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Hemiageusia: an unusual presentation of multiple sclerosis

Patients with multiple sclerosis (MS) rarely complain of taste disturbances, although electrogustatory examinations often demonstrate dysfunction of the taste pathway in patients with advanced disease, especially in those with prominent brainstem involvement.¹

A 25 year old native American man presented with a two day history of gradually progressive loss of taste on the entire right half of his tongue (hemiageusia). One week later he developed numbness of the right inner cheek, double vision, and a tendency to fall to the left. He had clockwise rotatory nystagmus, right internuclear ophthalmoplegia, left central facial palsy, left hyperreflexia, and intention tremor with the right hand. The right hemiageusia was unchanged. The gag reflex was diminished on the right side, and the right palate and the right inner cheek were numb. Routine laboratory tests were all normal. CSF analysis revealed no red cells, five lymphocytes, 53 mg/dL protein, normal glucose, 5.6 ng/mL myelin basic protein (normal range 0-5.1 ng/mL), and three oligoclonal bands without correlates in serum.

Eleven days after the onset of the taste disturbance he developed paroxysms of pain around the right eye, perioral numbness and tingling sensation in the right cheek. New

findings were a decreased right corneal reflex and hypaesthesia to pain and temperature in the distribution of the right mandibular nerve. Cranial MRI demonstrated multiple, bilateral, periventricular areas of increased T2 signal. A similar lesion was found in the right medulla on the floor of the fourth ventricle (fig). In the CSF myelin basic protein was 16.2 ng/mL and five oligoclonal bands without serum correlate were identified. The patient was treated with a 10 day course of 1 gm/day intravenous methylprednisone followed by a two week oral prednisone taper, without improvement.

Thirteen months after presentation he developed acute left hemiparesis. MRI demonstrated an increase in the number of periventricular lesions of increased T2 signal and a new right sided lesion at the expected location of the internal capsule. Visual evoked potentials and lower extremity somatosensory evoked potentials showed prolonged wave latencies. CSF showed an elevated protein (63 mg/dL) and myelin basic protein (14.8 mg/mL), and five oligoclonal bands. The patient was treated with a 10 day course of IV methylprednisone and his hemiparesis recovered. On examination he still had the previously described deficits.

Hemiageusia involving the entire right half of the tongue is usually explained by a lesion in the ipsilateral nucleus solitarius, where fibres from the lingual nerve (anterior two thirds of the tongue) and fibres from the glossopharyngeal nerve (posterior third) come together.² Recent evidence suggests the presence of an accessory taste pathway through the trigeminal nerve.³ Hypogeusia is found in 5-10% of patients with advanced MS¹ and is frequently associated with sensory involvement of the trigeminal nerve.³⁻⁵ Taste disturbance as the initial symptom of MS has previously been reported only by Harris.⁶ His patient, a 21 year old woman, developed numbness of the right side of her face and right sided hemiageusia, that persisted for one year. Four years later she presented with trigeminal neuralgia, but it was not until 10 years later, that multifocal symptoms led to the diagnosis of MS.⁶ The remarkable aspect of our case is that right-sided hemiageusia was the sole presenting symptom, although the investigation demonstrated multifocal central nervous system lesions. Right trigeminal sensory involvement occurred almost two weeks after the hemiageusia, and prominent, more widespread brainstem symptomatology developed only later.

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Paroxysmal kinesigenic choreoathetosis as presenting symptom of multiple sclerosis

Paroxysmal kinesigenic choreoathetosis (PKC) is characterised by attacks of uni- or bilateral choreoathetosis precipitated by sudden or fast movements. The acquired form of symptomatic PKC may be a manifestation of underlying structural or metabolic disease.¹ PKC has previously been reported in eight patients as the first symptom of multiple sclerosis.² We describe a patient with PKC as the presenting symptom of multiple sclerosis, in whom the lesions were localised by MRI.

A 35 year old female lawyer noticed diplopia and had minor attacks of sudden halts with her right foot when she started walking as if she had "stepped into glue". She noticed that the attacks were also precipitated by a sudden noise or the unexpected appearance of a cyclist or a car. She first sought medical attention five months later when she developed urgency of micturition and a diminution of dexterity of her right hand. The attacks now included slurred speech and rotational posturing of the head, right arm and leg lasting a few seconds and occurring five to 10 times daily. The attacks were provoked by emotion, acceleration of movement, speaking and writing. Neurological examination was normal. Two months later she was occasionally able to avert the attacks by completely arresting her movements as soon as prodromal symptoms occurred. The attacks occurred now five to 10 times per hour and lasted from about five to fifty seconds.

