Carotid gustatory syndrome in a patient with Holmes-Adie syndrome

Holmes-Adie syndrome comprises the “tonic” pupil and reduced or absent deep tendon reflexes, and it can be accompanied by various other symptoms such as segmental hypothaloid, 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, and the left pupil was fixed in the same direction. 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. The left 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 reflex 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 were normal. Lacrymation and saliva were normal.

Pressure on the left anterior neck overlying the carotid sinus immediately produced a "metallic" taste on the left posterior tongue, H-reflex and EOG 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 dry and moist. There was no phinphent sensation. 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, ESP, CRP, serum protein fraction, RA test, anti-nuclear of body, anti-DNA, anti-RO/NP antibody, LE tests and ECG were all normal or negative. During hyperventilation, the heart rate increased from 66 to 96 beats per minute. Nystagmus was noted in the abductor side.

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 with the H-reflex being 5 ms. Neither H-reflex nor N-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.5 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 response usually involved the carotid sinus.

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 fibers originating from the posterior third of the tongue. This condition may be called "carotid gustatory syndrome". Pathogenesis of Holmes-Adie syndrome differs 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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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 symmetry 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.8 but generally support the results of Zetisky and Janovic.9 Severity of the disease, particularly bradykinesia and degree of disability, appears to play a more 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.10 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 discriminated to differentiate subjects on these variables.

### Pseudotumour cerebri with focal neurologica1 deficit

Hemiparesis associated with facial nerve palsy developed in a patient with pseudotumour cerebri 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 predominant occipital dominance in the occipital region associated with some nausea. She experienced blurring of vision for 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 mmHg of water in fully extended position. She was obese (199 lbs), and had papilloedema and increased intracranial pressure. She 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 laboratory 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 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 (MG) cys-tyr-lys, and no specific histological 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 mmHg. 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 visual 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. Saks and Joynt 11 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 pericruciate area was also used. The diagnosis of pseudotumour cerebri was made based on the CT scan further supports this theory.1 In a review of the literature of 120 patients with benign intracranial hypertension, 10%, had equivocal neurological signs.12 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.13 Other symptoms such as tinnitus, paresthesias, difficulties in walking and transient visual obscurations have been reported to be related to PTC.14 This case represents a woman with two episodes of symptoms and signs consistent with