M.D., F.R.C.S.(C), DONATELLA TAMPIERI, M.D.,
YVES ROBITAILLE, M.D., F.A.C.P., ANDRÉ OLIVIER, M.D., PH.D., F.R.C.S.(C),
FREDERICK ANDERMANN, M.D., F.R.C.P.(C), AND ALAN SHERWIN, M.D., F.R.C.P.(C)

Montreal Neurological Institute and Hospital, and McGill University, Montreal, Quebec, Canada

The authors describe the association between an anterobasal temporal lobe encephalocele and medically intractable temporal lobe epilepsy in three patients treated successfully by surgery. Two men and one woman, aged 26 to 37 years (mean 31 years), had onset of complex automatism and generalized seizures in their second and fourth decades (mean age 22.7 years). They had been epileptic for 6 to 14 years (mean 8.3 years) before surgery. Preoperative electroencephalograms localized ictal epileptic activity to the left mesial temporal lobe in all cases, and neuropsychological testing revealed dominant temporal lobe dysfunction. Magnetic resonance (MR) imaging demonstrated an anteromedial basal temporal encephalocele extending into the pterygopalatine fossa through a bone defect at the base of the greater sphenoid wing in the region of the foramen rotundum and pterygoid process, a discrete center of embryonal chondrification. At surgery, the encephaloceles were found in front of the uncus, and an area of gliosis extended from the encephalocele to the amygdalohippocampal region. All patients have been seizure-free following anterior temporal resection and amygdalohippocampectomy including the encephalocele. These three cases delineate a condition of disordered embryogenesis wherein a developmental anterobasal temporal encephalocele acts as the substrate for temporal lobe epilepsy. This lesion may be diagnosed preoperatively with MR imaging and should be considered in the differential diagnosis of late-onset temporal lobe epilepsy.

Key Words • encephalocele • meningoencephalocele • temporal lobe • neural crest • seizure • embryogenesis

The surgical treatment of medically intractable seizures depends on the identification of an epileptogenic area in an expendable region of the brain. Epileptogenicity is frequently associated with structural anomalies such as a posttraumatic meningo-cerebral cicatrix, mesial temporal sclerosis, hamartomas, and indolent neoplasms. With the development of magnetic resonance (MR) imaging, structural lesions previously undiagnosed even with computerized tomodraphy (CT) scanning are being recognized with greater frequency.

We report three patients with medically intractable temporal lobe epilepsy associated with an anterobasal temporal encephalocele herniating through the base of the greater sphenoid wing in the region of the foramen rotundum and pterygoid process. In these patients, epilepsy was successfully treated by anterior temporal resection and amygdalohippocampectomy including the encephalocele. One patient (Case 1) was included in a previous report but is described here in greater detail, and two new cases with identical biological, clinical, and pathological features delineating a clinicopathological entity are presented.

Clinical Material and Methods

Patients were assessed according to our protocol for the investigation of medically intractable seizures. Surgery was performed under local or general anesthesia with intraoperative cortical mapping and electrocorticography as previously described. Resected tissue was fixed in 10% buffered formalin for 12 hours, embedded in paraffin, then serially sectioned and stained with Cajal's gold chloride sublimate, Klüver-Barrera, and hematoxylin and eosin. The distribution and severity of reactive glial changes were assessed on representative sections immunoreacted with glial fibrillary acidic protein and S-100 immunoperoxidase-labeled antibodies used according to the classic peroxidase-antiperoxidase technique of Sternberger.

Case Reports

Case 1

This 37-year-old woman presented with a 6-year history of two types of seizures. The first type was preceded by a prodrome of paresthesias affecting the fingers and feet, followed by loss of consciousness and urinary incontinence. The second consisted of psychomotor attacks and complex automatisms.

Examination. The patient's physical examination was normal. Neuropsychological evaluation was consistent with dysfunction in the left (dominant) temporal lobe. Electroencephalographic (EEG) recordings revealed epileptic activity from the left mesial limbic structures. A skull x-ray film was normal. A CT scan of the brain performed after intravenous infusion of contrast material demonstrated that the left lateral ventricle was slightly larger than the right, and a focal area of calcification without surrounding edema or mass effect, considered to be of no clinical significance, was present just anterior to the left central region.

