The Symptomatic Localization-Related Epilepsies: Problems with Subclassification

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Classification of different types of epilepsy based on region of seizure origin is particularly appropriate in patients being considered for epilepsy surgery. While some progress has been made, many of the specific seizure types are based on few, often inadequately documented, cases. Further progress requires that the region of origin be established and clinical characteristics documented. Two examples of carefully studied patients are described.

SUBCLASSIFICATION: THE SYMPTOMATIC LOCALIZATION-RELATED EPILEPSIES

Attempts to describe and categorize episodic attacks that we now call epilepsy began in ancient times [1]. While these early efforts at epileptic seizure classification were frequently tainted with superstitions and religious beliefs, the fact remains that different seizure types have long been recognized. The beginning of the modern era of epileptology is generally attributed to the teachings of John Hughlings Jackson, whose insightful observations brought understanding to functional anatomical relationships [2]. The next hundred years witnessed remarkable expansion in our knowledge of the epilepsies and epileptic mechanisms, yet no classification scheme has ever been entirely successful. Disagreements over terminology, definitions, and interpretations have led to polarized opinions and bitter arguments.

One of the more enduring schemes proposes to classify epileptic seizures on the basis of clinical characteristics [3,4]. Classifying seizures strictly on the basis of clinical features has definite limitations, however. Clinically similar seizures can occur in different conditions, the same condition can result in different seizures, and, finally, a number of different seizure types can occur in the same patient. Once criteria other than clinical features are included, classification involves epileptic syndromes rather than epileptic seizures. Modifiers or other criteria used to define seizures are numerous and include EEG results; patient age; seizure etiology, chronicity, and severity; precipitating factors; and localization features. Although syndromatic classification runs the risk of creating chaos [5], a scheme to classify epileptic syndromes has recently been proposed [6]. This proposal avoids the rather ambiguous term “partial epilepsy” and the sometimes misleading concept of focal epilepsy by including all seizures with probable localized origin in the category of “localization-related epilepsies.”

Symptomatic localization-related epilepsies are those in which the existence of an epileptogenic lesion located in part of one hemisphere is presumed. This is obviously a useful concept in the context of considering surgical intervention. The descriptions of
the symptomatic localization-related epilepsies were admittedly preliminary, however, and, in some cases, incorrect and misleading. For example, the description under the heading "frontal-lobe epilepsy" is of a specific type of complex partial epilepsy of frontal-lobe origin that has only recently been defined [7]. Other types of frontal-lobe epilepsy, e.g., cingulate, supplementary motor, orbitofrontal, are placed under separate localization-related headings. The descriptions of these latter types of frontal-lobe epilepsies were based on very few, often inadequately documented, cases.

In the revised proposal for classifying epileptic seizures [4], it was suggested that the scheme should be considered a "skeleton" to be "fleshed out" as more is learned. The same can be said for syndromatic classification. "Fleshing out" the "skeleton" will not be an easy task since it can only be valid if based on data from a significant number of carefully studied cases in which seizure onset localization is well-established and spread patterns are estimated. The following two case descriptions illustrate some of the difficulties involved in meeting these criteria.

Patient 1

This 41-year-old, right-handed woman had had seizures since age 13. The first seizure was generalized with tonic-clonic activity. Subsequent to starting anticonvulsant therapy, she had rare nocturnal generalized seizures and brief seizures while awake. The brief seizures occurred three to twenty times per day and consisted of her experiencing a "tight feeling" in her head followed by partial to complete loss of consciousness. When she maintained partial awareness, she could hear but could not speak. Some seizures were preceded by partial to complete vision loss. During seizures she had been observed to develop a grimace, clenched fists, and posturing of one or both arms. The attacks lasted about 15 seconds and, if standing, she would fall. She had sustained numerous injuries during seizures, some of them near fatal. Several times a month she would have almost continuous seizures for several hours. Seizures remained uncontrolled despite trials of numerous anticonvulsant drugs, alone and in combination. Her examination was consistently normal. Previous EEG reports described right-sided spikes variably located between frontal and parietal regions.

These data from the referring physician suggested several possible regions of seizure origin. Frequent, brief tonic seizures are usually associated with onset in the medial frontal region [8], but they can also occur following seizure origin in other areas [9,10,11]. Isolated tonic seizures, however, are usually not associated with temporal lobe origin [8]. The history of probable partial status epilepticus would also favor extratemporal origin [12]. Visual loss at seizure onset is usually related to occipital lobe origin [13,14]. Speech arrest does not necessarily lateralize seizures to the dominant hemisphere [15,16].

