

Cerebral blood flow-SPECT in a patient with Sneddon's syndrome

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We report a 50-year-old woman diagnosed with Sneddon's syndrome and examined by CBF scintigraphy several times for follow-up of the disease. There were no significant changes in her CBF scintigraphic findings or neurological status during the 6-year follow-up period. Sneddon's syndrome is a slowly progressive disorder in which livedo reticularis precedes cerebrovascular accidents. Because small cortical arteries are predominantly affected in Sneddon's syndrome, MR and conventional angiography often fail to show any abnormal findings, and MR imaging may not visualize decreased CBF in the early stage. Therefore, CBF scintigraphy should be performed in patients who have or are suspected of having Sneddon's syndrome.

Key words: cerebral blood flow scintigraphy (CBF scintigraphy), I-123-IMP, Tc-99m-ECD, Sneddon's syndrome

INTRODUCTION

SNEDDON'S SYNDROME is a rare disease characterized by livedo reticularis, hypertension, and cerebral blood vessel damage.¹⁻³ It is thought to be an antiphospholipid antibody syndrome because patients are positive for antiphospholipid antibodies such as anticardiolipin antibody.⁴⁻⁶ Here we report a case of Sneddon's syndrome in which cerebral blood flow (CBF) scintigraphy was performed for follow-up of the disease.

CASE REPORT

A 50-year-old woman developed a motor deficit in the right arm with speech disturbance on February 7, 1991. When the symptoms failed to improve, she consulted the department of internal medicine of our hospital. Initial examination demonstrated marked hypertension (250/120 mmHg). Net-like purpura was evident on the forearms and thighs (Fig. 1), and the skin lesions had persisted for several years. Brain CT demonstrated a low attenua-

tion area in the left corona radiata (Fig. 2), and the patient was immediately admitted with a diagnosis of cerebral infarction. T2-weighted MR images taken after admission demonstrated a high-signal intensity area extending from the left corona radiata to around the lateral ventricle (Fig. 3). Cerebral angiography demonstrated occlusion of the sphenoidal portion (M1) of the left middle cerebral artery, but the blood flow to the area was maintained through a collateral pathway from the anterior cerebral artery and the external carotid artery. I-123-N-isopropyl-p-iodoamphetamine (IMP) SPECT images taken during the same period showed blood flow reduction in the left temporal lobe and parietal lobe, and the area detected was larger than that shown by MR imaging (Fig. 4). Laboratory data showed positivity for two anti-cardiolipin antibodies, IgM antibody and IgG antibody. A diagnosis of Sneddon's syndrome was made based on the laboratory data and the presence of livedo reticularis, a cerebrovascular disorder, and hypertension. The patient responded well to antihypertensive and anticoagulant therapy after admission, and her symptoms improved.

The patient was discharged one month later, but the purpura on the arms and thighs persisted. She was followed up carefully because of the possibility of a major stroke and intellectual impairment. CBF scintigraphy was performed twice with I-123-IMP and once with Tc-99m-ethyl cysteinyl dimer (ECD) during a 6-year follow-up period. The scintigraphic findings did not change

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significantly compared with the findings at the time of initial diagnosis. Quantitative analysis by Tc-99m-ECD showed almost normal mean cerebral blood flow (42 ml/100 g/min), but decreased regional CBF was observed in the left temporal and parietal lobe (Fig. 5). MR images and MR angiography 6 years after discharge also showed no major changes compared with the studies at the time of the initial diagnosis. No neurological deficits have developed thus far.

DISCUSSION

In 1965, an English dermatologist, J.B. Sneddon, reported 6 cases of a syndrome which involved unstable hypertension and cerebral ischemia accompanied by livedo reticularis. He considered it to be a type of arterio-occlusive disorder similar to Takayasu's arteritis,¹ and ever since the disease has been called Sneddon's syndrome. Sneddon's syndrome is a rare disease, and to our knowledge less than 200 cases have been reported in the literature.¹⁻¹¹ The small number of cases may reflect unfamiliarity with the syndrome rather than its true incidence. Age at the first cerebrovascular accident has ranged from 18 to 81 years old, and young women are predominantly affected. Various persistent neurological symptoms, especially sudden motor deficit, aphasia, and dysarthria may occur. Cerebral blood vessel damage in Sneddon's syndrome often progresses slowly, and almost half of the patients develop functional disability and intellectual impairment. There is no specific treatment for this syndrome, but symptomatic treatment with such as antihypertensive agents, anticoagulants and antiplatelet drugs, may prevent dementia. Sneddon's syndrome is thought to be a treatable form of dementia.⁷

