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Hemiageusia from an ipsilateral multiple sclerosis plaque at the midpontine tegmentum

The exact location of the pontine gustatory pathway has not yet been clarified, probably because there are so few studies of taste in patients with well localised brainstem lesions.[1–3] Here we report on a patient with isolated hemiageusia and trigeminal sensory neuropathy from a single small pontine lesion.

A 46 year old woman experienced a burning sensation on the left side of the tongue. The next day she discovered a loss of taste on the entire left half of her tongue and numbness on the left side of her face. Neurological examination was normal except for hypaesthesia to pain, touch, and temperature sensation in all three divisions of the left trigeminal nerve, with decreased left corneal reflex and no weakness of masseters. Taste sensation was tested using separate solutions of 1.2 M NaCl, 0.47 M glucose, 0.17 M citric acid, and 2.52 mM quinine HCl. Marked disturbance on the left side involving the anterior two thirds and posterior one third of the tongue was noted with all four substances. Routine laboratory tests were normal. Analysis of CSF showed 8 lymphocytes/mm^3, 46 mg/dl protein, increased quantitative intrathecal IgG, and oligoclonal bands. Visual evoked potentials, brainstem auditory evoked potentials, and somatosensory evoked potentials after both median and posterior tibial nerve stimulation were normal. The blink reflex was normal on stimulation on the right, and when stimulated on the left, there was no R1 component and R2 responses were elicited normally. Masseter reflex was absent on the left side. Brain MRI demonstrated multiple bilateral hyperintense white matter signals in periventricular distribution on T2 weighted images, and a hyperintense small lesion in the lateral part of the left midpontine tegmentum that showed enhancement after gadolinium injection (figure A and B). After intravenous high dose methylprednisolone therapy there was no immediate improvement of the patient´s neurological symptoms. In the follow up MRI after 3 months, the gadolinium enhanced pontine image had disappeared, the blink and masseter reflexes were normal, and trigeminal neuropathy and the taste disturbance had gradually reduced.

The present case supports the finding that unilateral pontine lesions result in ipsilateral gustatory deficits, suggesting that gustatory fibres ascend from the solitary nucleus in the medulla up to the homolateral pontine tegmentum without decussating.[1–3] Lower midbrain level decussation is supported by a recent report.[3] Topography of gustatory pontine fibres has been discussed in cases with taste disturbance,[1 3] and, as found here, the absence of limb sensory involvement with normal somatosensory evoked responses contradicts the widely accepted notion that the pontine medial lemniscus conveys taste. On the other hand, Uesaka *et al*[2] described a patient who presented with ageusia and ipsilateral truncal ataxia presumably due to brachium conjunctivum involvement, and, therefore, it was suggested that the adjacent parabrachial nucleus might constitute a pontine taste area. Our patient developed left sided hemiageusia and trigeminal sensory disturbance, and electrical stimulation on the left elicited no R1 response and absence of masseter reflex. These electrophysiological abnormalities imply ipsilateral brainstem lesions at the trigeminal principal sensory and motor nuclei, respectively.[4] Moreover, MRI confirmed the existence of a new gadolinium-enhanced demyelinating lesion in the left midpontine tegmentum. The precise correlation between our patient´s symptoms and the electrophysiological and MRI abnormalities indicates that the involvement of the central tegmental tract, which is the anatomical structure adjacent to the sensory and motor trigeminal nuclei at the midpontine level (figure C), is probably important in causing taste disturbance. According to this hypothesis, Norgren[5] showed in primates that axons of neurons located in the solitary nucleus ascend in the central tegmental tract to the ventroposteromedial nucleus of the thalamus without terminating first in the pontine parabrachial nucleus.

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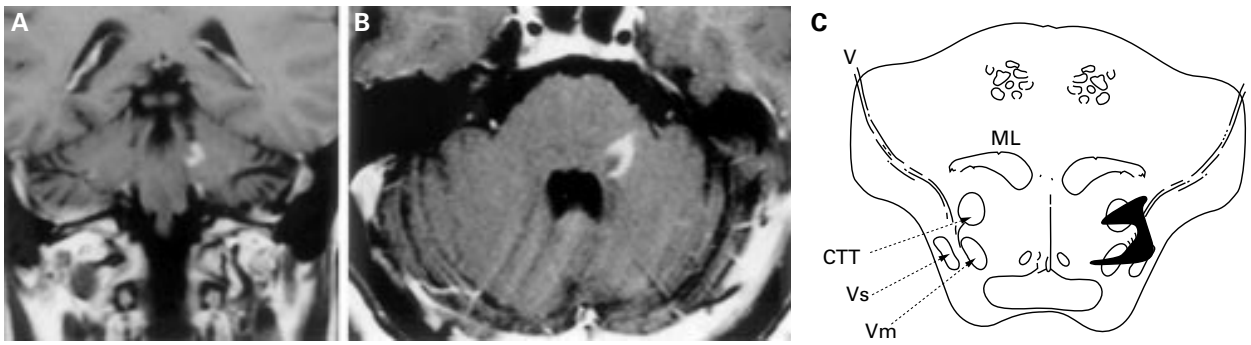
Use of lamotrigine to treat paroxysmal kinesigenic choreoathetosis