Operation. Electrocorticography confirmed the epileptogenicity of the left temporal lobe. During anterior temporal resection, which included the amygdala and anterior hippocampus, an encephalocele approximately 1 cm in diameter was encountered at the base of the greater sphenoid wing. The encephalocele was associated with a dural defect. The tissue was markedly firm and contained numerous pial-arachnoid adhesions. Gliosis extended from the encephalocele to the amygdalohippocampal region and well into the uncus. The encephalocele was resected and the dural defect repaired. A postoperative skull x-ray film demonstrated the vascular clips which had been placed in the encephalocele cavity within the pterygopalatine fossa. The patient's postoperative course was uneventful and she remained seizure-free 5 years after surgery.

Histological Examination. Histological examination of the encephalocele was performed on three fragments (0.5, 0.2, and 0.1 cm in size) roughly corresponding to the volume of tissue removed from the cavity. The examination revealed segments of isocortex and white matter which, despite the presence of moderately severe neuronal loss and gliosis, remained well laminated. A specimen labeled "amygdala" consisted mostly of the junction of the basomesial part of the amygdaloid complex with the parahippocampal gyrus, and revealed moderate neuronal loss and gliosis. Moderate Chaslin's gliosis was observed in the anterior temporal lobe.

Case 2

This 36-year-old man suffered a generalized tonic-clonic seizure at 16 years of age. Seizures recurred every 4 to 5 months and were sometimes preceded by an inner sense that they were about to occur, but there was no other recalled aura.

Examination. Physical examination was normal. Neuropsychological evaluation revealed left (dominant) temporal lobe dysfunction. Epileptic activity was recorded from the left inferomesial temporal region. Skull x-ray films revealed a conchal pattern of the sella turcica. The foramina spinosum and ovale were well visualized bilaterally, the left foramen rotundum was not seen while the right was well demonstrated, and the left inferior orbital fissure was enlarged.

Fig. 1. Case 2. Computerized tomography scans of the head with bone window, axial (left) and coronal (right) projections, showing erosion of the base of the left pterygoid process between the body and the greater sphenoid wing. The meningoencephalocele appears isodense. The letters a, b, and c (left) correspond to the enlarged views shown right.
Anterobasal temporal encephalocele

A CT scan obtained after intravenous infusion of contrast material revealed a defect roughly 1 cm in size in the medial aspect of the floor of the left middle fossa at the base of the greater sphenoid wing. The defect contained brain parenchyma which impinged upon the lateral wall of the left sphenoid sinus but did not breach it (Fig. 1). An associated hypodensity in the central portion of the encephalocele extended into the intracranial compartment but did not communicate with the temporal horn. Magnetic resonance imaging in the sagittal, coronal, and transverse planes using proton density and T1-weighted imaging modalities demonstrated a 2 × 2-cm portion of brain parenchyma extruding from the middle cranial cavity and entering the pterygopalatine fossa (Fig. 2). The anterior temporal lobe in continuity with the encephalocele had an area of decreased signal intensity on T1-weighted imaging, with a hyperintense peripheral rim on proton density imaging; this area appeared diffusely hyperintense on T2-weighted images (Figs. 2 and 3). Bilateral carotid angiograms were normal. Electroencephalography revealed sharp wave activity in the prefrontal and posterior temporal regions.

Operation. During anterior temporal lobe resection and amygdalectomization, a dural defect in the anteromedial wall of the middle fossa was appreciated, through which cerebral tissue herniated. The tissue appeared remarkably firm and gliotic, and the gliosis extended posteriorly in continuity with the amygdala, hippocampus, and parahippocampal gyrus. The encephalocele was biopsied and amputated at its point of exit from the middle fossa, and a dural repair was fashioned. The patient's postoperative course was unremarkable and he has remained seizure-free 1 year after surgery.

Histological Examination. The salient histopathological abnormalities were confined to the encephalocele and associated tissue (Fig. 4). The white matter lateral to the amygdaloid complex showed loss of myelin and scattered, ectopic, well-differentiated ganglion cells on Klüver-Barrera preparations. The cells were mildly dysplastic; no aberrant giant neurons or astrocytes were observed. However, a single 0.5 × 0.2-cm fragment of tissue harbored a few poorly laminated neurons and a paucity of myelinated fibers with marked isomorphic reactive gliosis. These changes were accompanied by increased numbers of dilated capillaries, which were also isomorphically arranged along their axis of myelinated fibers. Signs of inflammation were confined to thin perivascular cuffs of lymphocytes. The surrounding cortex and white matter showed mild to moderate neuronal loss and gliosis.

