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.

ACKNOWLEDGMENTS

We thank Dr. T. Masuzawa, Department of Microbiology, School of Pharmaceutical Sciences University of Shizuoka, Japan for confirming the serological diagnosis of the subject.

REFERENCES

1. Halperin JJ, Luft BJ, Anand AK, et al. Lyme neuroborreliosis: central nervous system manifestations. *Neurology* 1989;39:753–759.
2. Finkel MJ, Halperin JJ. Nervous system Lyme borreliosis: revisited. *Arch Neurol.* 1992;49:102–107.
3. Reik LJ, Smith L, Khan A, Nelson W. Demyelinating encephalopathy in Lyme disease. *Neurology* 1985;35:267–269.
4. Fernandez RE, Rothberg M, Ferencz G, Wujack D. Lyme disease of the CNS: MR imaging findings in 14 cases. *AJNR* 1990;11:479–481.
5. Folstein MF, Folstein SE, McHugh PR. Mini-mental state: a practical method for grading the cognitive state of patients for the clinician. *J Psychiatr Res* 1975;12:189–198.
6. Botez MI, Leveille J, Lambert R, Botez T. SPECT in cerebellar disease: cerebellocerebral diaschisis. *Eur Neurol* 1991;31:405–412.
7. Sun X, Tanaka M, Kondo S, Hirai S, Ishihara T. Reduced cerebellar blood flow and oxygen metabolism in spinocerebellar degeneration: a combined PET and MRI study. *J Neurol* 1994;241:295–300.
8. Demaerel P, Wilms G, Lierde SV, Delanote J, Baert AL. Lyme disease in childhood presenting as primary leptomeningeal enhancement without parenchymal findings on MR. *AJNR* 1994;15:302–304.

Anterior Operculum Syndrome Localized by SPECT

Yair Lampl, Adam Steinmetz, Ronit Gilad, Yehiel Eshel, David Chamovitz and I. Sarova-Pinhas

Departments of Neurology and Nuclear Medicine, Edith Wolfson Medical Center, Holon; and Tel Aviv University, Tel Aviv, Israel

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

J Nucl Med 1997; 38:1122–1124

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.

Received May 6, 1996; revision accepted Oct. 2, 1996.

For correspondence or reprints contact: Yair Lampl, MD, Dept. of Neurology, Edith Wolfson Medical Center, Holon 58100 Israel.

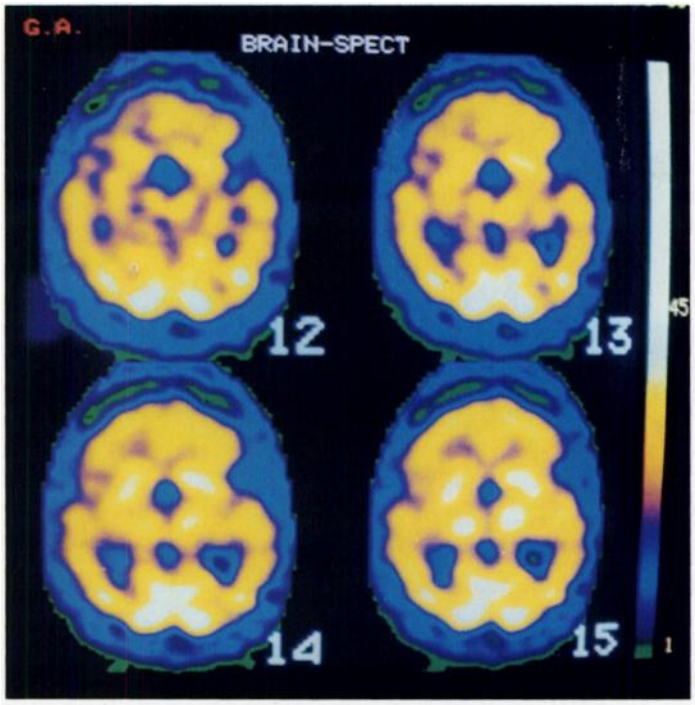

FIGURE 1. SPECT ^{99m}Tc-HMPAO brain image demonstrates perfusion defect in the opercular area and posterior frontal area in the left lobe.

The patient's tongue was of normal size without involuntary quivering; he moved it spontaneously, but not at all on command. His limb muscle strength was also normal. No fasciculations were noted. Sensation was mildly decreased in the limbs in the form of glove and sock distribution areas. Deep tendon reflexes were diffusely decreased. No pathological reflexes were induced, including limb and jaw jerk. Cerebellar function was intact. There was no limb apraxia of any type.

