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Brain Perfusion SPECT in Lyme Neuroborreliosis

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SPECT imaging brain perfusion using ^{99m}Tc-HMPAO was performed on a 38-yr-old woman with Lyme neuroborreliosis confirmed by autopsy. The patient had been suspected of spinocerebellar degeneration. Cerebral blood flow was diffusely decreased throughout cerebral cortices but cerebellar blood flow was not impaired, which indicated that the diagnosis was unlikely spinocerebellar degeneration. These findings suggested that brain perfusion SPECT provides useful information in diagnosing the patients with Lyme neuroborreliosis, especially when spinocerebellar degeneration is included in the differential diagnosis.

Key Words: Lyme neuroborreliosis; SPECT; HMPAO; spinocerebellar degeneration

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Lyme disease is a multisystemic disease caused by tick-borne spirochete Borrelia burgdorferi, and its invasion into the central nervous system develops a diversity of neurologic and psychiatric disturbances (*1,2*). In Lyme disease, the central nervous system involvement usually becomes involved in the second stage showing meningitis, multiple cranial nerve palsy, motor or sensory radiculoneuritis and polyneuropathy. In more severely affected cases, disseminated cerebromyelitis, leukoencephalitis and demyelinating encephalopathy have been found, which characterize the third stage of Lyme disease, known as Lyme neuroborreliosis (LNB). Findings in imaging studies with brain CT and MRI are frequently subtle and nonspecific (*3,4*).

This article describes previously unreported HMPAO-SPECT findings in a patient with LNB.

CASE REPORT

A woman, who had been in good health, became aware of unstable walking at 37 yr. She had not experienced any signs or symptoms of skin lesion or joint pain. She consulted a general hospital and was neurologically examined, but no abnormality was detected. However, her neurological symptoms gradually developed. Her gait became definitely ataxic, and she could not stand up for a long time. She had choreoathetotic movement in her upper extremities, spoke explosively and had dysdidochokinesis. Five months later, she could not stand up and memory disturbance became evident with inertia, and she was admitted to our hospital.

Neurological examination disclosed marked disturbance in coordination and dysmetria including intentional tremors. She was frequently drowsy, restless and disorientated as to time and place. She responded very slowly to any questions. No meningeal signs were detected. Mini-Mental State Examination score was 16/30 (*5*). Her ocular movements had limitation on upward and downward gaze. Her pupil was miotic but reactive. She was judged to have supranuclear bulbar palsy because of the absence of pharyngeal reflex. She had muscle weakness detected in general skeletal muscles that were hypotonic. Choreoathetotic movements were present in her upper and lower extremities. Deep tendon reflexes

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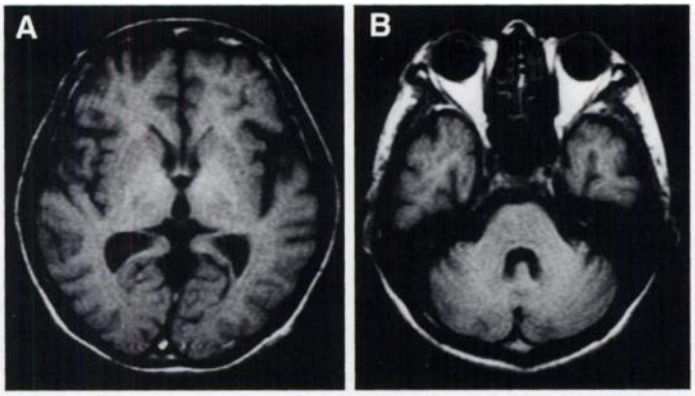

FIGURE 1. Axial MRI T1 weighted image at the level of (A) basal ganglia and (B) cerebellum shows mild brain atrophy. No abnormal signal intensity is detected.

were brisk bilaterally in all extremities with right-sided ankle pseudoclonus and positive Babinski's sign. Sensory disturbance was absent. The laboratory data were almost normal. Her electroencephalogram was normal and cranial CT and MRI showed only mild brain atrophy (Fig. 1). The initial clinical diagnosis was spinocerebellar degeneration (SCD).

