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SURGERY FOR EXTRATEMPORAL EPILEPSIES

Technologic advances have provided modern alternatives to resective surgery for medically intractable epilepsy, but none has supplanted surgical resection in efficacy. Advances in neurosurgical techniques and neuroanesthesia have made operative mortality a rare occurrence. Advances in functional imaging, stimulation brain mapping, and intraoperative image guidance help minimize the chance for neurological deficit. One of the most significant developments in the treatment of epilepsy has been the recognition of specific surgically remediable syndromes of epilepsy. Foremost among these conditions has been the syndrome of mesial temporal lobe epilepsy, characterized by distinct patterns of semiology, electroencephalographic signature, imaging correlates, and histopathology. The hallmark of this syndrome is hippocampal sclerosis, which underlies a hyperexcitable, recurrent, and pharmacologically resistant pattern of electrical activity. From the surgeon's standpoint, the significance of this syndrome is the feasibility of a uniform surgical approach to the disease. Unlike mesial temporal lobe epilepsy, extratemporal epilepsy comprises a variety of seizure types depending on the location of the epileptogenic cortex. Lateral temporal lobe epilepsy, medial and lateral frontal lobe epilepsy, and the more rare entity of parieto-occipital or posterior cortex epilepsy, all fall within the category of neocortical epilepsy. Extratemporal neocortical epilepsies can involve a variety of pathways and structures, and may involve or be in close proximity to eloquent cortex including motor, speech, and visual areas.

The ratio of temporal to extratemporal resections reflects not only the relatively high epileptogenicity of the temporal lobe but also the difficulties encountered in attempting to define the localization and extent of the epileptic zones in extratemporal epilepsy, which are often more diffuse and frequently overlap eloquent areas, thus preventing complete resections.

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EXTRATEMPORAL EPILEPSY

Extratemporal resections were more common in earlier series of epilepsy surgery. (Penfield W, 1958). In recent epilepsy series, however, these procedures account for only 15–20% of cases, whereas up to 80% of all surgical procedures involve the temporal lobe. (Engel J Jr, 1996). In adult epilepsy surgery series, extratemporal surgery represents 13% to 37% of operations. Of the extratemporal resections, 60% to 84% were frontal, 4% to 20% were parietal, 3% to 20% were occipital. (Eriksson S, 1999)

Extratemporal epilepsy presents a more complex challenge for the surgeon. Because there is no analogue to the distinct pathology found in mesial temporal lobe epilepsy, there is no uniform resection plan for extratemporal epilepsy. It is necessary to achieve seizure control and functionally feasible with no neurological deficit or a deficit that is acceptable to the patient).

This zone of planned resection may be delineated by using one or more of the following features: (1) seizure semiology; (2) focal abnormality on electroencephalography (EEG) or magneto-encephalography (MEG); (3) a focal abnormality on magnetic resonance imaging (MRI); (4) focal metabolic abnormality; (5) focal abnormality in blood flow on single-photon emission tomography (SPECT); and (6) concordance with neurocognitive impairment. The existence of a focal MRI abnormality is probably the most useful diagnostic modality for planning a surgical approach for extratemporal epilepsy. The presence of such a focal structural abnormality immediately classifies a case as extratemporal lesional epilepsy, which usually correlates with a better surgical prognosis.

The semiology of extratemporal neocortical epilepsy is less well characterized, even when the seizure focus is localized to a single lobe (frontal, temporal, or parietal). Extratemporal lobe epilepsies also tend to spread rapidly, thus making localization based on their clinical characteristics difficult. In some cases, especially in patients with frontal lobe epilepsy, seizures cross to the contralateral side rapidly, which makes it difficult to even lateralize the site of seizure onset.

Because of the risk for neurological injury, a standard resection strategy cannot be used. Instead, each resection must be tailored to the unique characteristics of each patient. In contrast to temporal lobe epilepsy, which frequently has the consistent underlying pathology of hippocampal sclerosis, extratemporal lobe epilepsies have a wide variety of underlying pathologies ranging from tumors and other space-occupying lesions to developmental abnormalities and trauma. As might be expected, surgical outcomes differ for patients with different underlying pathologic conditions.