Livedo reticularis results from irregular focal and persistent impairment of blood flow that is physical in nature, such as in arterial occlusion in atherosclerosis, vasculitis or Sneddon's syndrome. Recently, characteristic histopathological findings have been reported in skin biopsies from Sneddon's syndrome patients. They consist of inflammatory changes in the endothelium of small arteries followed by subendothelial cell proliferation leading to partial or complete occlusion.⁸ In almost all cases of Sneddon's syndrome, livedo reticularis precedes the neurological symptoms by several years.^{1-3,8,9} In our patient, similar net-like red and purple purpura was noted on the forearms and thighs before the cerebrovascular accident. Recognition of livedo reticularis is therefore important in the diagnosis of this syndrome in the early stage. Patients who have livedo reticularis should undergo skin biopsy.⁸ If examination of the skin biopsy specimen suggests Sneddon's syndrome, the patient should be evaluated for cerebral blood vessel damage. It is important to prevent, or at least delay, the onset of irreversible cerebral damage.⁷ Unfortunately, our patient was diagnosed after a cerebrovascular accident, but no irreversible cerebral

damage has occurred.

Left MCA occlusion and infarction of the left corona radiata were observed in our patient, but these are rare findings in Sneddon's syndrome. The cerebral blood vessel damage in this syndrome is characterized by microembolism and a hypercoagulable state in the end-perfusion territory of cortical arteries.^{8,9} It is considered that a thrombogenic factor is located in the vessel lumen in this syndrome, rather than in the vessel wall, and therefore small arteries are affected more often than large arteries in Sneddon's syndrome.⁹ The region of the cerebral arteries affected is different from that in atherosclerotic stenosis.

Stockhammar et al. reported MR angiographic findings of Sneddon's syndrome. They observed patency of the carotid arteries, basilar artery, circle of Willis, and proximal portions of the cerebral arteries in 16 of 17 patients.⁸ Tourbach et al. reported that the basal ganglia, which are usually affected in leukoencephalopathy associated with hypertension or other vascular risk factors, were impaired in only 8 of 26 patients (30%) in his series.⁹ Therefore, an imaging method that can visualize CBF is required to evaluate cerebral involvement of Sneddon's syndrome, and CBF scintigraphy seems to be suitable for this purpose.

In some cases of Sneddon's syndrome, CBF scintigraphy showed blood flow reduction in areas where no abnormalities were evident on MR imaging.^{10,11} It has been also reported that CBF scintigraphy can detect abnormal findings at an early stage of the disease, before they can be visualized by CT or MR imaging.¹¹ CBF scintigraphy in the present case also showed blood flow reduction in areas broader than the infarcted areas revealed on MR images. Quantitative measurement of CBF can provide objective results. Unfortunately, quantitative measurement of CBF was only performed once in our patient. It would be useful to detect minimal changes in regional CBF during further follow-up. Because cerebral blood vessel damage in Sneddon's syndrome often progresses slowly, patients need to be followed up carefully on an outpatient basis over a long period. The examination method employed should be reproducible and should not be invasive. CBF scintigraphy, especially quantitative measurement of CBF, is also suitable for this purpose.

In conclusion, CBF scintigraphy should be performed whenever Sneddon's syndrome is present or suspected.

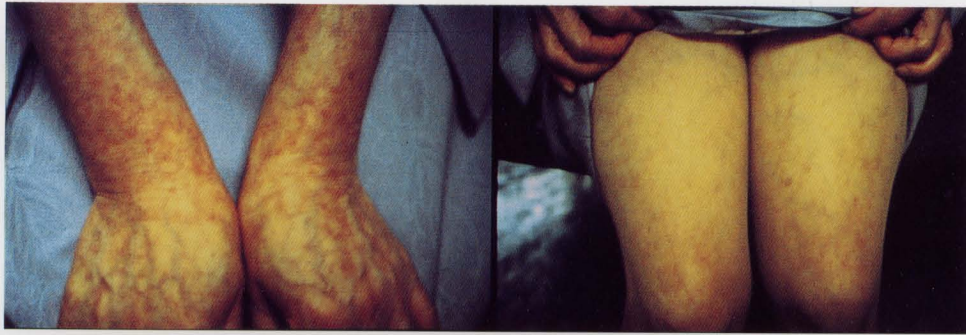


Fig. 1 Photographs of the skin showing livedo reticularis of the forearms and thighs.



Fig. 2 Brain CT before hospitalization demonstrating a low attenuation area in the left corona radiata.

