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Simple partial status epilepticus presenting with jargon aphasia and focal hyperperfusion demonstrated by ictal pulsed arterial spin labeling MRI

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Abstract

We report a case of 74-year-old lady, presented with recurrent jargon aphasia as simple partial status epilepticus (SPSE) which lasted for a few days to a few weeks, following a brain abscess removal from the left temporo-parieto-occipital region at the age of 71 years. The ictal activity on electroencephalogram was documented at left posterior quadrant, where marked hyperperfusion was clearly visualized by perfusion image acquired with magnetic resonance imaging (MRI) using pulsed arterial spin-labeling (PASL). Jargon aphasia as a primary feature of simple partial status epilepticus is so uncommon that only few cases have been reported. Furthermore, this report suggests that MRI using PASL is a promising method not only to localize the seizure foci but also to follow up the corresponding regional cerebral blood flow changes noninvasively.

Keywords: Jargon aphasia; non-convulsive seizure; simple partial status epilepticus, pulsed arterial spin-labeling-MRI

INTRODUCTION

The commonest cause of acute onset aphasia is vascular insult, however when the symptoms of aphasia occurs episodically or fluctuates, it may be suggestive of seizure. Jargon aphasia as the primary manifestation of focal epilepsy is uncommon, more so presenting as status epilepticus. It has been accepted that interictal regional hypoperfusion visualized by pulsed arterial spin-labeling (PASL)-MRI has diagnostic value in localization of focus in epilepsy. Ictal regional hyperperfusion on PASL-MRI should have more localizing value. However, there still has been only a few reports of ictal PASL-MRI. We report an elderly lady who presented with jargon aphasia as simple partial status epilepticus (SPSE), during which PASL-MRI revealed ictal regional hyperperfusion over temporal lobe and posterior cortices of language dominant side. We discuss ictal jargon aphasia and validity of ictal PASL-MRI.

CASE REPORT

A 74-year-old right-handed Japanese lady began to have paroxysmal episodes of 'strange speech' at the age of 71 years. Six months prior to this, she underwent surgical evacuation of brain abscess at left posterior quadrant of the brain, resulting in the right homonymous superior quadrantanopia. She could not speak properly, walk or carry out daily activities during the episodes. Her husband noted that the patient just suddenly had a speech arrest and later she uttered meaningless simple syllables and kept repeating it. These episodes lasted for several days to a week and spontaneously ceased. She had been treated with valproate and levetiracetam, which failed to prevent the recurrence.

She was first seen at our hospital when she was 73 years old and asymptomatic. The interictal EEG showed slow waves of delta range at left temporal, occipital and parietal regions (T3-P3-O1). MRI revealed post-operative changes such as enlarged dorsal horn of left lateral ventricle in addition to diffuse cerebral atrophy. Interictal PASL-MRI showed significant hypoperfusion at the left posterior quadrant (Figure 1).

Three months later, she was admitted to our hospital presenting with strange speech for eleven days. The EEG on the day of admission (Day 12) revealed ictal pattern at the left posterior regions repeating every 5 minutes (Figure 2). The ictal speech was characterized by meaningless fluent words and phrases such as "Jubemu-Atamaikan-Shakkin-Biehun", "Nehryan-Nehzan-Nehzan-ne".

