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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 reflexes, cardiac autonomic dysfunction, 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 after, 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 to 2 mm. In a dark room, 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 was normal with the 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 extraocular muscle palsy. Facial muscles, facial sensation and corneal reflexes were all normal, as was her hearing. Soft palate and tongue were normal. Lacrymation and salivation were normal.

Pressure on the left anterior neck overlying the carotid sinus immediately produced a "metallic" taste on the left posterior tongue. Heart rate on ECG was slightly decreased by the carotid pressure. Pressing the right side of the neck did not produce any gustatory sensation. Superficial sensation of the tongue and pharynx was normal, and taste sensation was normal over the tongue. Cranial nerves were otherwise normal. There were no motor or sensory abnormalities in the limbs except for loss of ankle reflexes bilaterally even with reinforcement. The skin was moderately moist. There was no sphincter impairment. The supine BP of 122/88 fell to 96/0 on standing although the patient did not com-

plain 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. Needle EMG showed no abnormality. 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 5.1 ms with the amplitude of 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*⁴ 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 varies 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 view of the association with 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 lateralisation 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 severity of disease and disability.² 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 the pool of 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 functional status varied widely. To secure 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 tabulated. For symptom and disability factors, clinical ratings were available for tremor, rigidity, alternating movements, bradykinesia and functional activities.

It was then determined which side of 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 ratings ranged from 0 to 8 (L mean = 5.9; R mean = 3.0, $t < 0.01$). For the right side dominant group they ranged from 0 to 7 (L mean = 2.2; R mean = 4.9, $t < 0.01$). The groups are significantly different for the side of the body most affected.

To determine the total symptom severity for each patient, ratings of bradykinesia and impaired function were added to those of tremor and rigidity.

A modification of the Randt Memory Test³ was used to evaluate memory functions. Several aspects of episodic memory were evaluated, including rote, associative, discourse and visual recognition. The modules used were general information, recall of five items, recall of a short story and recall of drawings of everyday objects. Cognitive functions were assessed by administration of the following subtests of the Wechsler Adult Intelligence Scale-Revised:⁴ Information, Digit Span, Vocabulary, Similarities, Picture Completion, Block Design and the Digit Symbol. The battery includes both verbal and non-verbal tests. Prorated Verbal and Performance IQ scores were also derived.

The left and right side dominant groups did not differ for age, age at onset, sex, 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

Table Left-right side differences in memory and cognition before (variance) and after (covariance) adjusting for symptom severity (left side N = 15, right side N = 10)

	Left side Mean	Right side Mean	Variance		Covariance	
			t	p	t	p
Information	8.8	7.1	-1.37	0.17	-0.30	0.77
Digit Span	9.0	6.9	-2.27	0.03	-1.48	0.14
Vocabulary	8.3	7.4	-0.77	0.44	0.30	0.76
Similarities	6.1	5.7	-0.30	0.76	0.76	0.45
Picture Completion	6.4	4.0	-2.10	0.04	-1.07	0.29
Block Design	6.7	4.1	-2.71	0.01	-1.70	0.09
Digit Symbol	5.1	3.6	-2.19	0.03	-1.20	0.24
General Information	12.2	11.7	-1.60	0.11	-0.60	0.55
Five Items	9.9	9.4	-0.37	0.71	0.82	0.41
Paired Associates	13.0	8.8	-2.69	0.01	-1.87	0.07
Short Story	7.6	5.7	-1.37	0.18	-0.54	0.59
Picture Recognition	6.8	6.6	-0.56	0.58	0.05	0.96

bradykinesia ($F = 5.38$, $p = 0.03$) and functional impairment ($F = 4.60$, $p = 0.04$) with the right side dominant group being both more bradykinetic and more functionally impaired. Consequently, concurrent analyses of variance and covariance were performed, the latter to adjust for the observed differences in bradykinesia and functional disability.

Before adjustment for symptom severity the right side dominant group scored significantly lower in five of 12 tests of memory and cognition and showed trends in the same direction for three others. After adjustment, there were no significant differences and only two tests showed trends in that direction. The differences, when noted, included both cognitive and memory functions, verbally and non-verbally mediated and were wide ranging, involving attention and concentration span, recent memory, alertness to stimuli, spatial-perceptual performance and associate learning, and involved tests with and without a motor component.

