Rapidly Progressive Dementia Caused by a Superior Sagittal Sinus Dural Arteriovenous Fistula: A Case Report

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(Received January 28, 2015; Accepted February 26, 2015)

We describe the case of a 72-year-old man who presented with dementia that had progressed rapidly over a few months. Laboratory analysis of blood and cerebrospinal fluid (CSF) showed no abnormalities, with the exception of a slightly increased CSF protein level. Results of routine magnetic resonance imaging (MRI) including diffusion-weighted imaging (DWI) and magnetic resonance angiography (MRA) were unremarkable. However, detailed neuroimaging studies including contrast-enhanced T1-weighted MRI and conventional angiography revealed a superior sagittal sinus (SSS) dural arteriovenous fistula (DAVF). After endovascular embolization and surgical interruption of all arteries feeding the DAVF, the dementia reversed. We should be aware of the possibility of DAVF as the cause of rapidly progressive dementia even if routine MRI reveals no or only minimal abnormality.

Key words: rapidly progressive dementia, treatable dementia, dural arteriovenous fistula, magnetic resonance imaging (MRI), angiography

INTRODUCTION

Slowly progressive dementia is usually caused by a neurodegenerative disorder, such as Alzheimer's disease. Rapidly progressive dementia, to the contrary, can develop from various disorders, such as vascular, autoimmune, infectious, metabolic, and neoplastic conditions [1]. Because some rapidly progressive dementias are treatable, early diagnosis of the underlying cause is of great importance. However, identification of the cause in patients is not easy because rapidly progressive dementia is a nonspecific symptom and the potential causes vary widely.

Rapidly progressive dementia can be caused by an intracranial dural arteriovenous fistula (DAVF) [2–5], which is an abnormal shunt between dural arteries and a dural venous sinus or cortical (leptomeningeal) vein [6]. The most common manifestations of DAVF are pulsatile tinnitus, headaches, focal neurological deficits, and visual disturbances [7]. Although a definitive diagnosis of DAVF should be based on angiographic findings, the malformation can sometimes be diagnosed preliminarily by routine magnetic resonance imaging (MRI); venous ectasia [3–6, 8, 9], intracranial hemorrhage [2, 6, 8–10], and/or cerebral parenchymal changes [3, 5, 6, 8, 9, 11–14] due to venous hypertension are reported diagnostic clues [6, 9].

We report herein a case of rapidly progressive dementia caused by DAVF in which routine MRI revealed only minimal abnormality.

CASE REPORT

A 72-year-old, right-handed Japanese man who had experienced rapidly progressive memory disturbance over a few months was transferred to our hospital after sudden onset of a generalized seizure. Although the seizure resolved with intravenous administration of an anticonvulsant medication, the patient was disoriented, anosognosic, and apathetic, and he showed memory loss. He had a Mini Mental State Examination (MMSE) score of 6, even after he completely regained consciousness. There was no pulsatile tinnitus, headache, nausea, or visual disturbance. He was afebrile, and his blood pressure was normal. Standard blood tests revealed no abnormality. Examination of cerebrospinal fluid (CSF) obtained by lumbar puncture revealed a slightly increased protein level of 67.2 mg/dL, with an opening pressure of 230 mmH_2O. Electroencephalography (EEG) showed no epileptiform discharges. Brain computed tomography (CT) and initial routine MRI, including diffusion-weighted (DWI) and fluid attenuated inversion recovery (FLAIR) imaging, magnetic resonance angiography (MRA), and magnetic resonance venography (MRV), did not depict any responsible lesion (Fig. 1). However, later T2-weighted imaging (T2WI) and contrast-enhanced T1-weighted imaging (T1WI) revealed abnormally dilated cortical veins in the medial aspect of both cere-