Frontal lobe epilepsies tend to be briefer, more frequent, and may involve the supplementary motor area (SMA), motor strip, or consist of brief lapses of awareness („pseudoabsences"), and the discharge can spread rapidly to other brain areas (Garcia PA, Laxer KD, 2001). Parietal and occipital seizures classically begin with visual and somatosensory auras, and may arise within or close to eloquent cortex (Geller EB, Heverly D, 2001).

Extratemporal epilepsies do not share a unifying pathologic finding. Lesions can be classified as developmental abnormalities including heterotopias, hamartomas, cortical dysplasias, and gyral anomalies; tumors including gangliogliomas, gliomas, and dysembryoplastic neuroectodermal tumor (DNET) among others; vascular lesions including cavernous malformations and arteriovenous malformations; and encephalomalacia due to trauma or ischemia (Table 1).

Table 1. Classification of Lesions

Developmental abnormalities:
- Cortical dysplasia
- Heterotopias
- Hamartomas
- Gyral abnormalities

Tumors:
- Grade I gliomas
- Grade II gliomas

Vascular lesions:
- AVMs
- Cavernous malformations

Encephalomalacia:
- Trauma (including scar)
- Ischemia
- Birth trauma

Other:
- Gliosis
- Granuloma
- Unspecified
- Normal brain (no lesion)

The presence of a lesion on preoperative imaging studies has a significant impact on the surgical prognosis. Seizure-free outcomes after lesional extratemporal epilepsy surgery are significantly better than those after nonlesional epilepsy surgery. MRI scanners with higher resolution offer promise for identifying anatomic abnormalities in more patients, so many surgical specimens from „nonlesional" epilepsy surgery are found to have abnormalities on subsequent pathologic analysis (Strandberg M, Larsson EM, 2008).

EEG findings vary in extratemporal epilepsies. Lobar localization may not be possible based on scalp recordings alone, and secondary spread is common. Although noninvasive measures can generally localize seizure foci, none can do so

with perfect sensitivity and specificity. Hong et al. (Hong K-S, 2002) compared ictal EEG, interictal FDG-PET, and ictal SPECT with invasive monitoring in 41 patients with intractable epilepsy and no visible lesion on MRI. Ictal EEG was most likely to correctly localize the seizure focus to the appropriate lobe, doing so in 67% of patients. FDG-PET localized the correct lobe in 43% of patients, and ictal SPECT localized correctly in 33% of patients. Boon et al. similarly examined patients with lesions on computed tomography (CT) or MRI (Boon P, 1994). Of 20 patients (including five with isolated MTS) this group found seizure semiology correlated with lesion location in 55%, interictal EEG lateralization was correct in 85%, ictal EEG lateralized correctly in 70% and localized in 50%, and interictal PET was congruent in 81%. The epileptogenic focus of epileptic activity may not correlate precisely with the imaging abnormality. Techniques for localizing the electrical abnormality include scalp EEG, which often only gives lobar localization, invasive EEG monitoring (utilizing surgically implanted electrode grids or strips on the cortical surface, or depth electrodes placed within the brain matter), and intraoperative electrocorticography (ECoG). Invasive monitoring requires proper placement of surface or depth electrodes. Ictal recordings are most useful in localizing seizure onsets, but prolonged monitoring may be required to record an ictal event. Intraoperative ECoG may vary depending on whether ictal versus interictal recording is obtained, and the presence of anesthetic agents.

Electrical abnormalities were excised by corticectomy using ECoG guidance or long-term extraoperative invasive EEG monitoring. Surgery is only considered for epilepsy uncontrolled by medications (intractable), or epilepsy associated with lesions for which surgical treatment is indicated regardless of associated seizures as with tumors and an arteriovenous malformation.