Case 3

At birth, this 26-year-old man had been delivered at term with forceps following prolonged labor. At the age of 21 years he had two generalized tonic-clonic seizures while asleep. The generalized seizures did not recur; however, the patient developed psychomotor seizures with complex automatisms during which he was mute and did not appear to comprehend verbal communication.

Examination. Physical examination was normal. Neuropsychological assessment demonstrated the left hemisphere to be dominant for speech and memory functions, despite left-handedness, and was consistent with dominant temporal lobe dysfunction. Epileptic activity was recorded from the mesial and anterior neocortical surfaces of the left temporal lobe. Skull x-ray films and CT scans were normal, while T1- and T2-weighted MR images demonstrated a meningoencephalocele extending from a defect at the base of the greater sphenoid wing and extending into the pterygopalatine fossa (Fig. 5).

Fig. 2. Magnetic resonance T1-weighted images in Case 2 (TR 450 msec, TE 30 msec), coronal (a) and sagittal (b) views, demonstrating the two components of the meningoencephalocele (arrows). The subarachnoid space, limited by the dura, has a low-intensity signal while the encephalocele has an isointense signal. A hypointense signal representing gliosis is seen in the anterior mesial temporal lobe.
FIG. 3. Magnetic resonance images, axial view, in Case 2. In the proton density images (a, b, and c), the meningoencephalocele (a, arrows) appears isointense, while in the T2-weighted images (d, e, and f) it appears hyperintense. Gliotic tissue can also be seen, becoming progressively hyperintense in views a to f.

FIG. 4. Photomicrographs of the encephalocele in Case 2. Left: Poorly laminated neurons (arrows) and a paucity of myelinated fibers with marked isomorphic reactive gliosis can be observed. H & E, × 142. Right: An increased number of engorged capillaries are arranged along the axis of residual myelinated fibers. Holzer, × 142.
Anterobasal temporal encephalocele

Operation. Electroencephalography confirmed the preoperative EEG findings, and a depth electrode in the amygdala revealed sustained epileptic activity. During anterior temporal and amygdalohippocampal resection, an area of brain herniation was encountered. This encephalocele protruded from the middle cranial cavity through a 2-cm defect in the greater sphenoid wing in the region of the foramen rotundum, and was lined by thickened, reactive dura. It was just anterior to the uncus and consisted of firm, markedly gliotic tissue, with the gliosis extending toward the amygdalohippocampal regions. The patient remained seizure-free 1 year after surgery.

Histological Examination. Histological examination of the encephalocele and adjoining temporal structures revealed a focus of severe arachnoidal fibrosis and adventitial fibrosis of the leptomeningeal vessels covering a mushroom-shaped microgyrus (Fig. 6). Its lamination was poorly defined and gliosis was rather widespread, resembling polymicrogyria rather than pachygyria. The adjacent white matter showed heavily collagenized small veins surrounding widened Virchow-Robin spaces. Gliosis was severe, and was associated with abundant diffuse corpora amylacea. The deep white matter harbored scarce ectopic aberrant pyramidal cells but no giant astrocytes. The surrounding temporal isocortex was normally laminated but showed mild to moderate Chaslin's gliosis. Mesial structures were not submitted for histological examination having been aspirated during the course of the resection.

Discussion

Literature Review

In a case similar to ours, Garcia3 reported a 30-year-old woman operated on for temporal lobe epilepsy. During left temporal lobectomy, an encephalocele was encountered leaving the middle fossa through a dural and bony defect in front of the foramen rotundum and lateral to the foramen ovale. The herniation entered the zygomatic or pterygopalatine fossa lateral to the sphenoid sinus. Our three cases and that of Garcia bear the following striking similarities: seizure onset was late; the seizures originated in the left temporal lobe; and the bony defect leading to the meningoencephalocele was in the base of the greater sphenoid wing in the region of the foramen rotundum and pterygoid process, allowing the encephalocele and its coverings to protrude into the pterygopalatine fossa. In this location, the meningoencephaloceles were not apparent to inspection, nor was CT scanning of the middle fossa sufficiently discriminating to demonstrate it in all cases.