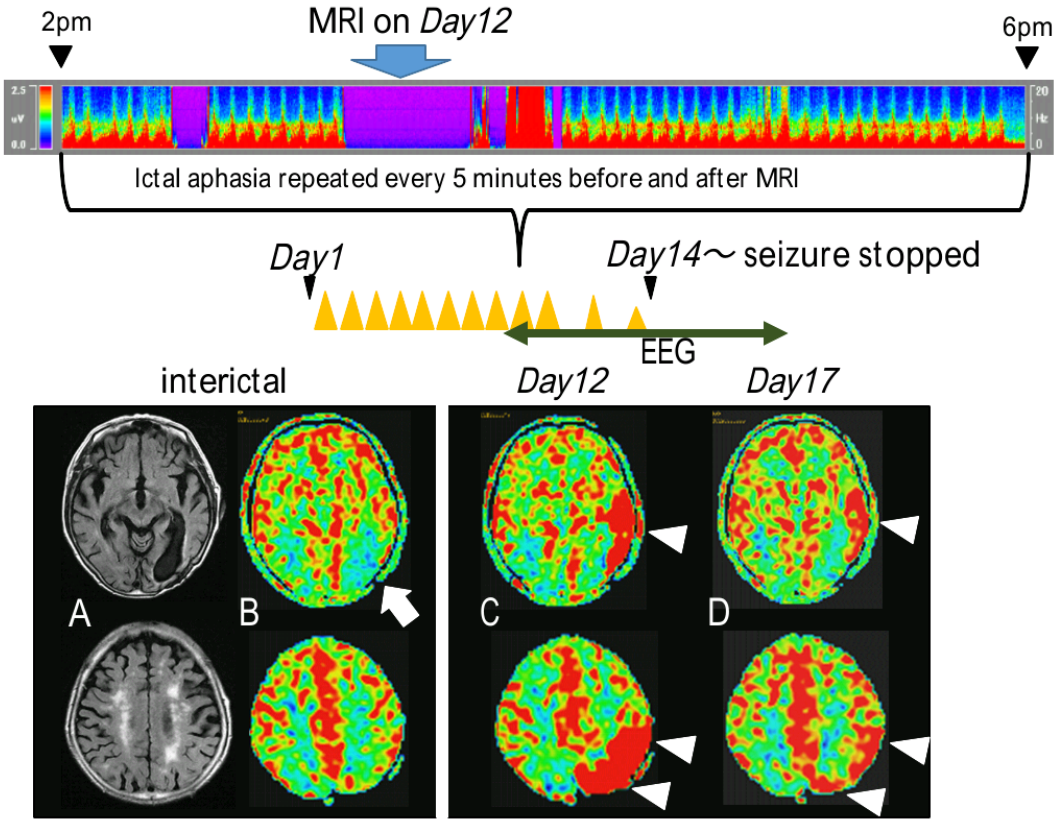

Figure 1. Colored density spectral array represents 4-hour EEG record from 2pm to 6pm on Day 12 of SPSE (upper). Interictal, ictal and periictal PASL-MRI (lower). (A) FLAIR images showed post-operative changes, enlarged dorsal horn of left lateral ventricle in addition to diffuse cerebral atrophy and ischemic change. (B) Hypoperfusion at the left posterior quadrant on interictal PASL-MRI (arrow) (C) Hyperperfusion at left posterior cortices on ictal PASL-MRI on Day 12 when aphasic seizures repeated every five minutes. (arrowheads) (D) Persistent but milder hyperperfusion at the same regions on Day 17, three days after aphasic seizure has ceased (arrowheads)

The prosody of her speech was preserved. In between bouts of seizures, she tried to explain haltingly how difficult situation she was in, using non-fluent, brief and meaningful words and phrases. Her spontaneous speech was significantly reduced. The jargon aphasia was associated with clumsiness of her right hand. She could not hold her chopsticks properly when epileptic discharges were present. She could skillfully use them again, soon after the discharge disappeared.

PASL-MRI was obtained in the middle of repetitive ictal discharges appearing every five minutes on Day12. The images were acquired using MRI GE Health care, Signa HDxt Optima Edition TwinSpeed 1.5T (Ver23) ▪ 8-ch, Brain Array Coil. The following parameters of PASL was used: 36 slices; 4 mm thickness, FOV: 24 cm, TE: 9.8ms, TV: 4955ms, NEX: 3.0, points: 512, arms: 8, bandwidth: 62.5, post-label delay 1525ms. The ictal PASL-MRI showed marked hyperperfusion over the left posterior cortices (Figure 1).

Intravenous midazolam was only temporarily effective to abort the seizure. Intravenous phenytoin loading finally stopped the cycling pattern of the seizures and there was no recurrence of ictal pattern on EEG after day-14. PASL-MRI was repeated at Day 17 when no ictal discharge was seen on EEG performed just before MRI, still showing hyperperfusion predominantly over the left temporal and parietal lobes, which was relatively reduced in intensity compared to the ictal PASL imaging on Day12.