Outcomes after surgery can be classified by the Engel system, with four main categories of seizure outcome (Engel J, 1993) or by newer ILAE classification system (ILAE I-VI).

The categories in Engel system are I, seizure-free; II, rare seizures (two to three per year); III, worthwhile improvement in seizure frequency (>90% reduction); and IV, no worthwhile improvement in seizure frequency (<90% reduction). Each category can be further subclassified.

Surgical strategies fell into several groups:
1. those with the goal of resecting the lesion,
2. those that resect both the lesion and the electrical abnormality, and
3. those that resect the electrical abnormality alone.
4. lobectomy
5. disconnecting the pathways of seizure spread

Lesionectomy is defined as standard surgical removal of the structural lesion alone. In some cases, this may involve removal of adjacent structurally normal brain, or a formal lobectomy. Incomplete lesionectomy and resection of electrically

active cortex alone appear less successful, with subsequent lesionectomy often having a positive effect.

A second approach is to remove the epileptogenic cortex alone. The rationale is that the structural abnormality seen on imaging may be unrelated to the seizure syndrome. Sometimes the lesion itself may not be resectable if it is located in inaccessible or eloquent cortex. In general, if a lesion is present in some relation to the ictal source, some attempt will be made at resection.

Other strategies aim to transect pathways along which seizures spread. Seizure surgery aims to remove the structural lesion, if one is present, as well as the epileptogenic cortex. The epileptic foci may be adjacent to or removed from any lesion that is present.

FRONTAL LOBE EPILEPSIES

The frontal lobe is the largest lobe of the brain, and it encompasses several distinct anatomic-functional units, including the primary motor region; supplementary motor areas; language areas in the dominant frontal operculum; the frontal eye fields; part of the cingulate gyrus; a component of the limbic system. Also, the orbitofrontal and ventromedial regions, which play a major role in the regulation of emotions; and the dorsolateral frontal region, which has major cognitive function, especially in executive functions and working memory. Seizures in other regions of the frontal lobe have shown significant variability, which has resulted in difficulty characterizing classic frontal lobe epilepsy syndromes.

Localization of a single resectable focus in patients with frontal lobe epilepsy is typically difficult because of multiple pathways that allow rapid ictal spread within the frontal lobe and to other lobes and the contralateral side. Medically intractable frontal lobe epilepsy can be difficult to treat, and outcomes after surgical treatment are less favorable than those with temporal lobe epilepsy (Weiser HG, 1995).

A systematic review of epilepsy surgery studies showed a seizure-free rate of 27% with frontal lobe surgery in all age groups (Téllez-Zenteno JF, 2005).

Intracranial monitoring may play a greater role in frontal lobe epilepsy to define the ictal origin. However, even with invasive monitoring, the ictal focus can be difficult to identify. Sampling error may occur due to the large size of the frontal lobe. Some authors find subdural recording is mandatory in frontal lobe epilepsy (Olivier A. 1995).

The frontal lobe lacks clearly defined anatomical markers associated with seizure patterns, and neurological morbidity can be high, particularly in the dominant hemisphere. Subdural recording allows recording around a known lesion as well as functional mapping. Depth electrodes can record from deep frontal lobe structures, but the sampled area is small. Surgical options are similar to those of the temporal lobe, and include lobectomy, corticectomy, and disconnection of pathways.

Most authors reported a variety of surgical strategies, performing ECoG and invasive monitoring in some cases, and partial or total lobectomies in others, making it difficult to compare strategies. Olivier reported his experience with 88 patients

with frontal lobe seizures, using ECoG as the primary means of localizing the epileptogenic focus, but adding invasive EEG monitoring if no lesion was present (Olivier A. 1995).

Posterior cortex epilepsies encompass a group of epilepsies originating from the occipital and parietal lobe and also the occipital border of the temporal lobe, or from any combination of these regions.