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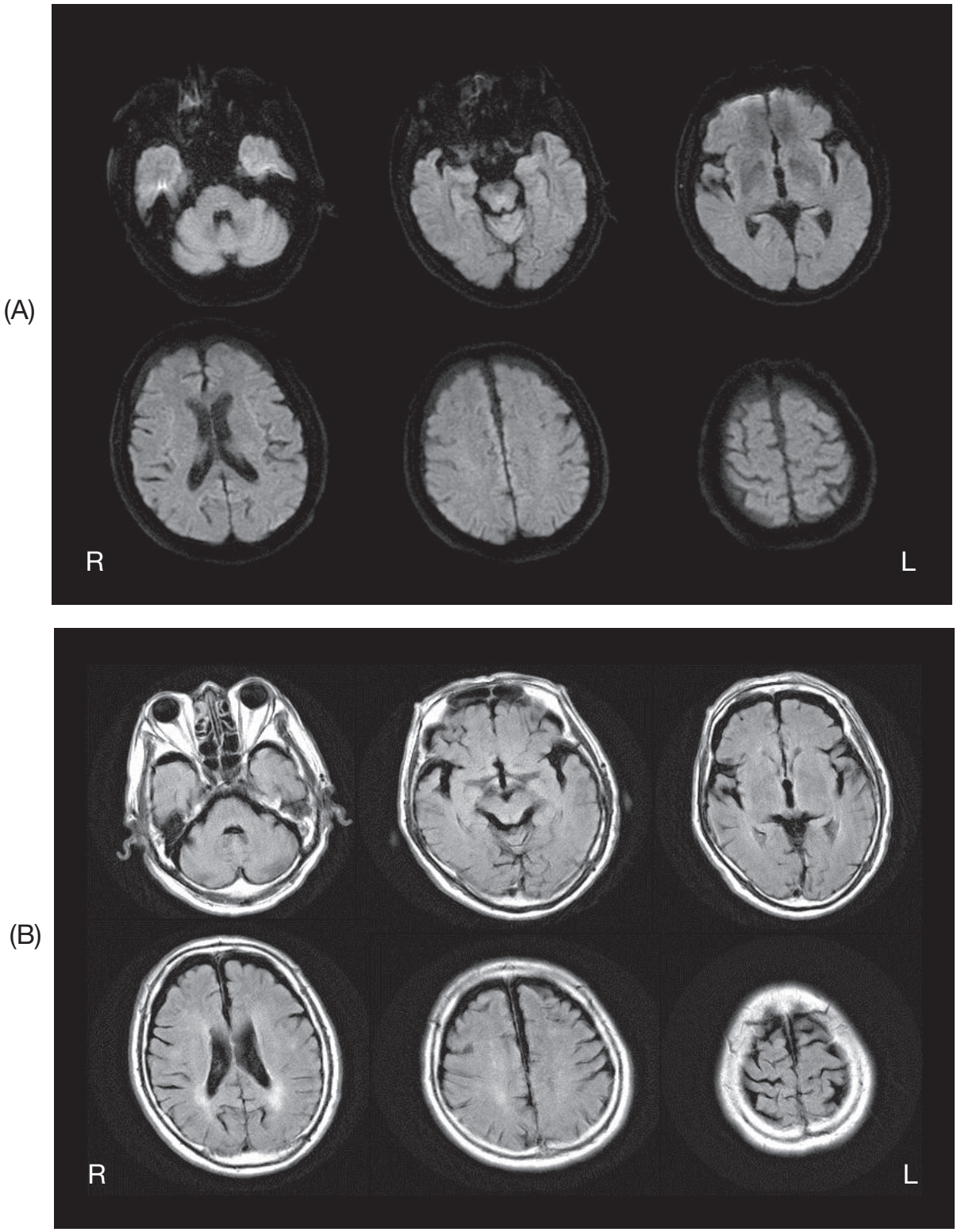

Fig. 1 Routine neuroradiologic images.
No abnormality is apparent on the diffusion-weighted MR image (A) and FLAIR image (B) obtained initially.