We report the use of lamotrigine to treat paroxysmal kinesigenic choreoathetosis. Paroxysmal cases of kinesigenic choreoathetosis were first described in 1962 by Lishman *et al*[1] and the term "paroxysmal kinesigenic choreoathetosis" was first coined by Kertesz[2] in 1967. Recently, a more universal and potentially useful classification of these disorders has been proposed amending the terminology to "paroxysmal kinesigenic dyskinesia".[3] However, we have used the more familiar terminology for our patient as it is more precise. The clinical features of paroxysmal kinesigenic choreoathetosis have been reviewed by Marsden and Luders.[4]

There is a male preponderance of the condition with about 50% of cases inherited as an autosomal dominant trait, the rest being sporadic. Attacks are precipitated by sudden movements or startle and may be unilateral, bilateral, or affect alternate sides. Often the cause is idiopathic but a few cases have been attributed to multiple sclerosis. There has been no clear evidence for seizure activity in this disorder even though the condition is very responsive to antiepileptic drugs such as phenytoin and carbamazepine.[4]

A 13 year old boy of unrelated parents and with no family history of neurological disorder presented with a 6 month history of muscle spasms affecting the left side of his body. Close questioning indicated that his symptoms were brought on during the initiation of sudden movements (such as starting



Gadolinium enhanced T1 weighted MRI of coronal (A) and axial (B) sections showing a high intensity lesion in the lateral portion of the left midpontine tegmentum. Diagram (C) showing the area clinically affected (shaded area). ML=medial lemniscus; CTT=central tegmental tract; Vs=trigeminal principal sensory nucleus; Vm=trigeminal motor nucleus.

to run) and manifested as spasm of the left side of the face and flexor spasm of the left arm and left leg. There were no premonitory symptoms and consciousness was normal during the attack. Attacks lasted for a few seconds only and he felt perfectly well afterwards. Neurological examination was normal. Brain MRI was normal and three EEGs performed over the course of the next year were all normal. There were no biochemical or haematological abnormalities; copper studies were normal. A diagnosis of paroxysmal kinesigenic choreoathetosis was made. He was allergic to carbamazepine and did not respond to sodium valproate.

Lamotrigine(3,5-diamino-6-[2,3-dichlorophenyl]-1,2,4-triazine) is a use dependent blocker of voltage gated sodium channels and has a similar anticonvulsant profile to phenytoin and carbamazepine in animal models.[5] Lamotrigine at a dose of 50 mg twice daily, completely abolished the involuntary movements. On two occasions, these returned on stopping lamotrigine and were abolished by reinstituting treatment. Our patient has taken this for over 2 years without ill effect. He now only experiences symptoms of paroxysmal kinesigenic choreoathetosis if he omits his medication. The response to lamotrigine may provide insight into the pathogenesis of the disease, suggesting that it may be caused by an ion channel defect; these are known to be responsible for some paroxysmal neurological conditions.[6] As far as we are aware, this is the first report of the successful use of lamotrigine to treat paroxysmal kinesigenic choreoathetosis.

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Chronic inflammatory demyelinating polyneuropathy as a complication of cat scratch disease

Cat scratch disease (CSD) was first described in 1950 as a benign regional lymphadenitis. This infection is caused by *Bartonella henselae*. The clinical range of CSD has expanded beyond the classic presentation. In 5%–20% of the infected patients the disease may spread to other organs. However, neurological complications associated with CSD are rare, with encephalopathy being by far the most common form (90%) of nervous system involvement. Encephalopathy occurs in 2%–3% of patients and is more common in adults than in children with the onset varying from a few days to months after diagnosis of CSD.[1] Other known neurological manifestations, often in combination with encephalopathy, are neuroretinitis, oculoglandular disease of Parinaud, myelopathy, radiculopathy or abducens nerve, and facial nerve paresis. We report on a 3 year old boy who developed chronic inflammatory demyelinating polyneuropathy (CIDP) 6 weeks after identification of CSD.

A previously healthy 3 year old boy presented in a paediatric clinic with regional lymphadenitis of the submandibular and suboccipital glands. He was afebrile and did not have any other symptoms. There was no hepatosplenomegaly. No neurological signs were present on physical examination. As the boy might have been scratched by one of his pet cats, in accordance with a small cut on his scalp, CSD was suspected. It was serologically confirmed (enzyme linked immunosorbent assay (ELISA) *B henselae* IgG> 850 U/l; IgM> 250U/l) and he was treated with clarithromycin.

