Letters

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Taste loss as an initial symptom of primary amyloidosis

Sir: One of the major clinical features of amyloidosis is polyneuropathy, especially of the sensory dominant type, observed in 90% of those with familial and 10–20% of those with non-familial amyloidosis.1 2 Taste is a chemical sense conducted via the sensory nerves, but taste disorders have not been described in the reported sensory disturbances of amyloidosis.1–3 We encountered a patient with primary amyloidosis who presented with taste loss as the initial symptom.

A 55 year old female had a history of hyperthyroidism at the age of 45 years. She noted the loss of taste at the age of 52, accompanied by loss of appetite and general fatigue. She was treated for depression, but her symptoms did not improve. She exhibited a slightly low plasma zinc level of 54 μg/dl (normal 65–110 μg/dl) and was treated with zinc sulfate (300 mg/day) for 6 weeks without appreciable effects. In June 1983, hyposmia developed; in July, transient episodes of dizziness on standing were noted. She began to lose weight gradually in 1984 from 59 to 42 kg. In June the same year, she suffered from recurrence of orthostatic hypotension, which deteriorated and was occasionally accompanied by syncope. She subsequently developed constipation in August, liver dysfunction and slight hepatomegaly in October, urinary disturbance in March 1985, and anosmia and sensory disturbance of the limbs in June.

She was admitted to Kagawa Central Hospital in June 1985. Taste was completely lost in all areas of the chorda tympani, glossoaryngeal, and vagus nerves. Electrogustometry was negative at the maximal stimulation of 32 dB (normally positive at more than 8 dB). Qualitative and quantitative clinical gustometry (using filter-paper discs4) were all negative for 80% sucrose (sweet), 20% saline (salty), 80% tartaric acid (sour), and 4% quinine HCl (bitter). These concentrations are several to more than ten times the normal threshold levels. Touch sensation of the tongue and the oral cavity was normal, and no local tongue lesions such as macroglossia were noted. Other neurological findings included left anosmia, left mydriasis, loss of left light reflex, decreased salivation and lacrimation, areflexia of distal extremities, and peripheral neuropathy of the socks-gloves type.

The motor nerve conducting velocity showed a slight decrease, being 38.9 m/s for the right median nerve and 38.6 m/s for the left posterior tibial nerve. Sensory nerve conduction could not be evoked. Other clinical findings were goitre, hepatomegaly, nephrotic syndrome, constipation, neurogenic bladder, and orphothastic hypotension. Ultrasonic echocardiography showed thickness and granular sparkling texture of the interventricular septum and posterior wall, suggesting cardiac amyloidosis. Laboratory findings in addition to the abnormal values corresponding to the clinical findings above were positive serum electrophoresis for M-protein in the gamma-globulin fraction, and immunoelectrophoretic analysis demonstrating M-proteinaemia IgG-L type. Urine concentrated 25-fold also yielded M-protein and Bence Jones protein of lambda type. Plasmacytes of the bone marrow showed a slight increase to 4–4% without abnormal nuclei. Biopsy of the minor salivary gland and thyroid gland revealed deposition of amorphous substances which were stained with congo-red and showed green birefringence under polarising microscopy. These substances were identified by electron microscopy as scattered amyloid fibrils and a diagnosis of primary amyloidosis IgG-L type was made.

Upon diagnosis, treatment was initiated with merpharan (2 mg/day), prednisolone (10 mg/alternative day), and dimethylsulfoxide (10 ml/day). Although this treatment produced moderate improvements in the sensory disturbance of the upper extremities, the loss of taste persisted and other symptoms either remained unchanged or deteriorated. Moreover, oedema, anaemia, petechiae, and nausea developed thereafter, and slight enlargement of the tongue was noted in November 1985.

