Case Report

Single Photon Emission Computed Tomography (SPECT) in a Patient with Wilson's Disease

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Single photon emission computed tomography (SPECT) with ^{133}Xe inhalation was studied in a patient with Wilson's disease, who had the low-density lesions in the bilateral ganglia and a small calcified lesion in the left basal ganglia on computed tomography (CT). SPECT showed the regional cerebral blood flow (rCBF) of the basal ganglia area. The values of the rCBF in these areas are 77 to 91 ml/100g/min in the right, and 56 to 77 ml/100g/min in the left. Further study is needed to prove the pathological role of rCBF for the basal ganglia lesion in the patient with Wilson's disease.

Key Words: Regional cerebral blood flow, Computed tomography, Calcification

Although computed tomography (CT) has shown low-density areas in the basal ganglia of patients with Wilson's disease, regional cerebral blood flow (rCBF) has not been adequately studied. In a patient with Wilson's disease, with such low-density lesion in the bilateral basal ganglia and a small calcified lesion in the left basal ganglia, single photon emission computed tomography (SPECT) was therefore performed using ^{133}Xe in order to gain some insight in to the pathogenesis of such lesion.

CASE REPORT

A 18-year-old Japanese girl was seen for liver dysfunction and depression. She had episodes of hemolytic anemia, liver damage, and gall stone two years ago. On examination, she was cheerful under the antidepressant treatment, without any neurological abnormalities. Hepatomegaly and Kayser-Fleischer ring were noted.

Serum copper level was 33 μg/dl, serum ceruloplasmin 3 mg/dl, and urine copper excretion over 1,000 μg/day during the loading of penicillamine 1 gm per day. There was no evidence of parathyroid dysfunction. Serial CT showed low-density areas in the bilateral ganglia with a small calcification in the left, ventricular dilatation, and cortical atrophy (Fig. 1a). SPECT with inhalation of ^{133}Xe was studied according to the method of Shirahata et al. (Fig. 1b)[1].

She became depressive, while the administration of 1 to 1.2 gm penicillamine daily was continued, and she developed finger tremor, marked rigidity, and muscle spasm in her legs. She could not open her eyes without using her fingers. After a year of treatment, neurological signs somewhat improved and stabilized.

DISCUSSION

The calcification in the basal ganglia on CT has been shown to be associated with diseases or may occur independently without known cause. In patients with Wilson's disease, such finding has not been documented so far. Only one case with calcifications in the frontal lobe and dentate nucleus was reported by Chen et al.[2]. The relationship between the calcification and Wilson's disease has therefore remained uncertain. Occur-

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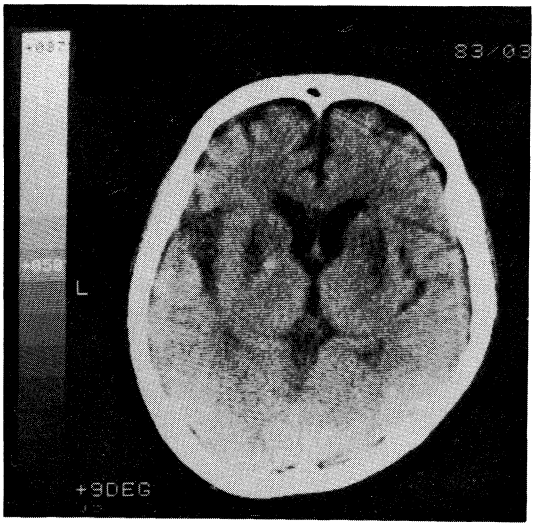

Fig. 1a. CT scan (3 cm above the orbitomeatal plane, slice thickness 1 cm) showing low-density areas in basal ganglia, a small calcification in the left basal ganglia.

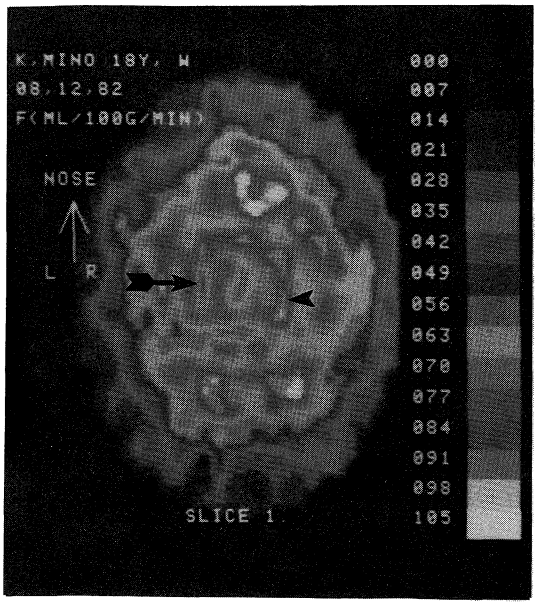

Fig. 1b. Tomographic rCBF by SPECT with ^{133}Xe inhalation through the plane (3 cm above the orbitomeatal plane, 2 cm thickness). Arrow and arrow head showing rCBF of basal ganglia.

rence of calcification of the basal ganglia would be readily explained in a patient with association of Wilson's disease and hypoparathyroidism[3]. But such is evidently not the case in this patient. Although it is possible that cerebral vascular accident is responsible for the calcification, but symptoms and signs supporting it are lacking.

Neuronal loss and consequent cavitation are thought to be a cause of the low-density lesion. Recently, improvement of low-density on CT by the treatment suggest that extensive loss of dendrites of glial cells and edema are the causes for the low-density lesion[4]. No evidence, however, is available as to whether or not rCBF change in these lesion.

Although spacial resolution of SPECT is not as high as that of CT, SPECT is an advanced method for detecting rCBF in the deep region of the brain. In the present, the comparison between CT and SPECT suggests that the rCBF of the areas, showing with arrow and arrow head in Fig. 1b, might represent the rCBF of the basal ganglia. The value of rCBF in the right, arrow head shows, is 77 to 91 ml/100 g/min and in the left, arrow shows, 56 to 77 ml/100g/min. Although averaged rCBF of the cerebral hemisphere of normal adults in the slice, 3 cm above the orbitomeatal plane, was 58± 8 ml/100g/min[1], there is no available data to compare with our data.

Further study is needed to prove the pathological role of the rCBF for the lesion of the basal ganglia in the patient with Wilson's disease. It is known that the low-density lesions are improved by the treatment in some cases with Wilson's disease and in the others not. No adequate explanation on such difference has got yet. Study by SPECT may offer further information as to discrepancy.

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