The finding that disease severity, particularly bradykinesia and impaired function, are associated with memory and cognitive differences rather than laterality of symptoms are contrary to those reported by Direnfeld *et al.*¹ but generally support the results of Zetuský and Janovic.² Severity of the disease, particularly bradykinesia and degree of disability, appears to play a most significant role in the behavioural deficits observed in both left and right hemi-Parkinsonian patients. In these studies, it is important to describe and equate the pattern of symptoms, stage of disease and even personal variables, such as education when comparing left and right side dominant patients on behaviour functions, since groups vary widely in these areas. The current findings also lend support to the notion that subcortical lateralisation is less definitive than for the cortex.³ Further research is needed, preferably with groups more definitively classified as having left or right side dominant symptoms and who are sufficiently advanced in the disease so that memory and cognitive deficits are present and can be discerned to differentiate subjects on these variables.

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Pseudotumour cerebri with focal neurological deficit

Hemiparesis associated with facial nerve palsy developed in a patient with pseudotumour cerebri¹ and resolved after treatment.

A 29 year old right handed obese female had onset of headaches, numbness of the right side of the face, altered taste sensation, and tingling and weakness of the right arm and right leg ten days before admission on 20 January 1988. The headache was described as a severe pressure-like sensation with predominance in the occipital region associated with some nausea. She experienced blurring of vision four days before admission which gradually became worse. Five days before the admission the weakness and numbness also increased.

Past medical history included a left Bell's palsy in May 1986 which resolved completely in one and a half months. At that time she also had blurring of vision and severe bifrontal headache associated with nausea and vomiting. The diagnosis of pseudotumour cerebri was made based on the CT scan which showed small ventricles and the lumbar puncture which revealed an opening pressure of 300 mm of water in fully extended position. She was obese (199 lbs), and had papilloedema and increased central scotoma. The patient was treated with tapering doses of prednisone and Diamox. She had complete resolution of headache and visual problems after reducing 40lbs over a period of four months. The patient has remained free of symptoms. There was no history of taking oral contraceptives, tetracyclines, vitamin A nor of any head injury.

Abnormal findings on admission included: obesity (250 lbs), papilloedema, reduced right corneal reflex, decreased sensations in

the right half of the face, right complete lower motor neuron type of facial palsy, right hemiparesis with 4/5 power, and decreased sensations in the right half of the body, right sided hyperreflexia and equivocal plantar response. Routine laboratory blood tests were all within normal limits. A CT scan of the brain with brain stem cuts, with and without contrast, and MRI were all within normal limits. A lumbar puncture revealed 450 mm of water of opening pressure with normal laboratory examination including oligoclonal bands, myelin basic protein, immunoglobulin G and CSF cytology. Within half an hour of the first lumbar puncture her headache responded dramatically and her facial weakness improved remarkably. She also had subjective improvement of the right sided numbness. However, the arm drift persisted for another three hours. She was started on prednisone 80 mg and Diamox 250 mg three times daily. In 24 hours her facial weakness was very mild, the numbness of the right side had disappeared and the arm drift was no longer observed. Two days later a repeat lumbar puncture was done due to the recurrence of headache. There was an opening pressure of 430 mm with no specific laboratory abnormalities. After the second LP the severe headache was relieved but the visual symptoms persisted to a less severe degree. A third lumbar puncture carried out three days later revealed an opening pressure of 220 mm. She was discharged home and continued on a tapering dose of prednisone and Diamox 250 mg three times daily. Within two weeks the visual symptoms completely resolved. On examination after one month, there was no evidence of neurological deficit. She is no longer on prednisone or Diamox and has reduced her weight by another 7 lbs over a period of three weeks. She continues on a programme of exercises and weight reduction.

The patient experienced a right hemiparesis with right facial palsy associated with benign intracranial hypertension. There was no evidence of mass lesions, obstructive hydrocephalus, infections, demyelinating disease or hypertensive encephalopathy. Unlike other patients previously reported, with presenting symptoms of headache, visual impairment and diplopia, this patient had right hemiparesis associated with the symptoms described. Although it has been presented that any focal neurological deficit excludes the diagnosis of pseudotumour cerebri, there have been reports of focal symptoms and signs involving cranial nerves.

Saahs and Joynt² first provided histological evidence of oedema in the brain with biopsy specimens taken at the time of subtemporal decompression for benign intracranial hypertension. The use of MR imaging showing focal areas of increased signal intensity in the periventricular white matter that could not be detected on the CT scan further supports this theory.³ In a review of the literature of 120 patients with benign intracranial hypertension, 10% had equivocal neurological signs.⁴ The association of pseudotumour cerebri and facial pain with hyperaesthesia and diminished corneal reflex with a dramatic response of symptoms and signs by lowering the intracranial pressure has been reported.⁵ Minor symptoms such as tinnitus, paresthesias, difficulty in walking and transient visual obscurations have been reported to be related to PTC.⁶

This case represents a woman with two episodes of symptoms and signs consistent