Neurological examination revealed internuclear ophthalmoplegia, cerebellar gait, dystonia of the right leg and hypertonic hemiparesis of the right side. Cerebrospinal fluid contained 26 mononuclear leukocytes, a slightly increased protein content with an increased IgG level and non-specific multiple oligoclonal bands in the alkaline region at isoelectric focussing. CT scan of the head showed a paraventricular hypodense area in the region of the caudate nucleus. MRI revealed in the proton density images and the T₂ weighted pictures high signal emitting lesions paraventricular, in the putamen and

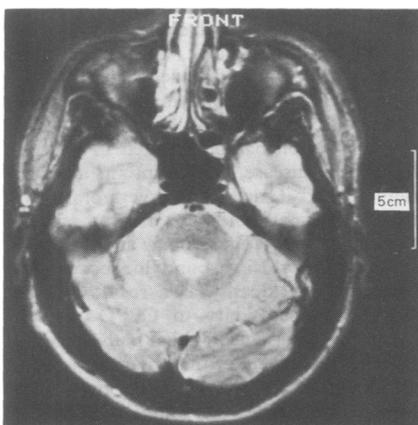


Figure 1 T2-weighted axial MRI scan (1.5 tesla). Increased signal noted by the floor of the fourth ventricle.

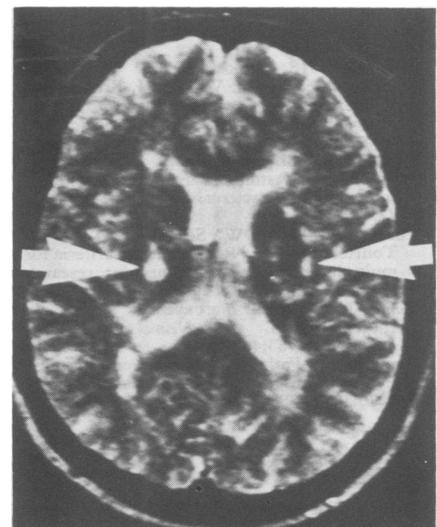


Figure 2 Transverse section through the lateral ventricle with T₂ weighted images of the MRI demonstrates high signal emissions in the right globus pallidus and thalamus and in the subcortical paraventricular regions in both hemispheres.

globus pallidus in both hemispheres (fig). High signal emitting lesions were also present in the left insular cortex and the thalamus, the crus posterior of the left internal capsule, the mesencephalon, the lateral parts of the cerebral peduncles, and the periaqueductal grey matter. These lesions suggest a demyelinating process.

A diagnosis of PKC as result of laboratory-supported definite multiple sclerosis was made.³ She was successfully treated with phenytoin sodium 300 mg daily.

Symptomatic PKC has been associated with structural or functional impairment of the central nervous system.¹ PKC has been described in multiple sclerosis,² neonatal asphyxia and head injury. Reversible PKC has been reported in hypoparathyroidism, insulin-dependent diabetes mellitus, thyrotoxicosis and hypernatraemia, but in most cases no underlying or associated disease has been detected. Phenytoin sodium is the drug most commonly reported to relieve the symptoms.⁴ It is tempting to correlate the findings of the MRI with the clinical symptoms. Neuropathological studies in symptomatic dystonia have revealed lesions in the putamen, globus pallidus, subthalamic nucleus or thalamus.⁵ This is in good correlation with our MRI findings.

As far as we know this is the first patient in whom lesions probably responsible for the PKC have been documented by means of MRI. Further studies with MRI may help clarify the anatomical lesions of this rare disorder.

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Cervical myelopathy due to calcium pyrophosphate dihydrate deposition disease

Calcium pyrophosphate dihydrate deposition disease (CPPD) is characterised clinically by arthritis (pseudogout), radiographically by chondrocalcinosis (heavy punctate and linear

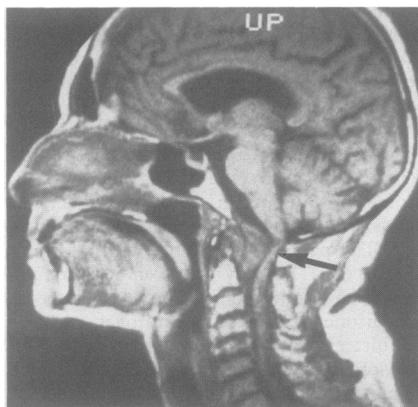


Figure 1 T1-weighted MRI, mid-sagittal plane, demonstrating extra-axial soft tissue mass at the cervicomedullary junction causing cord compression.

radiodensities seen in both hyaline and fibrocartilage), and pathologically by the identification of CPPD crystals in the synovial fluid.¹ In addition to synovial membrane and joint capsule involvement, the disease may also occur in tendons and ligaments.