The neuropsychological examination showed: Minimental Score examination = −28/30; Hamilton Depression and Anxiety tests = normal; Wechsler Adult Intelligence Scale: verbal = 90, performance IQ = 105; and Raven's test was scored on the sixtieth percentile.

iagnostic workup included: EEG, which was normal and electrophysiological examination, which showed a decrease of sensory distal latency and nerve conduction velocity in ulnar nerve and the motor conduction velocity in the peroneal nerves. EMG of the first dorsal interossei, deltoid and quadriceps muscles were normal.

A therapeutic trial with neostigmine methyl sulfate 0.5 g i.m. for 3 days was negative. Chest radiograph, blood and CSF examinations, and brain CT and MR scans were normal.

A regional brain perfusion scan was performed after intravenous injection of 800 MBq (22 mCi) ^{99m}Tc-HMPAO while reducing visual and acoustic stimuli to minimum. SPECT acquisition began 60 min after injection on rotating gamma camera with low-energy, high-resolution collimator (APC-45). Data were acquired in 64 × 64 pixel matrix, 60 frames over a 360° orbit 40 sec each frame. Raw data were corrected for uniformity and decay, and then filtered with a two-dimensional Metz filter set at 14 mm FWHM (fifth order). These prefiltered data were reconstructed into transaxial slices with ramp filter and reoriented to orbito-meatal, coronal and sagittal slices of 3.4-mm wide.

On the scan, hypoperfusion of the left posteriofrontal lobe (Fig. 1), especially of the inferior portion was observed, with possible involvement of the adjacent left anterior temporal lobe (Figs. 2,3).

DISCUSSION

Neurological deficits including total anarthria, dysphagia and orofacial dyspraxia are well-documented in brain lesions from cerebrovascular strokes and head trauma (*3*). Speech and swallowing disabilities with orofacial dysfunction are commonly linked to motor neuron disease associated with additional typical signs, such as tongue atrophy and fasciculations.

Severe impairment of voluntary movements of the buccolingual musculature, supranuclear palsy of the seventh, ninth and tenth cranial nerve, and twelfth cranial nerve palsy was described as anterior operculum syndrome. Foix et al. (*4*) and Mariani et al. (*5*) reported two patients having facial weakness, drooling, palatal and tongue movement dysfunction, and failure of speech caused by cerebrovascular events (*3*). Tumor, trauma, congenital anomalies and herpes encephalitis were reported as other causes of anterior operculum syndrome (*6*). Developmental anterior operculum syndrome was described in identical twins having bilateral perisylvian cortical dysplasia (*7*).

Only a few reports described similar symptoms without additional cognitive or motor deficits in the degenerative process of the elderly.

A single case of orofacial dyspraxia with loss of speech output and extrapyramidal symptoms was described with cortico-basal-ganglionic degeneration (*8*). Other studies, which examined the role of apraxia of different types in this disorder,

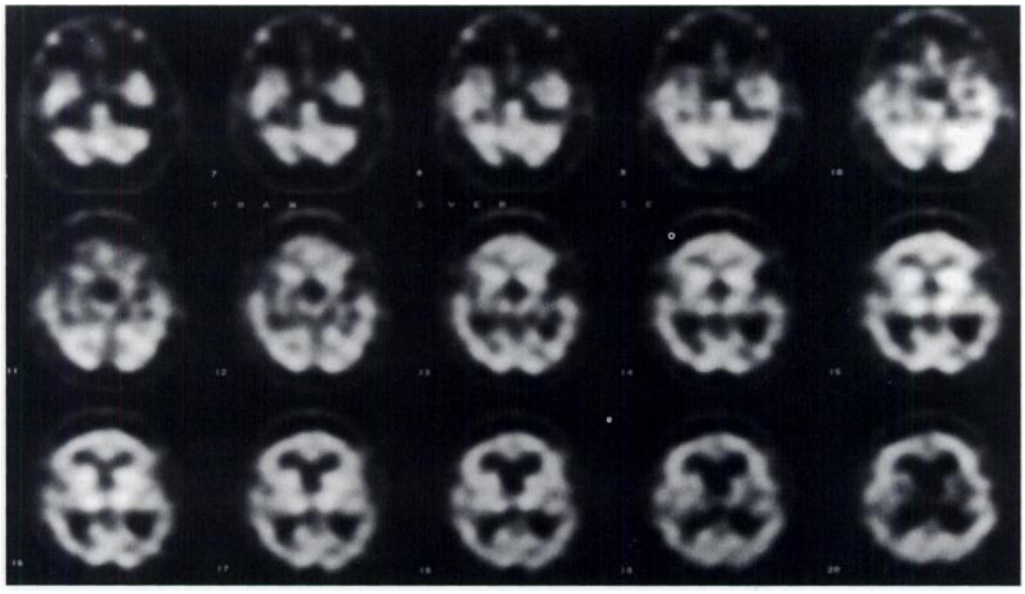

FIGURE 2. Selected transverse slices of the brain. Slice thickness is about 0.34 cm. Note the perfusion defect in the left lower posterior lobe. Involvement of the adjacent temporal lobe was suspected.

