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Short communication

Low signal intensity in U-fiber identified by susceptibility-weighted imaging in two cases of progressive multifocal leukoencephalopathy

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ABSTRACT

Magnetic resonance imaging (MRI) is a useful tool for diagnosing and monitoring progressive multifocal leukoencephalopathy (PML). Although characteristic MRI findings of PML are well known, we noted a potential new finding for this disease on susceptibility-weighted imaging (SWI). Two patients with PML were studied and followed using MRI. SWI revealed low signal intensities in U-fibers adjacent to the white matter lesions of PML. These findings progressed along with the disease progression. The cause underlying these findings remains unclear. This new finding suggests that SWI is useful for the diagnosis of PML. It can provide a helpful clue in a clinical setting.

1. Introduction

Progressive multifocal leukoencephalopathy (PML), a progressive demyelinating disease of the brain caused by oligodendrocyte, astrocyte, and neuron damage resulting from reactivation of JC virus (JCV) [1], was once regarded as a fatal disease characterized by progressive neurological deficits, leading to death within 2.5–4 months of diagnosis in the absence of treatment [2]. Recently however, reports have described that in certain clinical conditions such as natalizumab therapy in multiple sclerosis patients, an early PML diagnosis, preferably in a presymptomatic/asymptomatic stage, but even in a symptomatic stage, is associated with a favorable prognosis [3,4]. The diagnosis of PML is based on clinical information and detection of JCV in cerebrospinal fluid (CSF) or brain tissue, but a combination of specific findings from magnetic resonance imaging (MRI) enables us to suspect PML, including multifocal white matter lesions involving U-fibers, faint or absent peripheral enhancement of lesions, minimal or no mass effect, and hyperintensity on the periphery of lesions on diffusion-weighted imaging (DWI) [5,6]. Regarding the diagnosis of PML, the official American Academy of Neurology (AAN) criteria on PML diagnosis were proposed recently based on a combination of data related to clinical features, imaging findings, and CSF polymerase chain reaction (PCR) JCV [7]. For PML diagnosis, MRI is therefore regarded as an important tool. To date, no report has described specific findings for PML obtained using susceptibility-weighted imaging (SWI). This report is the first of a potential new finding by SWI in two patients with PML.

2. Case reports

2.1. Case 1

A 56-year-old man treated for repeated erysipelas and otitis externa was hospitalized with left paresis. Neurological examination showed left unilateral spatial neglect. Initial brain MRI revealed a hyperintense lesion on DWI in the subcortical lesion of the right occipital lobe. The initial diagnosis was acute brain infarction. He was treated with aspirin and edaravone for 2 weeks. He subsequently experienced repeated infection, such as that of *Pneumocystis carinii* pneumonia. The left paresis progressed gradually. Serological blood tests revealed previously unsuspected infection with human immunodeficiency virus (HIV). The patient was admitted to our hospital for additional examination and treatment. The findings were highly suggestive of PML. Therefore, lumbar puncture was performed. PCR testing of CSF showed positive results for JCV in the Laboratory of the National Institute of Infectious Diseases (201,750 copies/mL). The patient condition deteriorated progressively. Follow-up MRI performed at 22, 77, and 105 days after the first MRI

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examination confirmed the lesion progression. MRI findings from DWI, T1-weighted imaging, and 3D fluid-attenuated inversion recovery (FLAIR) were all typical for PML, but SWI identified multiple low signal intensity lesions in U-fibers adjacent to PML white matter lesions (Fig. 1). Based on the official AAN criteria for PML diagnosis [7], a definite diagnosis of PML was made. The patient was treated with highly active anti-retroviral therapy, but developed immune reconstitution inflammatory syndrome. Despite intensive medical treatment, the general condition of the patient deteriorated daily. He died 141 days after admission. His family did not consent to autopsy.