The brain perfusion SPECT imaging was performed using a high-resolution SPECT system with three rotating cameras equipped with high-resolution, low-energy, fanbeam collimators. Seven hundred and 40 MBq dose of ^{99m}Tc-HMPAO was injected with patient's eyes closed. Acquisition was started from 10 min postinjection. A total of 90 projection images were obtained in a 128 × 128 matrix over 360° with 50 sec per view, then a series of transaxial slices were reconstructed using a ramp filter with Butterworth filter (order 8, cutoff frequency 0.65 cycles/cm). Attenuation correction was not performed. The first brain perfusion SPECT was normal (Fig. 2). However, the second SPECT performed 6 mo later, when repeated cranial MRI showed only brain atrophy and no abnormal signal intensity, revealed diffusely decreased perfusion in cerebral cortices (Fig. 3). However, cerebellar perfusion was not impaired. Cerebellar hypoperfusion in patients with SCD has been reported (*6,7*). The diagnosis of SCD was considered doubtful from these SPECT findings.

Further laboratory examination of her serum disclosed an elevated titer of antibody against Borrelia burgdorferi by either Western blotting or enzyme-linked immunosorbent assay (11.8 pg/dl), IgM titer was elevated. The review of her past history disclosed that the patient had stayed in an area in Minnesota, where Lyme disease is endemic, for 1 yr when she was 28 yr.

She was treated with high dose penicillin that was effective for her choreoathetosis and reduction in her serum titer of Borrelia burgdorferi. She died of acute cardiac failure at the age of 40. Autopsy revealed a chronic inflammatory change in the brain and the presence of silver-impregnated organisms, which confirms the neuropathological diagnosis for LNB.

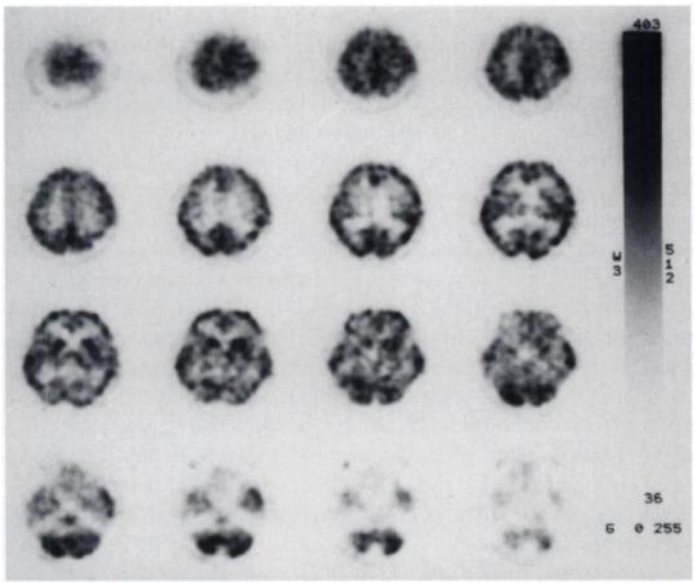

FIGURE 2. Initial brain perfusion SPECT images using ^{99m}Tc-HMPAO shows no apparent perfusion abnormality including cerebellum.

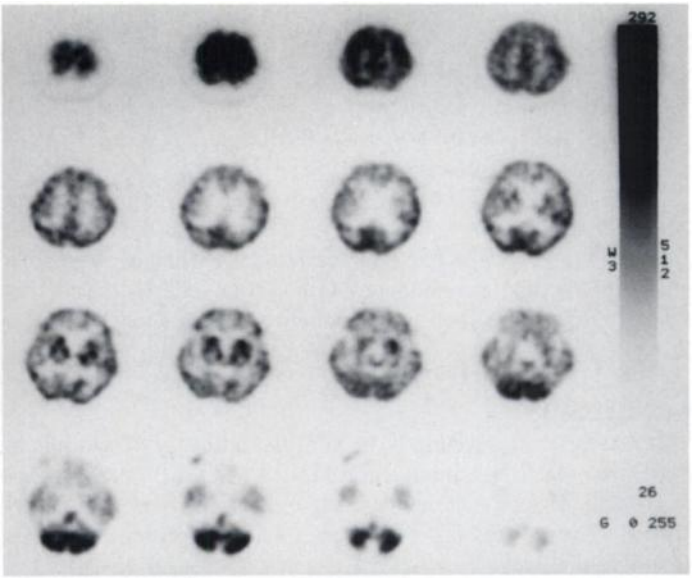

FIGURE 3. Second brain perfusion SPECT images demonstrate diffuse blood flow reduction in cerebral cortices compared to Figure 1. Note that cerebellar perfusion is well preserved.

