Supplementary motor area epilepsy

Supplementary motor area (SMA) epilepsy is a well-known clinical condition.

Seizures arising from the supplementary motor area (SMA seizures) are a clinically distinct entity occurring mainly during sleep and characterized by tonic posturing of extremities with preservation of consciousness.

Interictal electroencephalogram (EEG) findings are often normal, and furthermore, an ictal EEG change, if any, may be difficult to be detected by scalp recording, which may lead to the misdiagnosis as pseudoseizures.

Differential diagnosis

Nonepileptic seizures

Outcome

Long-term surgical outcome reports are scarce and correspond to small series or isolated case reports.

Surgery for SMA epilepsy guided by ECoG using a multidisciplinary and multimodality approach is a safe, feasible procedure that shows good seizure control, moderate morbidity, and no mortality [1].

Case series

2017

Retrospective descriptive study (1999-2014), 52 patients underwent lesionectomy and/or corticectomy of the Supplementary motor area (SMA) that was guided by electrocorticography (ECoG). The clinical, neurophysiological, neuroimaging, and pathological findings are described. The Engel Epilepsy Surgery Outcome Scale was used to classify surgical outcome. Descriptive statistics, Student t-test, and Friedman, Kruskal-Wallis, and chi-square tests were used.

Of these 52 patients, the mean age at epilepsy onset was 26.3 years, and the mean preoperative seizure frequency was 14 seizures per month. Etiologies included low-grade tumors in 28 (53.8%) patients, cortical dysplasia in 17 (32.7%) patients, and cavernomas in 7 (13.5%) patients. At a mean follow-up of 5.7 years (range 1-10 years), 32 patients (61%) were classified as Engel Class I, 16 patients (31%) were classified as Engel Class II, and 4 (8%) patients were classified as Engel Class III. Overall seizure reduction was significant ($p = 0.001$). The absence of early postsurgical seizures and lesional etiology were associated with the outcome of Engel Class I ($p = 0.05$). Twenty-six (50%) patients had complications in the immediate postoperative period, all of which resolved completely with no residual neurological deficits.

Surgery for SMA epilepsy guided by ECoG using a multidisciplinary and multimodality approach is a

safe, feasible procedure that shows good seizure control, moderate morbidity, and no mortality [2].

2002

Ikeda et al. analyzed the epileptogenic zone at/or adjacent to the SMA in four patients with clinical SMA seizures. All four patients had noninvasive presurgical evaluations (long-term video/EEG monitoring, MRI, and neuroimaging with radioisotopes), which provided convergent results between ictal semiology and the epileptogenic area, and thus, they had chronically implanted subdural electrodes, and finally had focus resection with a follow-up period of more than 2 years.

Three patients had lesions shown by MRI outside the SMA, and one patient had a lesion within the SMA. Interictal epileptiform discharges were seen at/or outside the SMA. Ictal EEG pattern originated from the SMA in one patient, from the high lateral frontal area in two patients, and from the precuneus in one patient. In the latter three patients, the ictal EEG pattern immediately spread to the SMA. Those ictal onset zones were consistently localized within/or just adjacent to the lesions revealed by MRI. Only one patient had SMA resection, and three had the resection of epileptogenic zone by preserving the SMA. No neurological deficits developed and good seizure control was achieved.

Among surgical candidates for intractable SMA seizures, frontal cortex other than SMA or even parietal cortex can be epileptogenic, and thus, the SMA itself may not necessarily have to be resected. This notion is clinically important when selecting surgical candidates as well as when planning presurgical invasive evaluation in patients with intractable SMA seizures [3].

1996

Baumgartner et al. studied propagation of epileptic discharges in five patients with supplementary motor area (SMA) seizures with subdural grid electrodes implanted over the dorsolateral frontal neocortex and in the interhemispheric fissure. We found that both interictal and ictal epileptic discharges occurred synchronously in the SMA and the primary cortex. The actively involved electrodes were separated by silent electrodes. The time lag between the SMA and the primary motor cortex averaged 25 msec for interictal and 100 msec for ictal discharges. Cortical stimulations of the affected electrodes showed motor effects in corresponding body parts. All patients underwent resections of the EEG onset zone within the SMA while sparing the primary motor cortex and experienced a significant (>90%) reduction of seizure frequency. We conclude that epileptic activity is propagated between the SMA and the primary motor cortex by a somatotopically organized monosynaptic pathway [4].

1988

The clinical and EEG features of 11 patients with seizures arising in the supplementary motor area (SMA) were reviewed. All patients underwent prolonged EEG with simultaneous video recording. Three patients had recordings and electrical stimulation of the SMA using subdural electrode arrays. All patients had preservation of consciousness during the seizure unless it became secondarily generalized. Tonic posturing of the extremities was present in all patients, and in seven it was present bilaterally. Adversive movements were not seen unless the seizure became secondarily generalized. Interictal and/or ictal abnormalities were present at or adjacent to the midline in ten patients.

Seizures arising from the supplementary motor region are clinically distinct, and the diagnosis can almost always be verified with prolonged EEG/video recording [5].

Case reports

A 6-year-old girl with attention-deficit hyperactivity disorder (ADHD) who had been abused by her mother in infancy developed supplementary motor area (SMA) epilepsy. The seizure was characterized by bilateral tonic seizure of the upper and lower extremities, speech arrest, preserved consciousness and a lack of postictal confusion. The duration of the seizure was usually 10-60 seconds. The seizures sometimes clustered. She was diagnosed as having SMA epilepsy based on the characteristic clinical symptoms, interictal EEG, ictal video-EEG and ictal SPECT. Though her seizure was initially improved by anti-epileptic drugs, the symptoms appeared again after discharge. Since her clinical course indicated that her seizure was aggravated by her mental state, treatment included both medication with anti-epileptic drugs and the adjustment of her living environment in cooperation with a child guidance clinic. Thereafter both her epileptic seizure and ADHD symptoms improved. These changes may be related to each other, because both conditions are associated with frontal lobe dysfunction. It was interesting that the adjustment of the environment improved frontal lobe epilepsy, which in turn ameliorated ADHD symptoms [6].

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