She developed another episode of SPSE in the following month triggered by decreased level of phenytoin. This time, the EEG showed continuous ictal discharges predominantly over the similar regions for five days, which later developed into periodic lateralized epileptiform discharges (PLEDs) for the following three days (Figure 3).

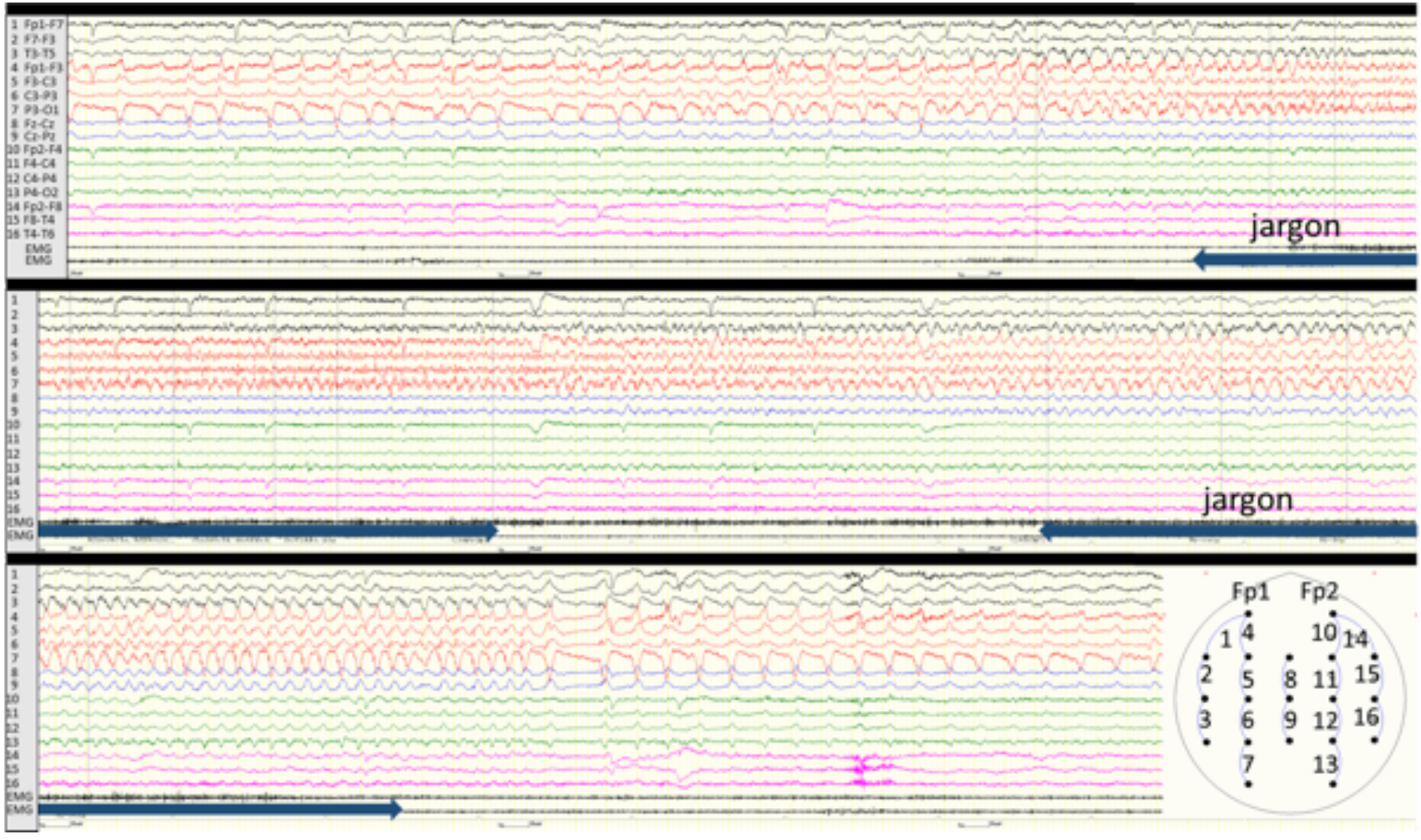

Figure 2. Ictal EEG (longitudinal montage)on Day 12. Focal ictal discharges over the posterior quadrant and the evolution in the frequency of the discharges.

