Body temperature was normal. White blood cells were elevated (15000/mm^3) and became normal (5800/mm^3) within 3 days. CSF analysis revealed 19 white cells/mm^3 (with 85% lymphocytes and 3% plasma cells), normal glucose, lactate and protein content without IgG oligoclonal bands. The EEG was normal. On day 2, serum antibody testing (ELISA) revealed positive mumps virus IgM titres up to a dilution of 1:320 and negative IgG (for dilutions $\geqslant$1:40). CSF antibody testing was negative (for dilutions $\geqslant$1:10). On day 51, mumps virus IgG titres (ELISA) in the serum were positive for dilutions up to 1:640 and negative for IgM (for dilutions $\geqslant$1:40). There was complete clinical recovery within 7 weeks and thermic stimulation showed improved vestibular function on the left (hyporesponsiveness of 43%). CSF was normal.

Sudden deafness with or without abnormal caloric responses is a possible complication of epidemic parotitis.[1 2] Several investigators[3] have suggested that subclinical mumps infections without parotitis may produce sudden unilateral hearing loss sometimes with vertigo in adults, but the relationship of the mumps infection proven by positive blood serology and the onset of symptoms was not clear.

In our patient, hearing was spared and only unilateral vestibular paralysis accompanied the serologically proven acute mumps infection. This condition has not been described previously. The Kilham strain of the mumps virus and a neurotrophic mumps strain (isolated from the CSF of a child with mumps meningitis) may infect the endolymphatic structures of the labyrinth.[4] Neurons of the vestibular and cochlear ganglia were regularly infected by the Kilham strain but only occasionally by the neurotrophic strain.[4] Infections of the labyrinthine endolymphatic structures or of the vestibular ganglia may be followed by vestibular paralysis and such a mechanism seems most likely in our patient.

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Ataxic hemiparesis with cheiro-oral syndrome in capsular infarction

Ataxic hemiparesis (AH) is defined as an unusual combination of ipsilateral pyramidal and cerebellar signs. AH is ascribed to lacunar stroke in both supratentorial and brainstem locations.[1] Several reports[2–6] have established that accompanying hemihypesthesia indicates a thalamocapsular rather than a pontine location for lesions causing the AH syndrome. We describe a variant of AH with sensory impairment restricted to a cheiro-oral topography, in association with contralateral small posterior capsular infarction.

A 73 year old woman, known to have been hypertensive and diabetic for 10 years, suddenly developed slurred speech and a tingling sensation around the left corner of the mouth and in her left thumb, index and middle fingers. Soon after she noticed weakness in her left arm and leg. On admission the next day, general physical examination was normal. Blood pressure was 220/110 mmHg. She was alert and had normal mental status. Her speech was dysarthric. There was slight left central facial weakness and a grade 3/5 left hemiparesis. Reflexes were increased on the left, with Babinski's sign present. Pinprick and light touch sensation were diminished in the left tongue, buccal mucosa, lips and cheek, and in the thumb, index and middle fingers of the left hand. Proprioceptive sensation was unimpaired. The patient was unable to walk alone and fell to the left. ECG and routine laboratory tests were normal except for a blood glucose level of 207 mg/dl.

By the sixth day, her power improved to grade 4/5. It was at this time that she appeared to have moderate dysmetria and intention tremor of the cerebellar type in performing the heel-shin and finger-nose tests on the left. The ataxia was more apparent in the leg than in the arm. CT scan obtained at that time revealed a small hypodense area in the right posterior limb of the internal capsule with possible involvement of the adjacent lateral thalamus (see below). Parietal-recorded somatosensory potentials following contralateral median nerve stimulation were studied two weeks after clinical onset. The short-latency components from the left side showed decrease in amplitude of N20 to 35% of the normal side and slight attenuation of the subsequent peaks. Latencies were normal.

Eight months after onset, there was no weakness but minimal ataxia of the left extremities persisted. Paresthesiae remained in the tips of the left first to third fingers and hypoesthesia for pain and light touch continued in the left perioral region. CT scan again revealed the capsular hypodense lesion (figure). Somatosensory evoked potentials did not show any asymmetry or abnormal responses.

