Carotid gustatory syndrome in a patient with Holmes-Adie syndrome

Holmes-Adie syndrome comprises the “tonic” pupil and reduced or absent deep tendon reflexes,1,3 and it can be accompanied by various other symptoms such as segmental hypohidrosis, impaired sudomotor and vasomotor function, orthostatic hypotension and peripheral neuropathy. We report a patient with Holmes-Adie syndrome who noticed an unusual taste sensation in the left posterior part of the tongue. When the neck was pressed on the same side. The patient was a 57 year old Japanese woman who was found to have dilated and non-reactive pupil in the left eye. Shortly afterwards, she noticed that pressing on the left anterior neck elicited a “metallic” taste on the left posterior part of her tongue. Otherwise she was asymptomatic.

General examination was normal. The left pupil was irregularly shaped and 6 mm in diameter, while the right pupil was 4 mm in diameter. The left pupil reacted to light very slowly and incompletely to a minimal diameter of 4 mm while the right pupil reacted promptly. When the neck was pressed on the left, the right pupil dilated promptly, but the left pupil was very sluggish in dilatation. Convergence reflex was moderately slow in the left while it was normal in the right. Accommodation near point being 10 cm bilaterally. The left pupil showed an excessive response to the application of 2.5%, methacholine and became much smaller than the right. Visual acuity, visual field and ocular fundi were normal. There was no extracocular muscle palsy. Facial muscles, facial sensation and corneal reflexes were all normal, as was her hearing. Soft palate and tongue function normal. Lacrimation and salivation were normal.

Pressure on the left anterior neck overlying the carotid sinus immediately produced a “metallic” taste on the left posterior tongue. Histamine (100 µg) was injected under the skin of the neck via the carotid artery but there was no reaction. The skin was moderately dry and moist. There was no sphincter impairment. The supine BP of 122/88 fell to 96/0 on standing although the patient did not complain of any dizziness and the pulse rate did not change. Laboratory tests including complete blood count, blood sugar, ESR, CRP, serum protein fraction, RA test, anti-nuclear antibody, anti-DNA antibody, anti-RNP antibody, LE tests and ECG were all normal or negative. During hyperventilation, the heart rate increased from 66 to 96 beats per minute. Neurological examination was also normal. The conduction velocity of the posterior tibial nerve and the sural nerve was 43 and 44 ms, respectively. The distal motor latency of the right posterior tibial nerve was 5 ms and the evoked EMG being 5 mV. Neither H-reflex nor T-reflex could be elicited in the gastrocnemius muscles.

The diagnosis of Holmes-Adie syndrome was made based on the presence of a tonic pupil associated with overactive response to methacholine and the absence of ankle jerks which was proved by H-reflex study. There were at least two more features in the present case: orthostatic hypotension and a unique syndrome consisting of gustatory sensation elicited by pressure applied to the carotid sinus of the same side. Johnson et al reported two patients with Holmes-Adie syndrome accompanied by orthostatic hypotension. Their cases were found to have afferent block from baroreceptors in contrast to the efferent autonomic block found in most other cases of idiopathic orthostatic hypotension. In our case, it is less likely that the orthostatic hypotension was due to the afferent baroreceptor block because the heart rate responded normally to the carotid massage. The most unusual feature in our case was the “metallic” taste sensation elicited in the left posterior part of the tongue by pressing the anterior neck on the same side. To our knowledge this phenomenon has not been described previously. The gustatory sensation elicited by the carotid pressure can be best explained by postulating a misconnection, within the glossopharyngeal nerve, of the afferent impulses from carotid baroreceptors with the sensory impulse from taste fibres originating from the posterior third of the tongue. This condition may be called “carotid gustatory syndrome”. Pathogenesis of Holmes-Adie syndrome is from case to case, and it may be associated with collagen disease, especially systemic lupus erythematosus. In our case, however, we could not demonstrate any underlying cause. In summary, the present case of orthostatic hypotension and unilateral glossopharyngeal neuropathy, it is most likely to be a part of systemic poly- and multiple neuropathy involving autonomic and somatic peripheral nervous systems.

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The relationship of memory and cognition in Parkinson’s disease to laterisation of motor symptoms

A recent report1 indicated that Parkinson’s disease (PD) patients with greater involvement on the left body side were more impaired than right body patients in memory, visuospatial performance, language and mental control. A question was later raised as to whether this was due to actual lateralised differences or more a function of symptom and disease and disability.1 In view of the apparent contradiction we explored other possible left-right hemisphere differences in psychological functions in the hemi-Parkinsonian patients to determine further the role of symptom severity and disability in such differences.

Fifty three consecutively tested patients with idiopathic PD served as subjects. All were outpatients receiving Sinemet treatment and almost half were receiving Symmetrel as well. The mean age of the group was 68 years (range 41–83). There were 21 females and 32 males. Mean length of formal education was 12.3 years (range 5 to 20 years). Mean length of illness was 12 years, five months (range three months to 57 years). Pattern and degree of symptoms and functions status varied widely. In addition to the other pertinent data, charts were reviewed for marital status, age, education, sex, length of illness and age at onset of illness. Dosage and time on medication for Sinemet and Symmetrel were also recorded. Disability factors, clinical ratings were available for tremor, rigidity, alternating movement, bradykinesia and functional activities. Cognitive functions of the body side that the body showed greater involvement by totalling the ratings for tremor, rigidity and alternating movement impairment for each side. If the total score was one rating point or more different between sides, the side with the higher score was deemed primarily involved. On this basis, 15 left side and 10 right side dominant patients were identified. For the left side dominant group the total ratio varied from 0.0 to 0.5 (mean = 0.25 ± 0.19, range 32 males. Mean length of illness, education, or length of time on and dosage of medication. They did not differ in total tremor, rigidity or alternative movement impairment, but did differ significantly in

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