Case Report

Multiple Cerebral Hemorrhagic Lesions Depicted by Susceptibility-Weighted Imaging in a Patient with Down Syndrome: Case Report

Kei Yanai, MD,* Yasuo Ishida MD, PhD,† Hajime Nishido, MD,* Shinya Miyamoto, MD, PhD,* Kazuto Yamazaki, MD, PhD,† and Katsumi Hoya, MD, PhD*

Our objective is to study a 53-year-old woman with Down syndrome presented with massive lobar hematoma in the left fronto-parietal lobe, and who underwent craniotomy and hematoma evacuation. Histopathological diagnosis of surgical specimen was amyloid angiopathy. Postoperative magnetic resonance studies were performed. The lesion this time showed mixed intensity on susceptibility-weighted imaging. In addition, multiple hypointense lesions were evident. An old previously unidentified hemorrhage in the right temporo-parietal lobe was accompanied by superficial cortical siderosis. Old bleeds were apparent in subcortical areas. These various kinds of hemorrhagic lesion were consistent with findings of amyloid angiopathy reported in the elderly. Most reported cases of Down syndrome associated with intracerebral hemorrhage have involved middle-aged patients. Magnetic resonance studies for Down syndrome patients before old age may disclose the degree to which amyloid angiopathy progresses in the brain of these patients.

Key Words: Down syndrome—amyloid angiopathy—susceptibility-weighted imaging

Introduction

Patients with Down syndrome (DS) show deposition of amyloid within the brain parenchyma, inducing early onset of Alzheimer's disease. Amyloid deposition also occurs in the cerebral vasculature in these patients.[1] This pathological process leads to amyloid angiopathy (AA), a well-known condition in the elderly and patients with Alzheimer's disease.[2] Intracerebral hemorrhage often occurs in elderly patients with AA, who reportedly show hypointensities on susceptibility-weighted imaging (SWI) as a form of magnetic resonance (MR) imaging.[3] Reports on MR images of histopathologically confirmed AA in DS patients are rare.

Case Report

A 53-year-old woman with DS presented with sudden right hemiparesis and total aphasia. Computed tomography showed massive lobar hematoma in the left fronto-parietal lobe. Computed tomography angiography did not show any vascular anomalies. The patient underwent craniotomy and hematoma evacuation. A specimen of brain from near the hematoma was obtained for pathological investigations. Histologically, Direct Fast Scarlet staining revealed amyloid deposition in the walls of small arteries, compatible with the diagnosis of AA (Fig 1, A, B).

Postoperative MR studies were performed at 3 T (Discovery MR750; GEHC, Milwaukee, WI). Left fronto-parietal hematoma was depicted as mixed intensity on SWI

From the *Department of Neurosurgery, Teikyo University Chiba Medical Center, Ichihara, Chiba, Japan; and †Department of Pathology, Teikyo University Chiba Medical Center, Ichihara, Chiba, Japan.

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Address correspondence to Kei Yanai, MD, Department of Neurosurgery, Tokyo University, 7-3-1, Hongo, Bunkyo-ku, Tokyo 113-8655, Japan. E-mail: yanai-tki@umin.ac.jp.

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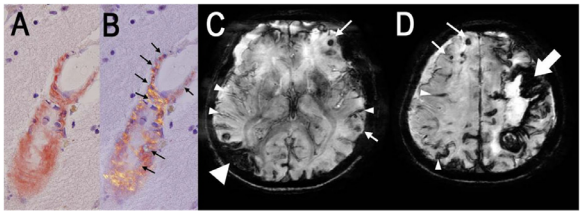

Figure 1. *(A, B) Direct Fast Scarlet staining for amyloid shows deposition of acidophilic substances in a small cerebral artery (A), with apple-green birefringence (B, black arrowheads) (original magnification × 100). (C, D) Susceptibility-weighted magnetic resonance imaging. Left fronto-parietal hematoma is depicted as a mixed-intensity lesion (large white arrow). Previously unidentified lobar hemorrhage in the right temporo-parietal lobe (large white arrowhead), superficial cortical siderosis (small white arrowheads) and subcortical bleeds of various sizes (small white arrows) are depicted as hypointensities.*

(T2* 3-dimensional gradient echo; repetition time, 52.2 milliseconds; echo time, 40.1 milliseconds). These images showed a hypointense area in the right temporo-parietal lobe, suggesting previously unidentified lobar hemorrhage. Superficial cortical siderosis was also apparent. Hypointense areas of various sizes, including microbleeds, were seen in subcortical areas (Fig 1, C, D).

Discussion

DS involves trisomy of chromosome 21. The gene for amyloid precursor protein is located on chromosome 21, and trisomy 21 results in overexpression and thus induction of AA.[1] MR images of cerebral AA in DS have been shown in a small number of reports.[4-6] The present case appears to represent the first report of MR images of pathologically confirmed AA in a DS patient.

SWI offers very sensitive depictions of intracerebral hemorrhage as hypointensity.[3] In addition to hematoma in this case, SWI showed various hemorrhagic lesions in the brain, including old lobar hemorrhage, superficial cortical siderosis and subcortical lesions, including microbleeds. These findings are consistent with descriptions of AA in elderly patients.[2]

Most reports of DS with intracerebral hemorrhage, including the present case, have involved middle-aged patients.[7-10] MR studies including SWI appear significant for middle-aged DS patients to evaluate the degree to which AA is progressing in the brain.

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