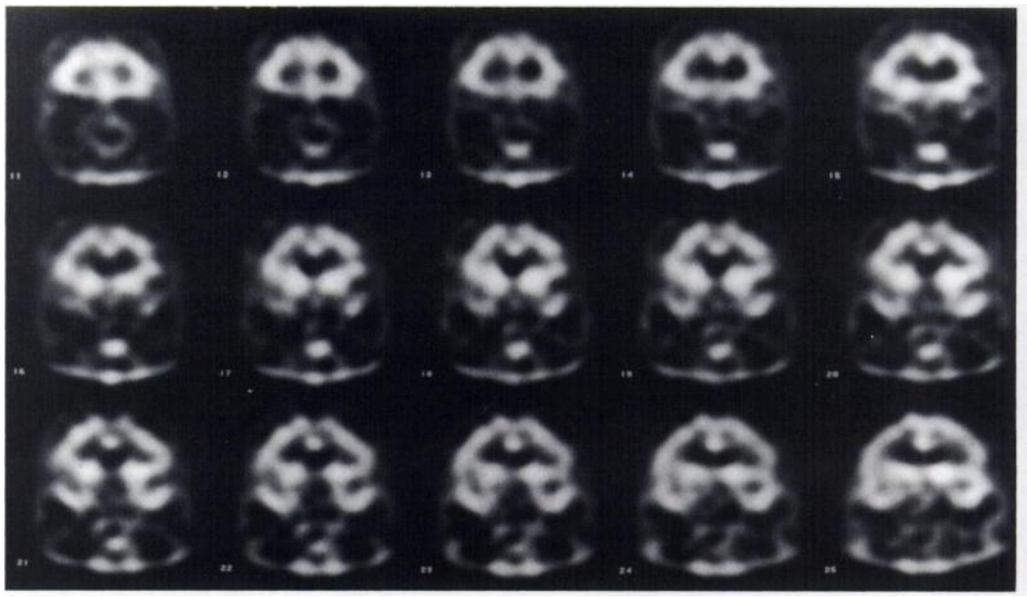

FIGURE 3. Selected 0.34 cm wide coronal slices. Note the same frontal lobe lesion as in Figure 1.

could not find any case of bucco-oral-facial apraxia and concluded that the apraxia of facial movements is not included in cortico-basal-ganglionic degenerative disease (*9*). Buccofacial and ideomotor apraxia with aphasia, word amnesia and stuttering were reported in one of three patients in the early stage of slowly progressive fluent aphasia (*10*). Tyrell et al. (*1*) described three patients who had progressive loss of speech output and orofacial dyspraxia in focal cortical degeneration. The anatomical location of the anterior operculum syndrome was found as bilateral damage of the anterior perisylvian area with involvement of frontal or parietal operculum (*11,12*). Lesions in the frontal lobes bilaterally (*13*), in the unilateral fronto-parietal lobe (*2*) and in the ventrolateral portion of the anterior frontal area involving also the parietal operculum and the anterior periventricular white matter (*1*) were described in those cases.

Whereas vascular, neoplastic or trauma causes of anterior operculum syndrome are easy to diagnose by CT scan or MRI, neurodegenerative changes are difficult to substantiate using these methods. The PET technique was used in these neurodegenerative processes to try to identify the location of the damage. Using PET O^{15} (*1*), frontal hypometabolic defects in the inferior and lateral areas were demonstrated in patients having focal cortical degeneration. In the case of a patient having cortico-basal-ganglionic degenerative syndrome with similar neurologic deficits, no changes were observed using the PET fluorodeoxyglucose technique (*8*). SPECT was not used in any of the previously reported patients.

The usefulness of SPECT in the evaluation of degenerative process of the brain was demonstrated in Alzheimer's disease and in other primary degenerative diseases (*14,15*). The three-dimensional information about surface cortical perfusion with a correlation to higher cortical malfunctions using ^{99m}Tc-HMPAO (*14*) was found to be an effective method for differentiating between the various degenerative processes of the brain cortex (*16*). Also, in slow progressive asymmetrical and focal brain atrophy, SPECT was reported to be an effective method (*17*). The key role of SPECT in the evaluation of degenerate diseases of the brain is accentuated by the fact that in a major number of those patients no abnormality can be demonstrated using other neuroimaging methods.