Cervical myelopathy is a rare complication of CPPD, and has only recently been documented to occur in the anterior cervical spinal canal.

An 85 year old woman, who had for many years had mild arthritis of both wrists, presented with a two year history of intermittent paraesthesiae of her right hand, exacerbated by prolonged sitting. One month before admission she developed neck and right shoulder pain. Two weeks later, she noted progressive upper extremity, followed by lower extremity, paraesthesiae. A week later, when she could no longer walk, she was admitted to hospital. Her weakness and paraesthesiae worsened, and three days before her transfer to New York Hospital, she developed urinary incontinence. Cervical spine MRI without gadolinium-DTPA (figure 1) revealed an extra-axial soft tissue mass at the cervico-medullary junction, causing cord compression and odontoid erosion. CT scan confirmed the presence of calcifications within the mass.

Her examination on admission to hospital showed minimal neck tenderness, spastic quadriparesis with diffuse atrophy, profound loss of proprioception and vibration, and a C2 level to pinprick. At surgery, there was an extradural mass at the base of the skull, associated with thickened fibrous scar tissue and dura, and with odontoid destruction. An extensive surgical resection of the mass was performed.

Histological examination of the surgical specimen revealed dense fibrous connective tissue and granulation tissue with fibrocartilaginous metaplasia. Within the metaplastic regions were haematoxyphilic deposits of CPPD crystals which were positively birefringent in polarised light (fig 2).

Subsequent radiographs revealed typical chondrocalcinosis of both wrists and knees. No associated metabolic disease was identified; serum calcium, phosphorus, blood sugar, electrolytes, creatinine, magnesium, iron studies, serum protein electrophoresis, liver and thyroid function tests were all within normal limits. Her ultimate neurological recovery included improved

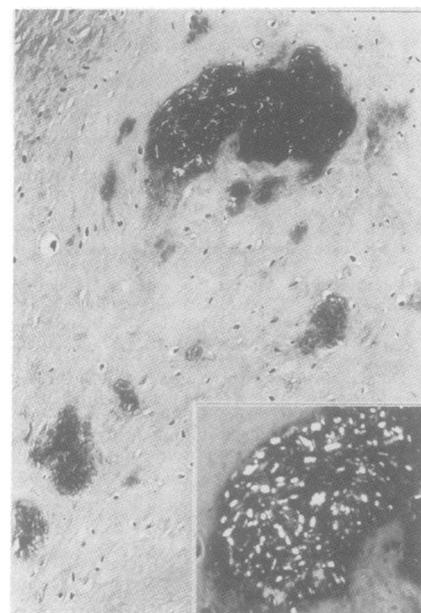


Figure 2 A high power photomicrograph showing several deposits of calcium pyrophosphate crystals (Hematoxylin and eosin stain, $\times 160$). Insert: Birefringence seen in polarised light ($\times 320$).

strength in all limbs, but proprioception remained unchanged, and she was unable to walk. She died approximately six months later, of aspiration pneumonia. No necropsy examination was obtained.

CPPD crystal deposition disease occurs with a population frequency of 1:1000. The prevalence increases with age, and approaches 45% for patients 85 years of age and older.¹ Sixty six per cent of patients have chronic arthritic complaints; two thirds of this group will have superimposed intermittent acute episodes. Fifteen per cent of patients are asymptomatic.² The disease can be sporadic, or associated with metabolic disease, trauma, or surgical procedures. Associated metabolic conditions include haemochromatosis, hyperparathyroidism, hypophosphatasia, hypomagnesaemia, hypothyroidism, neuropathic joints, and amyloidosis.¹

Various radiographic and clinical findings occur in the spine of patients with CPPD disease. Resnick² found radiographic abnormalities of the cervical spine in 52 of 57 patients, including disc space loss with adjacent vertebral body sclerosis and osteophytosis, apophyseal joint abnormalities, and subluxation. Calcification of the syndesmodontoid region has been noted in patients with CPPD disease, but symptoms are generally absent.³ Calcified intervertebral discs may be present, and are usually asymptomatic; rarely, spinal pain and stiffness mimicking ankylosing spondylitis occur.

Cervical myelopathy as a result of cord compression secondary to CPPD is rare. Only two patients with pathologically documented lesions at the anterior cervicomedullary junction have been previously reported;^{4,5} other patients had lesions at or below the third cervical vertebra, in the posteriorly located ligamentum flavum. All cases were clinically similar, and none were diagnosed before surgery.