Epileptic syndromes characterized by focal inhibitory motor seizures (ictal paralysis) are rare. This ictal paralysis is best documented with ictal video EEG recordings, which usually reveal hemiparesis or hemiplegia during the ictal EEG pattern without loss of consciousness. The most common foci, patterns of progression, and salient electrophysiological characteristics of ictal paralysis are still uncertain owing to the rarity of this seizure syndrome. We describe a patient with focal inhibitory motor seizures who was evaluated using invasive EEG recording with subdural electrodes implanted for presurgical evaluation.

Case Report

History and Examination. Episodic left paralysis developed in this 6-year-old girl when she was 4 years of age. Her developmental milestones had been normal, and the pregnancy had been uncomplicated. Following the first focal inhibitory motor attack, a series of antiepilepsy drugs (valproic acid, oxcarbazepine, and phenytoin) were prescribed but were ineffective. Initially, the seizures occurred 3–4 times a week, but later occurred as often as 4–5 times a day. Left-sided weakness was the first symptom, and she did not experience somatosensory sensations before weakness. Weakness in the arm was more obvious than that in the leg. About 20 seconds after the onset of paralysis, small tremulous movements of the left or right arm lasted several seconds. The whole ictal episode lasted 30–40 seconds, during which the patient did not lose consciousness. Neurological examination revealed that her left arm was thinner and had about four-fifths the strength of her right arm. Magnetic resonance imaging (Fig. 1) showed slight atrophy of the right centroparietal area. Long-term video EEG monitoring (Bio-logic Systems Corp.) was performed with the high-pass filter set to 0.1 Hz and the low-pass filter set to 100 Hz. Surface EMG activities from bilateral deltoid muscles were recorded simultaneously. During 1 day of monitoring, 5 spontaneous seizures were captured. Scalp EEG showed focal ictal epileptiform discharges and irregular slow waves at centroparietal electrodes C4-P4. Ictal EEG started with spike activity at the bilateral frontocentral area. Ictal activity was initially most prominent at C4. This initial ictal activity was followed by diffuse attenuation for about 20 seconds and then repetitive spikes on all channels with the highest amplitude at the right frontocentral areas (Fig. 2). Electromyographic recordings from the right deltoid muscle revealed that atonic phenomena occurred 1 second after ictal onset.
For presurgical evaluation, subdural electrodes covering the right frontocentral region were implanted, and seizure onset was documented by the electrodes depicted as darkened circles in Fig. 3. By stimulating the electrodes related to ictal onset, we induced clonic activity of the left fingers or the left corner of the mouth, but electrode stimulation could not evoke negative motor phenomena during functional mapping.

Operation. Cortex associated with ictal onset was resected, and a pathological diagnosis of focal cortical dysplasia was made.

Postoperative Course. The surgical range was about $3 \times 1.5$ cm on primary motor area, and it was demonstrated on postresection MR imaging (Fig. 4). The first week after surgery, the muscle strength of the left arm decreased to two-fifths of that in the right arm and gradually returned to preoperative levels at 2 weeks after surgery. The patient has been seizure free for about 3 years on oxcarbazepine.

Discussion

We describe a rare case of focal inhibitory motor seizures with left-sided limb paralysis associated with an epileptic focus in the right primary motor area. Inhibitory motor seizures, in which the conscious patient is unable to move or maintain muscle tone, are rarely observed in clinical practice. There have been occasional reports of focal inhibitory motor seizures, and scalp EEGs have documented ictal discharges in the frontal or centroparietal area during paralysis. Satow et al. described 2 patients with partial epilepsy manifesting atonic seizures, one with frontal lobe epilepsy and the other with parietal lobe epilepsy. Ictal EEGs showed low-voltage fast activity in the frontocentral area, and interictal fluorodeoxyglucose PET revealed hypometabolic regions consistent with EEG findings. Satow and colleagues suggested that the seizures may originate from the negative motor area or primary sensorimotor area. Other studies reported similar results from scalp video EEG monitoring, MR imaging, or PET. Noachtar et al. mapped seizure onset to a mesiofrontal lesion in a patient by using implanted sub-
dural electrodes. Electrical stimulation of the electrodes elicited sensory and supplementary motor-type responses but no negative motor responses (transient paralysis). These authors assumed that the focal akinetic seizures were caused by epileptic activation of the NMA. Indeed, electrical stimulation results supported NMA activation as the probable cause of motor inhibition. The NMA includes the primary and the supplementary NMA. The primary NMA is located in the inferior frontal gyrus, immediately rostral to the primary face motor area, whereas the supplementary NMA lies in the mesial portion of the superior frontal gyrus, immediately rostral to the face motor area of the supplementary sensorimotor area. Electrical stimulation of these areas in humans can elicit motor inhibition.

In contrast, we demonstrated ictal EEG activity arising from the primary motor area during focal inhibitory seizures. Furthermore, electrical stimulation of subdural electrodes close to the focus elicited only positive motor phenomenon, consistent with activation of the primary motor area. How inhibitory motor seizures arise from this area is unclear, however. It is possible that the epileptic focus involved only the superficial cortical layers, with hyperactivation restricted to the L2–3 pyramidal neurons. Elger et al. created epileptic foci at different cortical depths in the motor cortex of experimental rats. Foci involving all layers resulted in spinal discharge (output to spinal motor neurons), but foci confined to the superficial layer alone did not. Thus, if epileptiform discharges involve only the superficial layers of the positive motor area, spinal motor neurons may not be activated and convulsions will not occur. Elger et al. also demonstrated that the spikes localized in the superficial layer produced sustained inhibition of the output pyramidal neurons of the deep layers (L5–6), a phenomenon they called “vertical inhibition within the cortex.” In the patient in our case, small tremulous movements of the left or right arm were seen for several seconds starting about 20 seconds after ictal onset. This suggests that the initial epileptic discharges involved only the superficial layer, with concomitant feed-forward inhibition of L5–6 pyramidal neurons. Only when hyperexcitation spread to deeper motor output neurons were positive motor responses observed. Other results also support focal inhibitory seizures arising from the primary motor area. Ikeda et al. studied the mechanism by which inhibitory motor responses, such as cortical negative myoclonus, are generated in humans. From the results of direct cortical stimulation through subdural electrodes, these authors concluded that the inhibitory mechanism was within the primary sensorimotor area but not in the nonprimary motor areas. Matsumoto et al. obtained an ictal electrocorticogram with chronically implanted subdural electrodes in a patient with low-grade glioma in the right postcentral gyrus. During ictal paresis of the left arm, the epileptic discharges arose from the positive arm motor area of the right precentral gyrus. Based on the above results, we think that the concept of “negative motor areas” is not universally accepted. In
fact, the study by Matsumoto et al., is the only example in which electrical stimulation of a negative motor area elicited motor inhibition. But in patients with epilepsy, inhibitory motor seizures evaluated by subdural recording all support the notion of focal inhibitory seizures arising from the primary motor area.

For accurate diagnosis of this rare syndrome, focal inhibitory motor seizures must be differentiated from focal cerebral ischemia or migraine, and ictal video EEG recordings can help with this distinction. Moreover, epileptic negative myoclonus is similar to focal inhibitory motor seizures but usually lasts less than 500 msecs, so ictal EMG activity should be recorded simultaneously.

In summary, we have shown that intractable ictal paralysis can stem from epileptic foci in the primary motor cortex and can be successfully treated using resection.

Disclosure

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