Encephaloceles

Cranial encephaloceles and meningoencephaloceles occur most frequently in the occipital region where they arise from a defect of membranous ossification, and at the junction of the frontal and ethmoidal bones mainly in Southeast Asians.11,19 Basal encephaloceles are relatively infrequent and may be associated with midline cerebral anomalies such as absence of the optic chiasm and tract or agenesis of the corpus callosum, fornix, and septum pellucidum.11 They have been classified as follows:11 1) sphenopharyngeal or transphenoidal, when they protrude into the epipharynx and/or sphenoid sinus; 2) sphenoorbital, when the protrusion is through the superior orbital fissure into the posterior orbit producing unilateral exophthalmos; 3) sphenoethmoidal, when the cerebral mass extends through the sphenoid and ethmoidal bones into the posterior nasal cavity; 4) transethmoidal, when the encephalocele extends into the anterior nasal cavity; and 5) sphenomaxillary, when the meningoencephalocele passes through the superior orbital fissure into the orbit and through the inferior orbital fissure into the pterygopalatine fossa. Our cases do not fit into this classification system, as

![Fig. 5. Proton density (a) and T2-weighted (b) magnetic resonance images, axial view, in Case 3. The meningoencephalocele (arrow) extends from the left temporomesial region anteriorly toward the pterygopalatine fossa.](image)

![Fig. 6. Low-power photomicrograph of the encephalocele in Case 3. Marked and extensive arachnoidal fibrosis is seen covering a mushroom-shaped gyrus with poorly defined lamination and widespread gliosis. Kluver-Barrera, × 20.](image)
the bone defect occurred at the base of the greater sphenoid wing in the region of the foramen rotundum and pterygoid process, with the encephalocele and its covering protruding into the pterygopalatine fossa without involvement of the orbit. In one case, the inferior orbital fissure was enlarged ipsilateral to the defect, but this likely represented hypoplasia of its inferior border secondary to the osseous defect of the base of the greater sphenoid wing. Other forms of developmental temporal encephaloceles have been described, but these involve the region of the pterion and, less commonly, the asterion.9,14

Development of the Greater Sphenoid Wing
The greater sphenoid wing starts as a thickening of mesenchyme that surrounds the primitive brain vesicles. The bulk of its area ossifies in membrane, but its base and the root of the pterygoid process ossify after passing through a cartilaginous stage beginning in the 2nd month of intrauterine life.20 Ossification of the sphenoid bone arises from 14 centers of ossification.20 The first centers to ossify are those of the greater wing, the earliest residing within the cartilage about the foramen rotundum, forming the base of each wing and the root of the pterygoid process.2 At birth the sphenoid bone is in three parts: the greater wing and pterygoid process on either side, and the body and lesser wing in the middle. These fuse by the end of the 1st year of life.18

We believe that the bone defect in our cases results from a failure of chondrification and ossification of the base of the greater sphenoid wing which normally would produce the pterygoid process, foramen rotundum, and junction.16 Such a developmental defect must occur before the 2nd intrauterine month when chondrification begins. That these bone defects were not traumatic is supported by the absence of a history of head injury and of associated radiological findings, and by the absence of histological evidence of cerebral trauma. It is possible that the application of forceps may have promoted brain herniation through a pre-existent bone defect.

Altered Forebrain Embryogenesis and Epileptogenesis
The frequent presence of ectopic ganglion cells and abnormal networks of capillaries within ectopic brain tissue suggests early abnormalities of neuronal migration and possibly of the radial glia concomitant with focal altered interaction of the neural crest and surrounding mesenchyme during embryogenesis of the basal forebrain. The frequent finding of widespread architectonic abnormalities involving structures dependent on normal development of forebrain vesicles supports this hypothesis.5,9,16 The relationship of the meningoencephalocele to the genesis of temporal lobe epilepsy is strongly suggested by the extension of gliosis from the encephalocele in direct continuity to the amygdalohippocampal region, the most active epileptic area in our cases.

Conclusions
The cases described here are examples of a developmental defect acting as the substrate for temporal lobe epilepsy.7 Such defects can be readily recognized during life with MR imaging. The possibility of these lesions should be considered in the differential diagnosis of patients with medically intractable temporal lobe epilepsy, especially when there is no other obvious cause for epilepsy. Such patients can be rendered seizure-free after resection of the epileptogenic area.

References
Anterobasal temporal encephalocele


Manuscript received July 25, 1990.
Accepted in final form November 16, 1990.
Address reprint requests to: Richard Leblanc, M.D., F.R.C.S.(C), Department of Neurosurgery, Montreal Neurological Institute, 3801 University Street, Montreal, Quebec H3A 2B4, Canada.