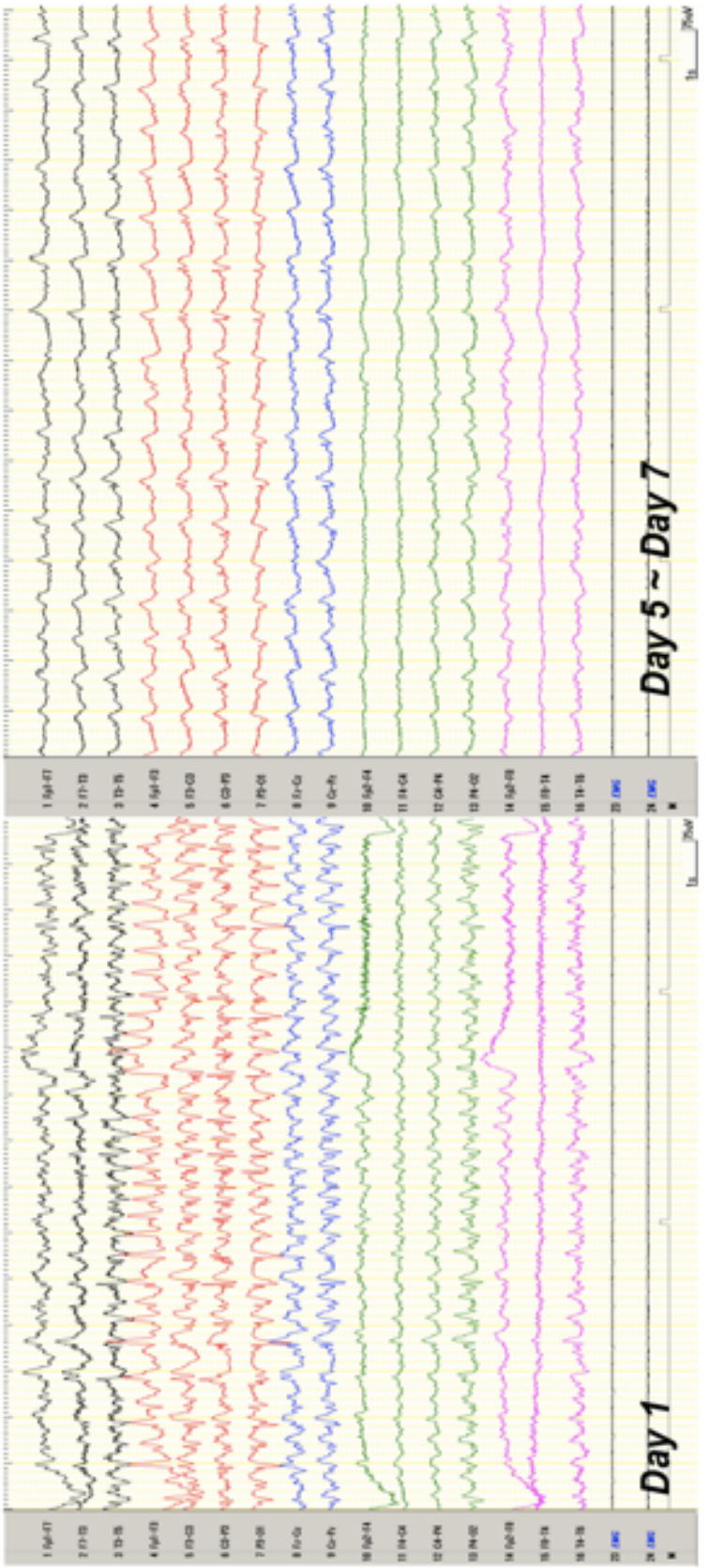

Figure 3. The EEG (longitudinal montage) showed continuous ictal discharges predominantly over the left temporal, occipital and parietal regions (T3-P3-O1) for five days, and the periodic discharges over the same regions for the subsequent three days.

