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Case report

Persistent verbal and behavioral deficits after resection of the left supplementary motor area in epilepsy surgery

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Abstract

An 8-year-old boy underwent a resection for focal cortical dysplasia at the left supplementary motor area (SMA) for the treatment of intractable epilepsy. The manifestations of SMA syndrome, such as transient mutism and right hemiparesis, resolved within a few weeks. Verbal disfluency and impaired executive function, accompanied by impulsivity and distractibility, persisted for more than 12 months. The verbal and behavioral problems caused serious difficulties in the school life of the patient, until they became less evident at 18 months after surgery. Tractography performed 18 months after surgery revealed a defect in the subportion of fronto-parietal association fibers within the left superior longitudinal fascicles. Verbal influency can persist with unusually long duration after resection of SMA during childhood. Although not discernible on the routine neuroimaging, white matter damage beneath the SMA region could result in serious disabilities in executive function. These complications should be recognized for the prediction and assessment of deficits in children after surgical intervention involving this region.

Keywords: Supplementary motor area; Transcortical motor aphasia; Fluency; Attention deficit hyperactivity disorder; Superior longitudinal fasciculus

1. Introduction

Acute lesions of the supplementary motor area (SMA) in the dominant hemisphere can result in transient contralateral hemiplegia and complete mutism, symptoms which compose SMA syndrome and subside completely within a month in most cases [1]. The SMA syndrome affects adults suffering from brain infarct or hemorrhage, or subjected to tumor resection, and has recently been recognized in children after epilepsy surgery [2]. In addition, this condition can be followed by a phase of several months or longer with reduced spontaneous speech and impaired word fluency—i.e., difficulty in word recall without visual cues [3–5]. This latter verbal disturbance is characterized by preserved abilities in verbal comprehension and word

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repetition, as well as the naming of objects presented visually. This discrepancy between word recall with and without visual cues is characteristic of the SMA syndrome and is in contrast to cases with dorsolateral frontal lobe lesions in which word recall both with and without visual cues are affected [4]. In the present study, we report the case of a boy who presented with postoperative SMA syndrome and persistent verbal deficits. Specifically, the patient exhibited academic and social difficulties that lasted more than a year, as a result of cognitive disabilities and inattentive behaviors that have not yet been described following SMA resection in literature.

2. Case report

An 8-year-old boy was referred to our epilepsy center for intractable seizures. He had exhibited normal psychomotor development and good social behaviors until 7 years of age, when he first experienced unprovoked seizures. The patient experienced tonic-clonic convulsions, involving the right face and extremities with versive posturing to the right side, that lasted 5–30 s and gradually increased in frequency to 15 times per day. The seizures were intractable to treatment with carbamazepine, valproate sodium, phenobarbital, zonisamide, topiramate, lamotrigine, clobazam, phenytoin, and potassium bromide.

Upon examination at the age of 8 years, the patient was a right-handed boy with normal neurological findings. Spikes and spike-waves appeared in the left-central regions on interictal electroencephalography, which evolved into diffuse voltage attenuation with fast activity in the same areas during ictal episodes. Magnetic resonance imaging (MRI) indicated focal cortical thickening with high signal intensity by fluid-attenuated inversion recovery imaging at the bottom of the left superior frontal sulcus, involving the supplementary motor area (SMA) (Fig. 1A–C). A band of high signal intensity in the subcortical white matter extending toward the lateral ventricle was also noted beneath the cortical lesion (Fig. 1C). After confirmation of seizure onset at the cortical lesion by intracranial recording with surface and depth electrodes, the posterior portion of the superior frontal gyrus (SMA), the middle portion of the cingulate gyrus, and the white matter underlying these areas were resected as the epileptogenic zones (Fig. 1D–I). Pathological examination confirmed the diagnosis of focal cortical dysplasia, type IIB. Although rare spikes were still observed in both the frontal areas on EEGs during sleep, epileptic seizures disappeared thereafter with carbamazepine administration.

The patient developed right hemiplegia postoperatively, which gradually resolved within 1 month. Complete mutism also appeared transiently, which soon subsided and was followed by a phase comprising a decrease in spontaneous speech and the emergence of echolalia for approximately 3 days. Verbal output, with good articulation and grammatically correct sentences, gradually increased. However, the patient often experienced difficulty in recalling single words; e.g., he would say, “something to void, in the abdomen,” instead of “pee,” or, “What day is it today?” instead of, “What is the date today?” In addition, marked inattentive, impulsive and distractible behaviors made it difficult for him to conduct daily activities; he opened the door of the car before it stopped, put his fingers on hot apparatus or into a fan, ran into an elevator moving in the opposite direction when he saw the door open, and went into the dining room and remained there when he initially intended to go to the restroom. Neuropsychological assessment conducted 3 weeks after surgery (Table 1) revealed a decrease in the verbal intelligent quotient associated with a decline in verbal comprehension and processing speed. The patient exhibited reduced spontaneous speech and difficulty with the repetition of longer words or sentences, as assessed by the Western aphasia battery (WAB). Specifically, he could only list as few as 2 words in a word fluency task requiring him to list the names of animals, whereas he could competently provide the name of objects upon visual presentation. Impairment in the skills of writing letters and words was also revealed. After discharge at 4 weeks after surgery, the patient returned to school, but experienced difficulties in the required academic and social activities. In addition to slowness in reading and writing, phonemic paraphasia and paragraphia, and impaired skill at calculation were also noted. His attention was easily distracted. They provided him special support with the attendance of an educational assistant in the classroom. He was unable to respond with appropriate responses in the context of conversation, and was unable to answer a phone correctly. Inattentive behaviors also persisted: he suddenly pushed the hazard lamp button while his family was driving on a highway, and bumped into a bicycle while he was watching an airplane and walking.

The verbal and behavioral problems gradually became less evident from 1 year after surgery. When examined 15 months after the surgery, the patient showed improvement in his intelligence quotient, WAB scores, and reading and writing skills, although he still had reduced abilities in processing speed, word fluency, and writing words (Table 1). He could not repeat pseudowords of longer than 6 characters (6 moras in Japanese), nor inversely true words beyond 2 characters (2 moras) at this period. Despite these residual disabilities, the patient behaved and spoke appropriately, and his academic scores attained an average level, or higher, in all subjects by 18 months. An MRI scan was obtained after surgery with a 3.0-T MRI system. Diffusion Tensor Images (DTI) were obtained with both 64-directional diffusion encoding (b value, 1000 s/mm^2 for each