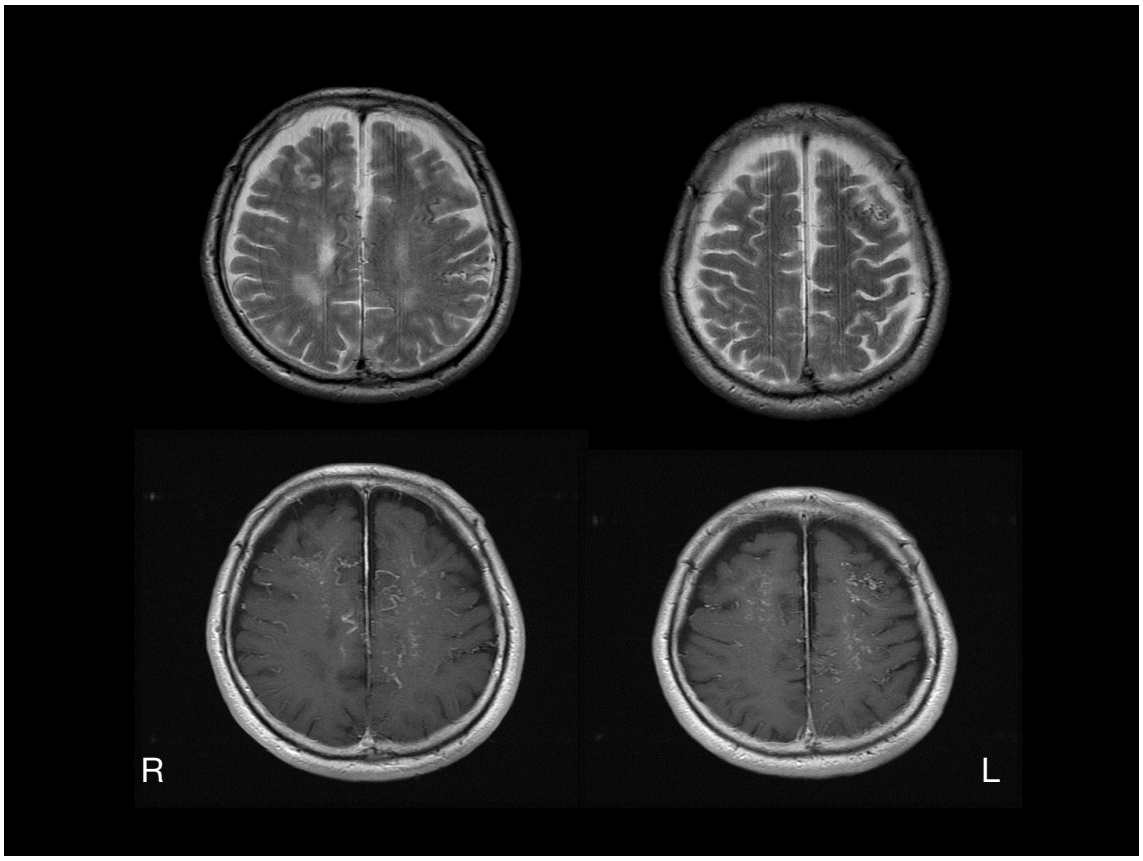

Fig. 2 Detailed MR images.
T2-weighted (top) and contrast-enhanced T1-weighted (bottom) MR images show dilated cortical veins in both cerebral hemispheres.

bral hemispheres (Fig. 2). Thus, DAVF was suspected, and superior sagittal sinus (SSS) DAVF was confirmed by conventional angiography. The lesion was fed bilaterally by the superior temporal arteries and middle meningeal arteries and unilaterally by the left occipital artery with delayed arterial circulation and cortical venous reflux mainly in both frontal lobes (Fig. 3). Iodine-123-N-isopropyl-p-iodoamphetamine single photon emission computed tomography (IMP-SPECT) revealed hypoperfusion of both frontal lobes (Fig. 4). The patient was treated by endovascular embolization using 20% n-buchyl-cyanoacrylate; during treatment, there was no angiographic evidence of dangerous anastomosis. Although the shunts did not completely embolized, delayed arterial circulation in frontal lobes was markedly improved after endovascular treatment. Then, surgical interruption of all feeding arteries was added. The patient's cognitive function was restored over the next 4 weeks, and repeat administration of the MMSE yielded a score of 23.

DISCUSSION

Embolization of SSS DAVF recovered arterial circulation in both frontal lobes in our patient. Our patient's rapidly progressive dementia improved after SSS DAVF treatment. Other possible causes of the dementia, such as encephalitis, metabolic encephalopathy, neoplasm, and epilepsy, were ruled out on the basis of laboratory, neurophysiologic, and neuroradiologic tests. Thus, we surmised that the rapidly progressive dementia in our patient was caused by the DAVF. The influx of arterial blood in the SSS was thought to bring about venous hypertension, leading to bilateral frontal hypoperfusion. This pathophysiologic mechanism was confirmed by IMP-SPECT.

Progressive dementia has been reported as a clinical manifestation of DAVF [2–8, 11–15]; progressive dementia is not uncommon, especially in elderly patients with DAVF [2–9, 11–12, 15]. Most dementias caused by DAVF can be diagnosed by routine MRI. However, routine MRI does not rule out the presence of a DAVF [6, 15]. In our patient, initial routine MRI study, including DWI, FLAIR, MRA, and MRV, failed to show the presence of a DAVF. Only detailed MRI was able to detect the cortical vein ectasia suggestive of DAVF. In examining patients with rapidly progressive dementia, we should be aware of the possibility of DAVF as the cause, and we should perform detailed neuroradiologic examinations including high-resolution T2WI, contrast-enhanced T1WI, and conventional angiography, even if routine MRI findings appear to be normal or only minimally abnormal.