Six weeks after the onset of CSD, he showed difficulty in walking, inability to run or climb stairs, and frequent falls. Slowly progressive pain and a gait disorder were noted over a course of a further 8 weeks. On admission to our department there were no general signs and only a small submandibular lymph node remained. Mental state and cranial nerve examination were normal. He showed a symmetric distal muscle weakness in all limbs. A marked sensory ataxia was noted. We found mild root pain on a straight leg raising test. Deep tendon reflexes were very low to absent, especially in the legs. Plantar responses were flexor.

Serum concentrations of liver enzymes and glucose, and a protein spectrum were normal. Laboratory tests for adeno, RS, corona, influenza 1–2–3, parainfluenza 1–2–3, sendai, mumps, measles, herpes simplex and varicella zoster viruses, *Mycoplasma pneumoniae, Chlamydia psittaci, Coxiella burnetti, Mycobacterium tuberculosis* and atypical mycobacteria were negative. Antinuclear factor and ANCA were negative. Levels of complement factor 3 and 4 were normal. ELISA *B henselae* IgG in serum, 3.5 months after outbreak of lymphadenitis, was still >850 U/l. IgM specific antibodies now were <200 U/l. Examination of CSF showed 13 mononuclear leucocytes/μl, no polynuclear leucocytes, raised protein concentration (558 mg/l, normally 160–310 mg/l), three oligoclonal bands in CSF on isoelectric focusing, and a slight intrathecal IgG synthesis (51 mg/l, normally 3.2–15 mg/l). IgG and IgM specific antibodies against *B henselae* were negative in two CSF samples. Polymerase chain reaction (PCR) on *B henselae* in CSF and in serum was negative. Enolase, myelin basic protein, S100, lactate and glucose concentrations in CSF were normal.

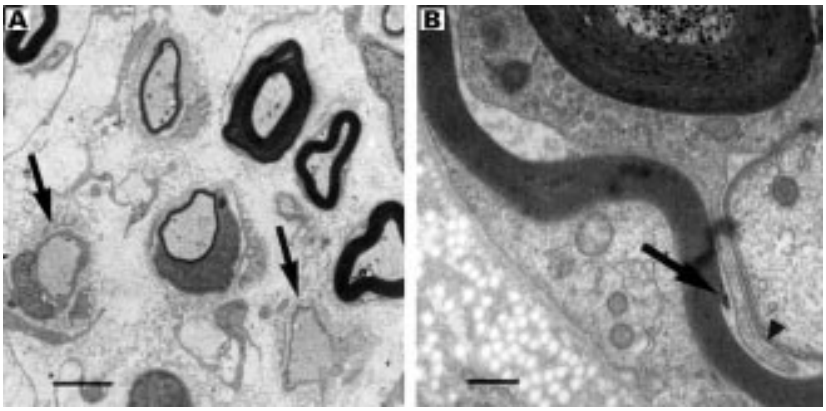
Electroencephalography, cranial CT, and gadolinium enhanced MRI of the thoracolumbar region were normal. Nerve conduction studies showed a marked decrease of motor nerve conduction velocities in all limbs (median nerve 20 m/s). In sensory conduction studies no response could be elicited. The EMG was normal.

The sural nerve biopsy showed many demyelinated axons and signs of early remyelination, but no onion bulbs (figure A). The density of myelinated fibres was slightly below normal, which could be attributed partly to intrafascicular oedema. Myelinated fibre size histogram was bimodal. There were no endoneurial infiltrates, or signs of vasculitis. Electron microscopy showed macrophage induced demyelination (figure B). Polymerase chain reaction (PCR) on *B henselae* in sural nerve fragments was negative.

The history and clinical presentation, combined with CSF findings and the results of EMG and light and electron microscopy studies, are compatible with the diagnosis of chronic inflammatory demyelinating polyradiculoneuropathy (CIDP).

He was treated with prednisone (15 mg every other day for 1 month) and the dose was reduced slowly in the course of 4 months. His motor and sensory function recovered completely and deep tendon reflexes reappeared. Nerve conduction studies 1 year after the onset of CSD were normal.

Neurological complications of CSD are rare and predominantly of the CNS. Myelitis has been described in combination with encephalopathy[2] and radiculopathy was reported only in combination with encephalomyelitis. Ophthalmological problems are oculoglandular disease of Parinaud[3] and neuroretinitis.[4] In a study by Carithers and Margileth of 76 patients with CSD and neurological complications, 15 patients showed signs of dysfunction of cranial or



(A) Sural nerve. Low power electron micrograph. Demyelinated fibres (arrows) and thinly remyelinated fibres. Bar=2.0 μm. (B) Sural nerve; detail of myelinated fibre. Macrophage has invaded the myelinated fibre; process of macrophage is extending between axon and myelin sheath (arrow head). The damaged inner major dense line is terminating as a small dark swelling (arrow). Bar=0.2 μm.