Evaluation at the Yale–New Haven/West Haven V.A. Epilepsy Unit disclosed additional pertinent history. At age 11, the patient developed episodes of a "dead" feeling and clumsiness of the left side accompanied by loss of half her vision (she could not remember which side), followed by a severe throbbing headache. She was told she had a type of migraine. She only had two of these episodes prior to the onset of her habitual seizures. On further questioning she remembered a similar left-sided "dead" feeling prior to some of her early seizures.

Neurological examination, including formal visual field testing, was normal. Neuropsychological testing demonstrated bilateral deficits but with focal findings suggesting right parietal and right temporal deficits. Scalp EEG monitoring revealed a
prominent right medial parietal interictal spike focus (Fig. 1) and less active independent right mid temporal and left frontal sharp foci. The major ictal change occurred over the right posterior quadrant (Fig. 2). Videotaped seizures revealed tonic contraction of the facial muscles, producing a grimace or pucker, tonic contraction of the hands and arms with clenched fists and both arms held in flexion across her chest. There was consistent deviation of her head to the right. During one seizure she experienced complete visual loss at the beginning of the attack. All attacks began with
a feeling of lightheadedness but no somatosensory feelings were reported. The attacks varied in duration and intensity but were objectively similar. CT scan with and without contrast was normal. T2 weighted MRI revealed a possible region of abnormality in the right posterior parietal region (Fig. 3).

At this stage of her evaluation, evidence favored right parietal or parieto-occipital seizure origin, but the findings were not thought compelling enough to justify direct surgical intervention. Subdural and depth electrodes were placed to sample left and right medial frontal and supplementary motor areas, right medial and lateral occipital regions, right medial and lateral parietal regions, and right medial and anterior temporal structures. Numerous clinical and subclinical seizures were recorded. Subclinical seizures were mostly confined to the right lateral parietal recording sites (Fig. 4), while clinical seizures spread rapidly from the right lateral parietal region to the right supplementary motor region. During some clinical seizures, right lateral occipital spread also occurred (Fig. 5). The patient reported no visual or somatosensory symptoms associated with any seizures recorded during intracranial monitoring.

The results of intracranial recording provided conclusive evidence of consistent right parietal seizure onset. At surgery, a lesion was detected with ultrasound beneath the cortex of the right lateral parietal lobe. Pathological examination of the excised lesion revealed a hamartoma. She has had no seizures in the eight months since surgery.

In retrospect, the episodes of migraine were probably seizures and provided the initial evidence for parietal localization. While episodes of ictal blindness are more commonly associated with occipital lobe seizure origin, they could be explained in this patient by postulating seizure propagation to both occipital lobes. This, however, was not completely verified during intracranial recording. The lack of prominent somato-
sensory symptoms after the initial episodes is surprising. The lesion was located outside the primary somatosensory area in a region of the parietal lobe that might be clinically silent with regard to seizures [8,11]. This fact does not explain why seizures stopped propagating to the nearby somatosensory area, but we have noted the disappearance of the initial seizure symptom in other patients.

Patient 2

This 27-year-old, right-handed man was the product of a difficult, prolonged labor followed by neonatal seizures. He then developed normally until the age of six years, when he began to have episodic staring spells and vomiting. Shortly thereafter, he had a generalized tonic-clonic seizure, and anticonvulsant medications were started. Seizures were never well controlled despite trials with multiple drugs. He described two types of seizures. The first consisted of painful, pulling sensations in his eyes followed by visual loss. These lasted ten seconds and consciousness was not impaired. The second type sometimes began in a manner similar to the first, then progressed to loss of consciousness with tonic posturing, followed by generalized tonic-clonic convulsions. In addition, association with noncompliance, he had three episodes of status epilepticus, one of which was consistent with complex partial status epilepticus. Seizures occurred six to twelve times per week. His examination was reportedly normal. Previous EEG reports described a left temporal focus.
FIG. 5. Clinical seizure recorded with subdural and depth electrodes. Subdural electrodes were those described in Fig. 4. Depth electrodes were linear 18 contact probes with number 18 contact at the tip. LSM and RSM were inserted near the midline anteriorly and directed posteriorly toward the left and right supplementary motor areas. LMT was inserted through the intermediate frontal region approximately 3 cm to the left of the midline and directed inferiorly toward the amygdala. RF was inserted near the midline and directed toward the right orbital frontal region. RPT was inserted through the occiput approximately 3 cm to the right of the midline and directed anteriorly along the length of the hippocampus. The seizure originated diffusely in the right lateral parietal region (all RL contacts) after active spike and slow activity in RL 3. The seizure spread rapidly to the right supplementary motor region (RSM 15-18). Some seizure activity was seen in anterior interhemispheric subdural electrode (RA) and in right occiput (RPT 2-4).