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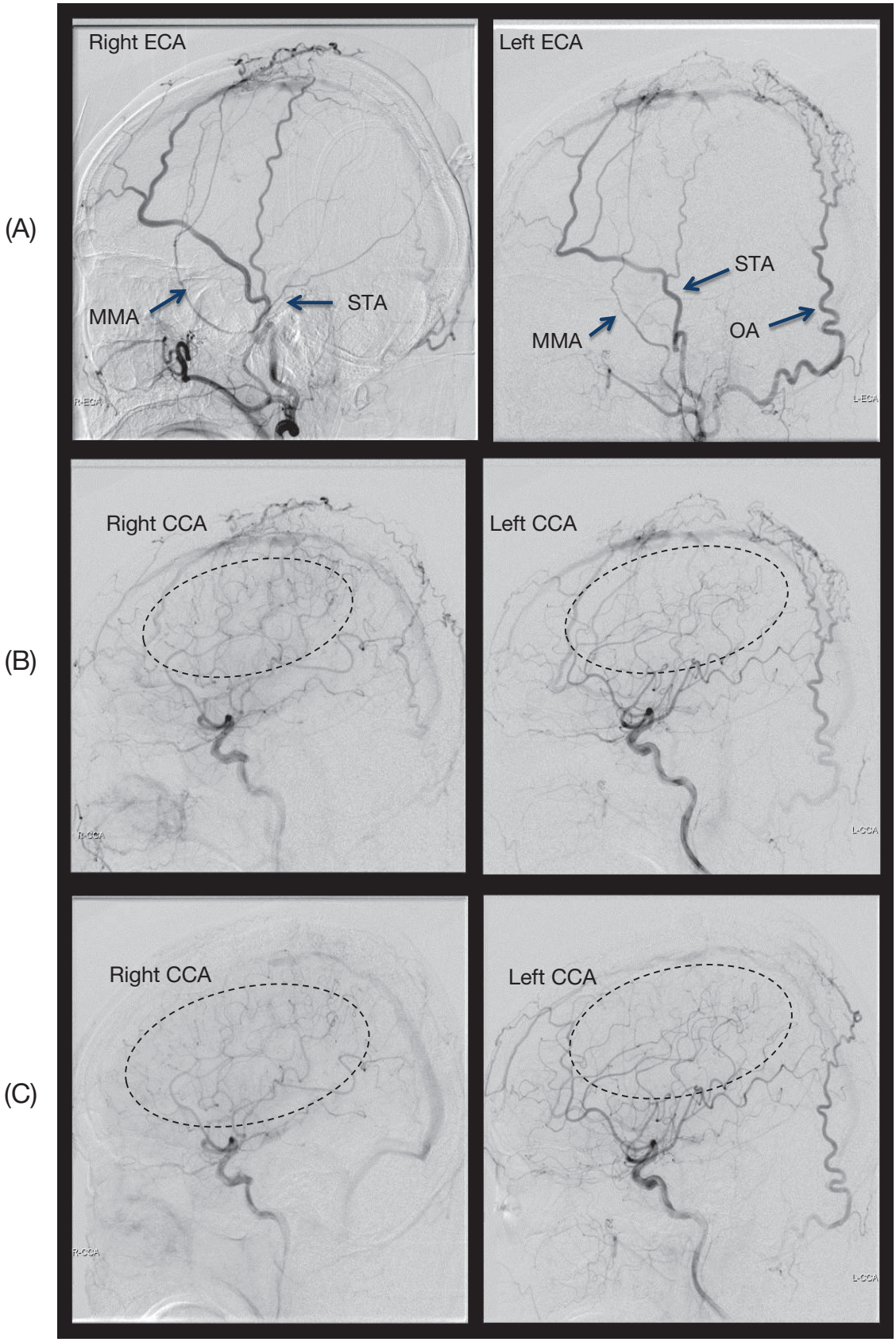

Fig. 3 Carotid angiograms.
External carotid angiogram (A) depicts the arteries feeding the arteriovenous fistula in the superior sagittal sinus: the middle meningeal artery (MMA) and superficial temporal artery (STA) (bilaterally) and the left occipital artery (OA) (unilaterally). Multiple shunt points are seen around the superior sagittal sinus. Common carotid angiogram demonstrated delayed arterial circulation mainly in both frontal lobes (dotted circle) (B). After endovascular treatment, the delayed arterial circulation is markedly improved despite incomplete embolization (dotted circle) (C). Right ECA = right external carotid angiogram; Left ECA = left external carotid angiogram; Right CCA = right common carotid angiogram; Left CCA = left common carotid angiogram.