Both PASL-MRI and N-isopropyl- p-[(123) I] iodoamphetamine single photon emission computed tomography (IMP-SPECT) were performed on Day 7, when PLEDs still existed on EEG, which revealed mild hyperperfusion at the left temporoparietal region (Figure 4).

The combination of phenytoin, valproate and clobazam has made her free from SPSE since then, except minor brief episodes of aphasia triggered by her missing drugs. Her speech became comprehensible enough for daily conversation, though some paraphasic errors remained and some difficulties in reading and writing words persisted.

DISCUSSION

Our patient's ictal presentation was jargon aphasia with preserved consciousness, accompanied by ictal discharges on EEG. Ictal jargon aphasia as the predominant feature of SPSE has rarely been reported.[1,2] The diagnostic criteria for ictal aphasia were first defined by Rosenbaum *et al*.[3] and then was modified further; (i) The patient has language production during the seizure. (ii) Language production shows aphasic features. (iii) Consciousness is preserved. (iv) The seizures are correlated with the aphasia, as documented by EEG monitoring and behavioral testing. (v) The aphasia resolves, or nearly so, concurrent with successful treatment of the seizures.[4] Our patient fulfilled the criteria as above.

Jargon aphasia is described as fluent, non-effortful flow of sounds and words but incomprehensible without meaning[5,6] and it is divided into undifferentiated jargon, asemantic jargon, paraphasic jargon, and circumlocutory anomic speech.[5] Our patient uttered meaningless speech fluently during seizure. The prosody of