In this case, the referral information also suggested occipital lobe onset because of the symptoms of ocular pulling and visual loss [13,14,17]. The tonic posturing indicated possible frontal origin [8,9], while the EEG reports implicated temporal lobe disease. The episode of complex partial status epilepticus favored extratemporal, possibly frontal, origin [7,12].

During the initial seizure unit admission, no additional pertinent history was elicited. On examination, a right inferior homonymous quadrantanopsia was detected. Neuropsychological testing indicated bilateral posterior impairment. Interictal scalp EEG disclosed rare independent left and right fronto-temporal sharp waves with the left side abnormality being more prominent. During ictal recording there was bilateral slowing with no lateralized onset. Two different types of seizures were recorded on videotape. In one he complained of eye pulling and visual loss. This was followed by confusion and fumbling automatisms. The other seizures consisted of right head and eye deviation, followed by fumbling automatisms with both hands, then strong, right, versive, tonic posturing with extreme deviation of the head and eyes, elevation, external rotation and flexion of the right arm, and extension of both legs. These seizures then progressed into generalized tonic-clonic convulsions. CT scan with and without contrast enhancement was normal. A T2 weighted MRI revealed an area of probable abnormality in the left medial occipital lobe (Fig. 6).
Although occipital lobe onset was suspected, the more dramatic seizures suggested medial frontal origin. In addition, the one seizure with prominent automatisms resembled a temporal lobe seizure. Depth electrodes were implanted to sample left and right medial temporal, left and right medial and lateral occipital, and left and right supplementary motor regions, and left and right medial frontal regions. Interictal recording revealed prominent independent paroxysmal foci in the left and right mid to posterior hippocampi and left medial occipital area. Seizures began in the left occiput and spread anteriorly to involve first both medial temporal lobes, then the left supplementary motor region (Fig. 7). Clinical seizures consisted of combinations of versive tonic activity and the automatisms described above.

The intracranial recording results combined with results of the prior evaluation established the left medial occipital origin of this patient’s seizures. At surgery a left medial occipital lesion was resected. Pathological examination revealed a hamartoma. There have been no seizures during the 15 months since surgery.

**CONCLUSION**

These two patients illustrate the importance of a complete and detailed evaluation for adequately defining and documenting the symptomatic localization-related epilepsies. In both examples, the more dramatic clinical features of the seizures were reflections of spread well beyond the region of seizure origin. This is a problem that plagues classification schemes, as it tends to direct attention away from the region of seizure origin and promote localization errors. The importance of the signal symptom, as repeatedly emphasized by Bancaud and colleagues [18,19], is well demonstrated in both examples; however, the signal symptom was inconsistent or remote in the first
patient. Visual symptoms in the second patient had not previously been thought important and indeed had been considered psychogenic at another epilepsy center.

Although classification of epileptic seizures, epilepsies, and epileptic syndromes has realized considerable progress since Jackson’s descriptions of uncinate fits and epileptic dreamy states [2], much remains to be learned. The epileptic seizure is a dynamic process and the regionally originating epilepsies are the most dynamic. Seizures can spread widely, sometimes very rapidly. Seizures originating in some areas such as the occipital and probably also the parietal lobes have the potential for variable propagation patterns [9,11,14]. Thus, seizures from the same region in different patients can have very different clinical characteristics. Similarly, different seizures from the same region of origin in a given patient can propagate via different patterns and suggest multifocal disease. These phenomena, while well described, are not widely recognized. Their importance in the development of classification schemes for epileptic syndromes is obvious.

The proposal for revising the classification of epileptic seizures recognized the important contribution of closed-circuit television and combined EEG monitoring to the study of seizures [4]. The recent proposal for classification of epileptic syndromes acknowledged the importance of video monitoring but also emphasized the need to use all clinical signs and symptoms as well as results of all investigations to define syndromes accurately [6]. Symptomatic, localization-related syndromes should not be categorically defined until data from sufficient numbers of adequately studied patients can be accumulated.

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