2.2. Case 2

A 78-year-old woman with chronic renal failure and hypopharyngeal cancer visited the Neurology Department because of myoclonic seizures in the upper left limb. Brain MRI showed a subtle white matter lesion in the right precentral gyrus (Fig. 2G). Five months later, the patient returned complaining of progressive palsy of the upper left limb. MRI showed multifocal subcortical high signal lesions on 3D FLAIR and DWI. SWI results showed multifocal low signal intensities in U-fibers adjacent to white matter lesions (Fig. 2). She was treated with steroids because

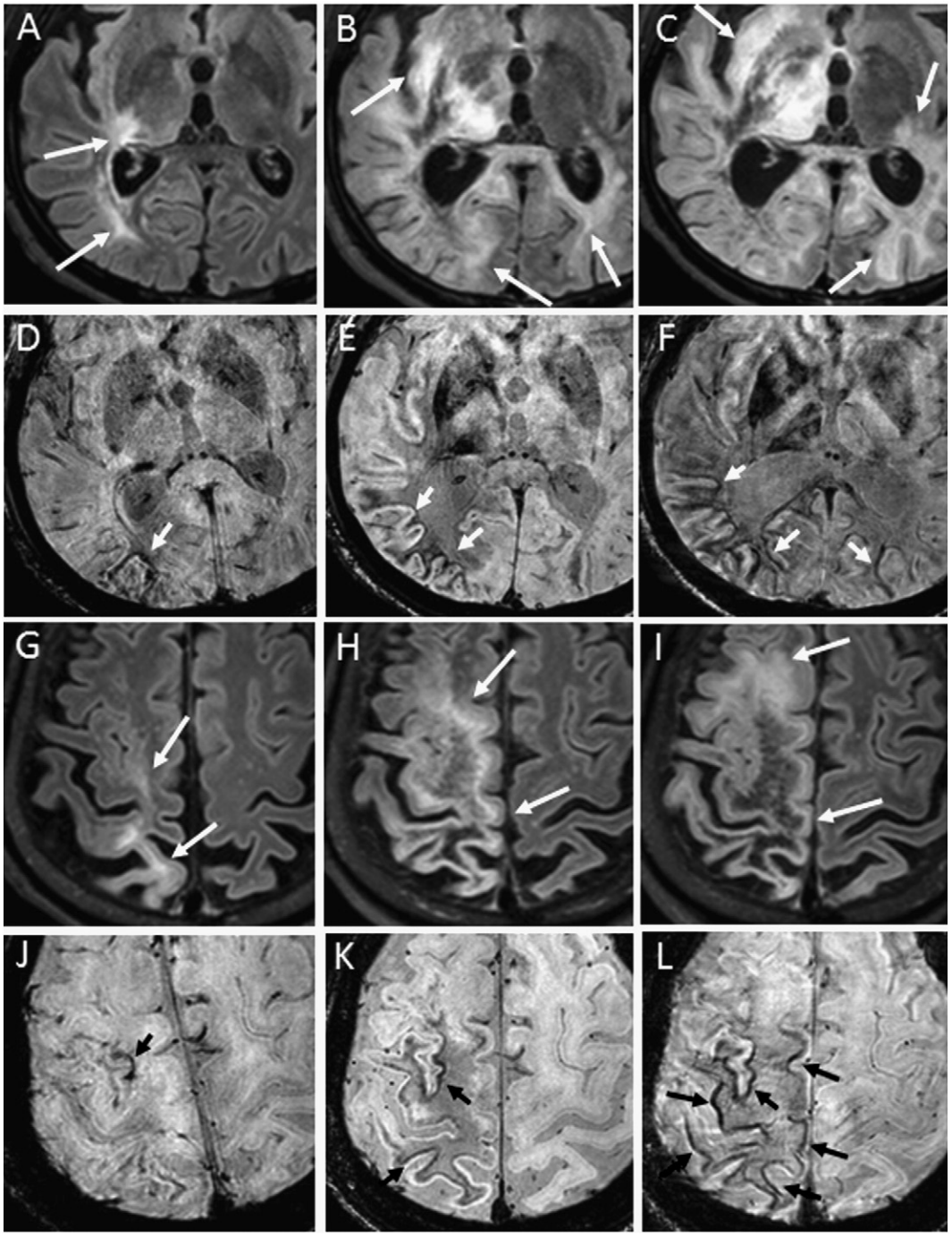

Fig. 1. MRI in Case 1. FLAIR (A–C, G–I) and SWI (D–F, J–L). MRI on day 22 after the initial MRI examination (A, D, G, J), and on day 77 (B, E, H, K) and day 105 (C, F, I, L). FLAIR shows a progression of white matter PML lesions (long arrows). SWI reveals progression of low signal intensities in U-fibers adjacent to white matter PML lesions (short arrows). Low signal intensities of the right thalamus and basal ganglia progress on SWI (D–F).