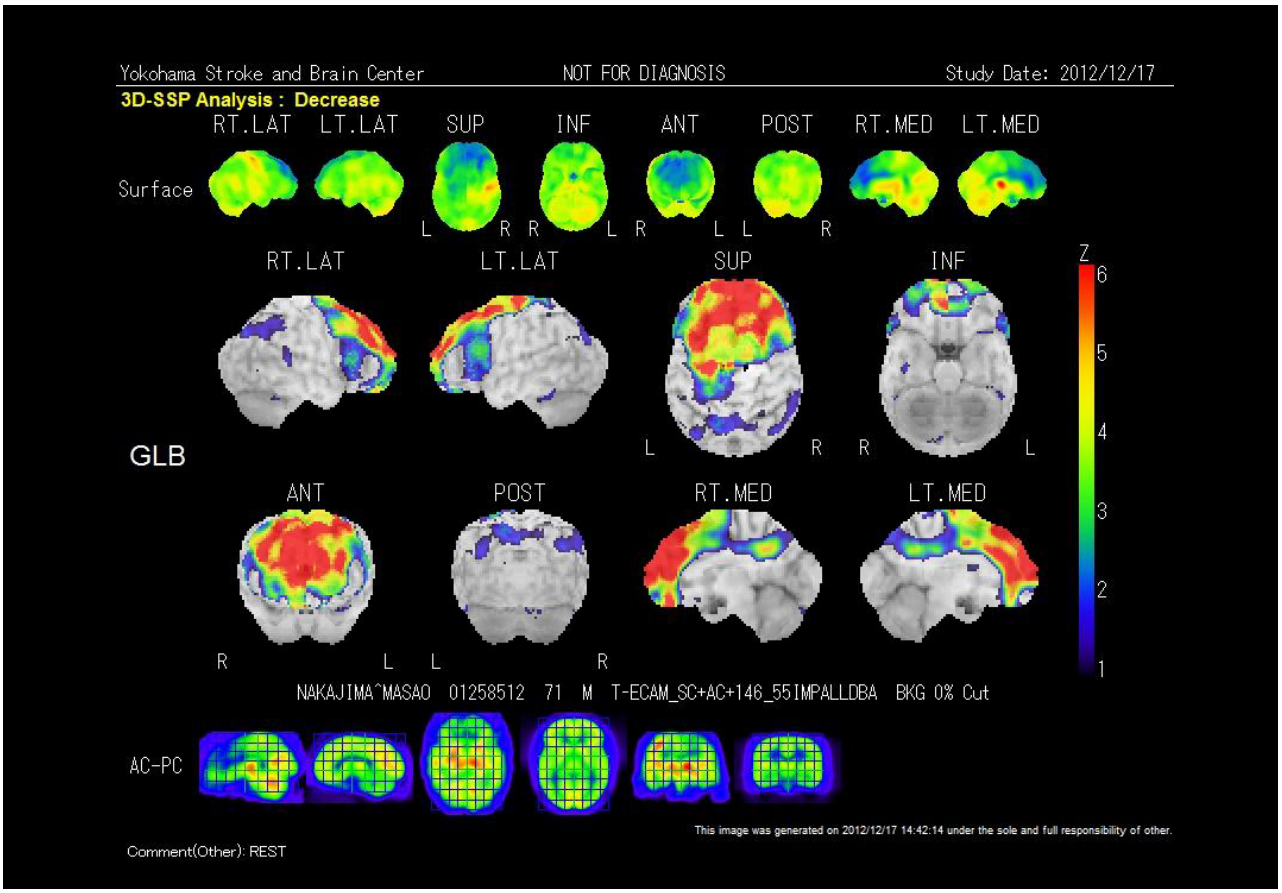

Fig. 4 SPECT perfusion images.
Brain 3D-stereotactic surface projection IMP-SPECT analysis revealed hypoperfusion in both frontal lobes.

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