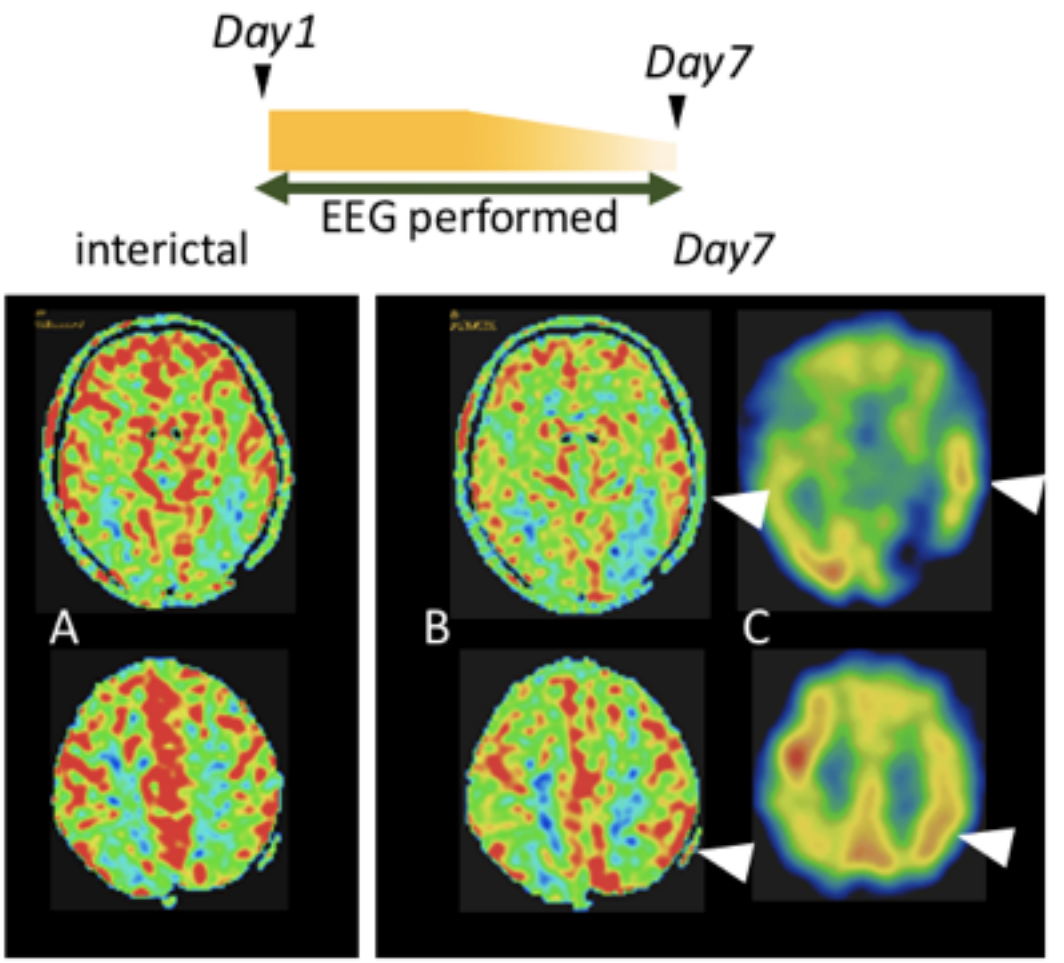

Figure 4. Periictal PASL-MRI (B) and IMP-SPECT (C) on Day 7 in the second SPSE, when left periodic discharges on EEG continued. (A) Interictal PASL-MRI. (B and C) Hyperperfusion at left temporal and parietal lobes was seen on both modalities. (arrowheads)

her speech was preserved. The type of aphasia observed during her seizures can be categorized into asemantic jargon, a fluent aphasia of Wernicke type, which consists of phonologically nonsense syllables or neologisms, however, it may sound grammatically organized foreign language due to its preservation of function words. Her speech consisted of a copious flow of complex, reiterative, phonologically related neologisms. The iterative pattern of phonemic variation is named as stereotypic pattern of alliteration and assonance by Green 1969[7] which is considered characteristic for the neologistic jargon.[6]

Bell *et al*. hypothesized that ictal neologistic speech automatism may implicate a focus in the hemisphere dominant for propositional language and the word's sounds are disconnected from word's meaning, producing neologism.[8] Posterior ictal aphasia can manifest either as fluent neologistic speech or as neologistic speech automatisms.[8] Other studies also noted that their patients had lesion at left dominant side particularly posterior quadrant.[9,10] Our patient has a post-operative lesion for brain abscess at the left posterior quadrant where sensory language area is close to.

PASL is a developing technique of MRI which is non-invasive and needs no radioactive tracer. It has been used to measure cerebral blood flow (CBF) which is helpful in the diagnosis of neurological diseases, such as stroke and cerebrovascular diseases, epilepsy, tumour, infection, neurodegenerative and neuropsychiatric disorders.[11-13] Quantitative nature of PASL is capable of quantifying local CBF by measuring the magnetically labelling inflow of the arterial blood at a certain point into the target area and it has been used for lateralization of the epileptic foci.[11,12] The PASL perfusion scans are able to show interictal hypoperfusion, which corresponding to hypoperfusion in inter-ictal positron emission topography (PET).[14,15] Meanwhile, there had been reports identifying the localizing value in ictal hyperperfusion of PASL-MRI while patients are having ongoing seizure[16-18] and up to five hours post-ictallly.[19]

In our case, PASL-MRI was performed during the sequence of repetitive seizures in an interval of 5 minutes, which was confirmed on EEG just before and after MRI. One or two seizures were likely to occur during PASL-MRI, as acquisition of PASL-MRI needed 4 minutes and 22 seconds. Marked hyperperfusion at left posterior region was likely correlated with the concurrent epileptic activity. In the second episode of SPSE, regional hyperperfusion shown in periictal PASL-MRI was equivalent to the IMP-SPECT's finding. A few authors also have described the concordance between ASL and SPECT, the standard method for evaluation of CBF in localizing the epileptogenic zone.[17,20]

Although jargon aphasia presenting as SPSE is rare and it is difficult to recognize, regional hyperperfusion on PASL-MRI would prompt physicians to consider SPSE and perform EEG for differential diagnosis. This report also suggests that PASL-MRI is useful in assessing CBF during seizures and it is as reliable as SPECT. It is a non-invasive technique requiring no radioactive tracer, thus it can be repeated over time to evaluate the progression of the cerebral perfusion pattern of the evolving seizure.

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