In our patient, disturbances in swallowing, speech and facial movements coupled with normal neuroimaging by CT and MRI re-inforce the assumption that the patient suffered from the unusual manifestation of focal cortical degeneration with anterior operculum syndrome. However, the pathology was localized by using SPECT. Unilateral hypoperfusion of the left posterior portion of the frontal lobe with extension into the temporal area is compatible with the expected anatomical locations. Since CT and MR images were negative in this condition, PET is frequently not available in most medical centers, and brain biopsy as a diagnostic method is not justified in these patients. SPECT can be a safe, noninvasive and efficacious technique for contributing to the documentation of unusual cases of isolated focal syndromes. SPECT may also further augment our understanding of the pathophysiology of these cases.

ACKNOWLEDGMENT

We thank Ms. Judy Brandt for her help with the English editing and skillful word processing.

REFERENCES

1. Tyrrell PJ, Kartsounis LD, Frackowiak RSJ, Findley LJ, Rossor MN. Progressive loss of speech output and orofacial dyspraxia associated with frontal lobe hypoperfusion. *J Neurol Neurosurg Psychiatry* 1991;54:351–357.
2. Natan P. Facial apraxia and apraxic dysarthria. *Brain* 1947;70:449–478.
3. Ma OC-C, Covll BM, Golper LAC, Rav MT. Anterior operculum syndrome. *Neurology* 1989;39:1169–1172.
4. Foix C, Chavancy JA, Marie J. Diplegie facio-linguo-masticatrice d'origiue cortico-sous-corticale sans paralysie des membres. *Rev Neurolo* 1926;33:214–219.
5. ariani C, Spinnler H, Sterzi R, Vallar G. Bilateral perisylvian softening: bilateral anterior opercular syndrome (Foix-Chavany-Marie syndrome). *J Neurol* 1980;223:269–284.
6. Van der Poel JC, Haenggli CA, Overweg-Plandsoen WCG. Operculum syndrome: unusual feature of herpes simplex encephilitis. *Pediatr Neurol* 1995;12:245–246.
7. Graff-Radford NR, Bosch EP, Stears JC, Tranel D. Developmental Foix-Chavany-Marie syndrome in identical twins. *Ann Neurol* 1986;20:632–635.
8. Lang AE. Cortical basal ganglionic degeneration presenting with a progressive loss of speech output and orofacial dyspraxia. *J Neurol Neurosurg Psychiatry* 1992;55:1101.
9. Leiguarda R, Lees AJ, Merello M, Starkstein S, Marsden CD. The nature of apraxia in corticobasal degeneration. *J Neurol Neurosurg Psychiatry* 1994;57:455–459.
10. Sakajiri K, Bandou M, Yamanouchi H, Ishii K, Fukusako Y. Slowly progressive dysarthria and impaired language function: a case report. *Rinsho Shinkeigaku* 1992;32:1107–1111.
11. Mohr JP, Pessin MS, Finkelstein S, Finkelstein HH, Duncan EW, Davis KR. Bucco aphasia: pathologic and clinical. *Neurology* 1975;28:311–324.
12. Schiff HB, Alexander MP, Naeser MA, Galaburda AM. Aphemia: clinical anatomic correlation. *Arch Neurol* 1983;40:720–727.
13. Grosswasser Z, Korn C, Grosswasser-Reide I, Solzi P. Mutism associated with buccofacial apraxia and bihemispheric lesions. *Brain Lang* 1988;34:157–168.
14. Hashikawa K, Matsumoto M, Moriwaki H, et al. Three-dimentional display of surface cortical perfusion by SPECT: application in assessing Alzheimer's disease. *J Nucl Med* 1995;36:690–696.
15. Talbot PR, Lloyd JJ, Snowden JS, Neary D, Testa HJ. Choice of reference region in the quantification of SPECT in primary degenerative dementia. *Eur J Nucl Med* 1994;21:503–508.
16. Talbot PR, Snowden JS, Lloyd JJ, Neary D, Testa HJ. The contribution of SPECT to the clinical differentiation of degenerative cortical brain disorders. *J Neurol* 1995;242:579–586.
17. Caselli RJ, Jack CR Jr, Petersen RC, Wahner HW, Yanagihara T. Asymmetric cortical degenerative syndromes: clinical and radiologic correlations. *Neurology* 1992;42:1462–1468.