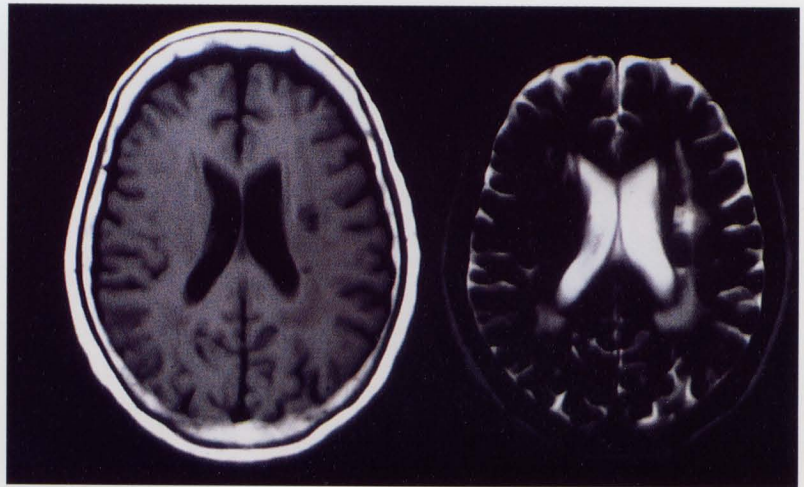


Fig. 3 T1-weighted (left) and T2-weighted (right) MR images obtained at the time of the initial diagnosis demonstrate multiple infarctions in the left corona radiata.

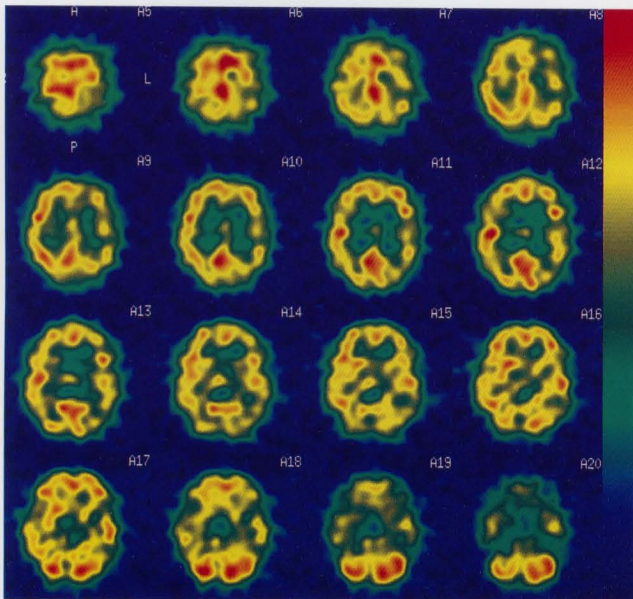


Fig. 4 I-123-IMP SPECT images obtained at the time of the initial diagnosis show reduction of left temporal and parietal perfusion and detect an area broader than shown by MR imaging.

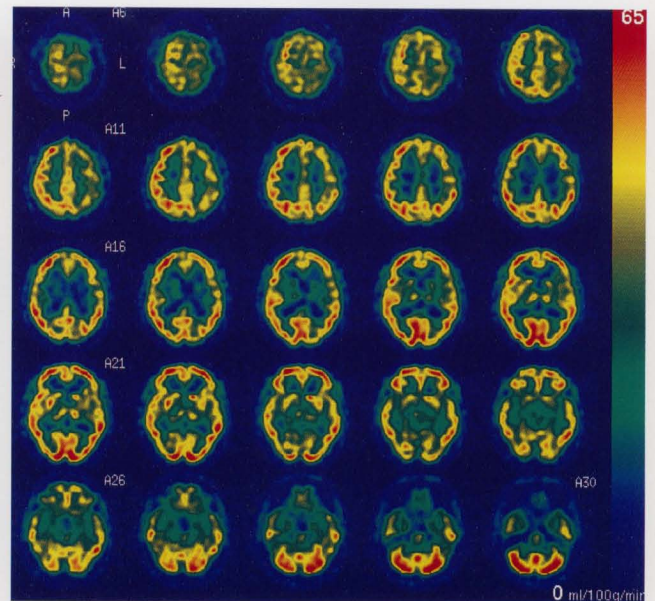


Fig. 5 Tc-99m-ECD SPECT images obtained at 6 years later show blood flow reduction in the region extending from the left temporal lobe to the parietal lobe. There were no interval changes compared with Figure 4, except for the prominence of hypoperfusion in the left parietal lobe.

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