The patient diagnosed as having primary amyloidosis IgG-L type was noted particularly for showing loss of taste as the initial presenting symptom. Taste disturbances may be caused by head trauma, tumour, lesions of the conduction route for taste, systemic disorders such as diabetes mellitus, cold, psychological disorders, and side effects of drugs.5 The loss of taste in our patient could be readily differentiated from gustatory disorders arising from these causes. Zinc deficiency, another frequent cause of taste disturbance,6 was also excluded since the administration of zinc sulfate was ineffective. Acute panadynia-autonomia also causes taste loss, but the other symptoms are confined to the autonomic nervous system and recovery may occur.7 Taste disturbances have not been described in reports of amyloidosis,5 8 and thus their severity or incidence has still to be clarified. However, the tongue is one of the frequent sites of amyloid deposition and may show various pathological conditions such as induration, yellowish nodules, haemorrhagic bullae, and aphthous ulcer.8

Amyloidosis, therefore, may well be expected to complicate taste disorders. Our patient presented with a tendency to macroglossia in November 1985, suggesting amyloid deposition also involving the tongue. The loss of taste may be ascribed to this condition, and the appearance of taste loss preceding that of macroglossia by 3 years in our patient indicates that a small amount of amyloid is sufficient to induce loss of taste. Therefore, taste loss may be an initial symptom of amyloidosis. Our experience emphasises that quantitative gustometry is necessary for investigating amyloidosis, and it may allow an earlier diagnosis of amyloidosis.

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References

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Sinoido-atrial arrest due to temporal lobe epilepsy

Sir: Autonomic events may accompany many types of epileptic seizure. They can occur as a normal response to the stress of an attack, but may also result from seizure activity spreading to autonomic centres. Consequent alterations in heart rate or rhythm are among the possible causes of death during a seizure. Bradycardiac arrhythmias have been noted less often than tachyarrhythmias and, to our knowledge, there have been only three documented cases of sino-atrial arrest, each in association with temporal lobe epilepsy. We here report a further case, in which the temporal lobe epilepsy was secondary to an intracranial tumour.

A 35 year old man was admitted to the Coronary Care Unit, Rochdale, in May 1982, having blacked out at the wheel of his car. He recovered quickly and there was no head injury, but he had two further brief losses of consciousness on the way to the Unit. During these he had a vacant stare with some jerky movements of the limbs, and the ambulance man could not detect his pulse. Clinical examination on admission was normal, but he soon had a further attack witnessed by nursing and medical staff. He reported a sudden feeling of déjà vu, at which time his pulse rate was unchanged at 50-60 beats per minute. Over the next 15 seconds there was progressive bradycardia, leading to asystole for a further 20 seconds, accompanied by loss of consciousness and slight twitching of the limbs. He was given intravenous atropine, his heart rate quickly returned to normal, and he developed a reactive hyperaemia as consciousness returned. He had five similar attacks over the next hour until a cardiac pacemaker was inserted. During each he had sufficient warning to remove his false teeth and notify staff.

There was no history of previous syncope but a month earlier he had been knocked out during a game of football and, a few days later, had an attack of déjà vu unaccompanied by any disturbance of consciousness. Several similar attacks occurred on the day before admission.

Clinical examination and haematological and biochemical investigations were essentially normal. His ECG, between attacks, was also normal. However, his EEG recorded 6 days after admission showed an excess of irregular slow activity over the left temporal region. Over the next 48 hours he had no further attacks and the pacemaker was removed. Subsequent monitoring showed occasional second degree heart block (Wenkebach phenomenon) at night only.

After 2 weeks' observation, he was transferred to the Southern General Hospital and was later discharged taking phenytoin. However, he had further fits and had to be re-admitted 15 months later, complaining of left-sided headaches. Examination revealed right optic atrophy and a CT scan and magnetic resonance imaging showed a suprassellar cystic lesion, though to be a craniopharyngioma (fig).

At operation in September 1983, the lesion proved to be an epidermoid tumour adherent to the optic chiasm and under surface of the hypothalamus and frontal lobes. The caviy was evacuated and postoperative recovery was uneventful. He has remained well and free of fits or syncope since then, with resolution of the previous visual field defect.

A recent survey of statistics concerning sudden death in epilepsy showed that in 3 to 31% of recorded cases, death was sudden, unexpected and without anatomically demonstrable cause. It was suggested that cardiac arrhythmias during a fit provide the most likely explanation for such deaths, particularly in patients with subtherapeutic anticonvulsant levels or co-existent cardiac disease. Furthermore, prolongation of the electrocardiographic QT interval, with its associated risk of ventricular arrhythmias, may have a higher incidence among epileptic patients. Transitions between sleep stages and the emotional and physical stress of an attack, could act as arrhythmogenic factors through the intermediary of the autonomic system. In addition, ready pathways for seizure activity to directly disturb autonomic function exist in the various conditions.