DISCUSSION

This case demonstrated previously unreported diffuse cerebral cortical flow reduction in a patient with pathologically proven LNB. It is uncertain whether the diffuse cerebral blood flow reduction seen in this case is typical for LNB. The mechanism of central nervous system involvement is not clear, but direct invasion of spirochetes into the cortices and vasculitis caused by spirochetes have been proposed (*8*). By neuropathological examination, focal encephalitic change was not severe in the cerebral cortices. Therefore, vasculitis may have caused the flow reduction in this case.

Early in the course of disease, our patient manifested a clinical syndrome indistinguishable from SCD. However, the

TABLE 1
Common Symptoms and Brain Perfusion in LNB and SCD

	LNB	SCD
Common symptoms		
Ataxia	+	++
Parkinsonism	−	− ~ ++
Cognitive impairment	++	+
Brain perfusion		
Cerebral cortex	↓	→
Cerebellum	→	↓

LNB = Lyme neuroborreliosis; SCD = spinocerebellar degeneration.

information obtained from the SPECT study was different from the cerebellar hypoperfusion typical for SCD (*6,7*). Our data showed that brain perfusion SPECT provided valuable information to the clinician early in the course of this disease with cerebellar ataxia. Brain perfusion SPECT could be of help for differentiating this disease from SCD.

Lyme disease is endemic in New England, pacific states, Minnesota, Wisconsin, Europe, Australia and in some areas in East Asia (*2*). Now increased international exchange of persons and development of transportation systems have spread this disease around the world. Our patient was from Japan.

CONCLUSION

If a patient suspected to have SCD has a history of visiting an endemic area, LNB should be placed in the differential diagnosis and further evaluation with brain perfusion SPECT as well as serologic testing should be considered promptly for this treatable disease.

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Anterior Operculum Syndrome Localized by SPECT

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The aim of this case report was to present a patient with complete anarthria and orofacial apraxia without other relevant neurological deficit. The clinical features are compatible with anterior operculum syndrome. **Methods:** A regional brain perfusion scan was done using ^{99m}Tc-HMPAO and a SPECT gamma camera. A brain CT scan and an MRI were also performed. **Results:** Brain CT and MRI were not diagnostic. On brain SPECT, hypoperfusion of the left inferior area of the frontal lobe was noted. **Conclusion:** The patient studied showed an uncommon case of anterior operculum syndrome of focal degenerative origin localized by SPECT. SPECT may be a useful and effective method for diagnosis of this unusual neurological deficit.

Key Words: anterior operculum syndrome; orofacial apraxia; anarthria; SPECT; isolated cortical atrophy

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The combination of anarthria and orofacial dyspraxia presented as isolated neurological deficits is a very uncommon and little known disorder. Decrease in cognitive functions, abnormal motor ability and praxia, and symptoms indicating motor neuron diseases are the most common neurological findings associated with anarthria and orofacial dyspraxia. These clinical features are described as an anterior operculum syndrome in vascular events, tumors or head trauma, but these findings are uncommonly seen as a focal degenerative disease. Only one report describing three cases of anarthria and orofacial apraxia as isolated symptoms of focal cortical atrophy have been published (*1*).

We describe a patient having focal cortical atrophy with the first published description of the anatomical location pinpointed using the SPECT technique. The finding of an abnormal focal perfusion by SPECT correlated with the anticipated defect.

CASE REPORT

A 72-yr-old man, born in Romania, presented with a 2 yr history of speech and swallowing difficulties. He had suffered from diabetes mellitus for 20 yr. The initial symptom was dysarthric speech after extreme effort without disturbance in language skills. Swallowing difficulties appeared simultaneously. After approximately 1 ½ yr, the patient had complete anarthria and could only eat and drink by using a special maneuver of the lips and jaw.

On examination, complete anarthria was noted. When requested to repeat certain sounds, syllables or full words, an indistinguishable voice was produced either by air flowing from outside into the lungs or vice versa. His comprehension was intact. Basic writing was normal in spontaneous writing, copying or on dictation testing. Word selection was correct in two languages in which the patient was familiar.

Neurological examination showed normal functioning of the first to sixth cranial nerves, including normal eye movements, even on command. When the patient was asked to move his facial muscles, he was not able to mimic any natural facial movements, such as swallowing, kissing or yawning. He could not complete these movements after verbal commands or in the presence of spontaneous coughing or laughing. He could, however, mimic the expression of anger or disappointment and could copy the movements of other persons. Gag reflex was normal, but the patient was not able to swallow under normal circumstances, particularly with liquids, although no nasal regurgitation was observed. To swallow, the patient had to hold the fluid in his mouth, pressing both lips together with his fingers and pushing the jaw posteriorly with his other hand.

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