PARIETAL EPILEPSIES

Parietal lobe epilepsy is relatively rare in series of epilepsy surgeries, perhaps because of an innate resistance to seizures in the region; the difficulty of localizing these seizures, which may be accompanied by symptoms referable to other lobes; and the reluctance of surgeons to resect tissue in this area. Seizures originating in the parietal area have not received as much investigation as frontal lobe or temporal lobe seizure syndromes.

Parietal lobe seizures are typically characterized by somatosensory auras, pain, paresthesias, vertigo, head and eye deviation, complex visual hallucinations, sensations of body movements, and actual complex movements of the extremities. In general, resections in the parietal dominant hemisphere should take into consideration the high probability of language deficits in the vicinity of the sylvian fissure and more superiorly in regions such as the angular gyrus. The nondominant parietal lobe mediates important visuospatial functions. Large resections in this lobe cannot be undertaken without severe impairment in spatial cognition. Seizure control outcomes after parietal resection are reportedly slightly better than outcomes after frontal lobe resection. A systematic review of epilepsy surgery studies showed a seizure – free rate of 46% with parietal lobe surgery in all age groups (Téllez-Zenteno JF, 2005). Results vary widely depending on the presence and nature of a structural lesion as well as surgical strategy

The lack of typical seizure characteristics and frequent spread make localizing these epilepsies difficult. Scalp EEG can be misleading and, at best, may lateralize the seizure focus (Cascino GD, 1993). EEG may show abnormalities not only in parietal and occipital lobes, but also in ipsilateral or contralateral temporal and frontal lobes. Neuroimaging is critical to define structural abnormalities and guide the hunt for the seizure focus. Functional mapping may play a key role in surgical planning to avoid or minimize neurologic deficits from injury to the somatosensory cortex, association cortex, language areas.

OCCIPITAL LOBE EPILEPSIES

Occipital lobe epilepsy is also rare. Clinically, occipital lobe epilepsy is characterized most often by visual auras. These visual auras are elementary in nature and are described as lights, spots, or simple shapes that can be flashing or moving. Formed visual hallucinations are more suggestive of temporal lobe seizure. Another clinical feature of occipital lobe epilepsy is episodic blindness, which can involve

half the visual field or the entire visual field. Other signs observed with occipital seizures include blinking and tonic or clonic eye deviation (Williamson PD, 1992).

Occipital lobe seizures can spread rapidly to the temporal lobe, thus making localization of their onset difficult and occasionally leading to relatively ineffective temporal lobectomy. Given the frequent spread of occipital lobe seizures to the temporal lobe, resections are sometimes designed to include some portion of the temporal lobe (Kuzniecky R, Gilliam F, 1997). Several surgical techniques have been reported for patients with intractable epilepsy, with or without lesions, and an occipital focus. Most of these cases were associated with a lesion, often developmental. Surgical strategies included partial or total occipital lobectomy, lesionectomy, and temporal lobectomy. Thus, for parietal and occipital lobe intractable epilepsy, no clear surgical strategy emerges for all cases. Invasive monitoring was useful for localizing seizure onsets in some patients with unclear foci. Lesionectomy alone was often successful, particularly for tumors. Patients who underwent ECoG generally had better outcomes if abnormal spiking was reduced or absent after the resection. Pursuing temporal lobectomy alone in patients with occipital foci generally yielded less favorable results. A systematic review of epilepsy surgery studies showed a seizure-free rate of 46% with occipital lobe surgery in patients of all ages (Téllez-Zenteno JF, 2005).

CONCLUSION

In summary, no clear surgical strategy fits every case of extratemporal neocortical epilepsy. Outcome depends on many factors, including the presence and pathology of a lesion, the location of the seizure focus, and the relationship to functional cortex. As neuroimaging techniques improve, the number of cryptogenic declines, and increasingly subtle lesions are identified. Strategies to improve outcomes include the use of ECoG and invasive monitoring to both identify the seizure focus as well as adjacent functional cortex. Although no single surgical strategy is ideal for every patient, as imaging and recording techniques improve, more tools are available to the neurosurgeon to tailor the best strategy for each individual patient.

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