Originally, persistent sensory disturbances were not considered as part of the AH syndrome.[1] In subsequent reports,[2–6] the clinical spectrum of AH was widened to include cases with persistent hemisensory deficit ipsilateral to the cerebellar-like ataxia and pyramidal tract weakness. This has been termed "hypesthetic ataxic hemiparesis". The sensory loss involves several sensory modalities, more often of the spinothalamic than the dorsal column type. Huang and Lui[2] indicated that patients with capsular AH are likely to have sensory loss, while those with a pontine lesion do not. Hypesthetic AH was attributed to anterior choroidal artery territory infarction by Helgason *et al*,[3] with lacunar infarcts localised by CT or MRI to the posterior limb of the internal capsule. This anatomical distribution is the same as that found in our case. A small haemorrhage of the posterior limb of the internal capsule is thought to produce an AH with hemisensory loss.[4] Hypesthetic AH has also been correlated to infarction[5] or haemorrhage[6] of the thalamus.

AH with cheiro-oral sensory deficit, as seen in our patient, has not been previously reported. The cheiro-oral syndrome is a sensory disturbance affecting both the hand and the corner of the mouth on the same side, but without motor impairment. It usually occurs as a result of a vascular lesion in the region of the ventroposterior nucleus of the thalamus and in the brainstem sensory pathways projecting to this nucleus.[7] Selective involvement of some, rather than all, fingers (as described in our case), may also occur as a result of parietal cortical infarction.[8 9] In cheiro-oral syndrome due to a thalamic infarct, the sensory deficit exactly conforms to the representation of body surface in the ventroposterior nucleus, the fingers being represented adjacent to the tongue in the most medial pole of the nucleus.[10] In our case, involvement of the ventroposterior nucleus might be responsible for the sensory deficit, because CT revealed infarction in the posterior limb of the internal capsule bordering the adjacent thalamus. However, the foot could have sensory change rather than the face and fingers, in view of the fact that the foot is represented laterally in the ventroposterior nucleus and most adjacent to the internal capsule. This suggests that the patient's symptoms are most probably due to involvement of sensory thalamocortical radiation, which occupies the posterior part of the posterior limb of the internal capsule.[11] Abnormalities in short-latency components of somatosensory evoked potentials, as those observed in our patient, have been reported as being the same in lesions of the thalamus and thalamo-cortical radiations.[12]

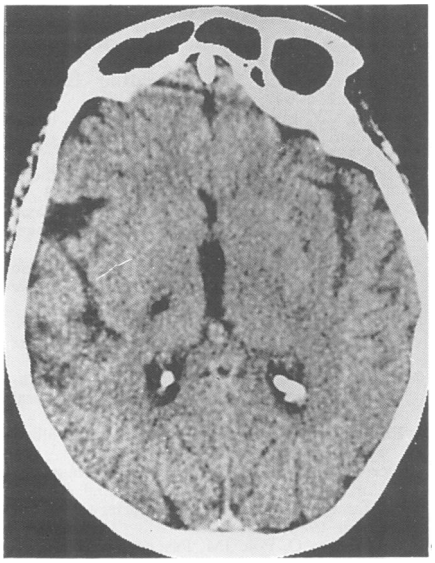

Figure CT scan taken at 8 months of onset showing lacunar infarct in the right posterior limb of the internal capsule.

In summary, ataxic hemiparesis associated with ipsilateral cheiro-oral syndrome is a previously undescribed symptom complex following capsular infarction. This association should be included in the spectrum of clinical syndromes presumably related to anterior choroidal artery territory infarction.

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Pavor nocturnus from a brainstem glioma

Pavor nocturnus or night terrors usually occur in the absence of identifiable neuropathology. This report documents pavor nocturnus associated with a brainstem lesion.

A 15 year old boy with headaches and ataxia had a brainstem tumour on CT (see fig). The patient underwent resection of a grade I cerebellar astrocytoma arising from the fourth ventricle and adherent to the brainstem. His postoperative examination disclosed dysarthria, spontaneous vertical nystagmus, bilateral sixth nerve paresis, decreased sensation on the face bilaterally, facial diplegia, sensorineural hearing loss, mild dysmetria and ataxia, and decreased reflexes with bilateral Babinski responses.

Postoperatively he developed a sleep disorder where he suddenly sat up in bed, screamed, and appeared to be staring in fright. During these episodes he was agitated and would try going over the rails of the bed or would thrash about in bed screaming. After one or two minutes, he promptly fell back to sleep. The patient had incomplete recollections of these episodes. Sometimes the only evidence of an episode was injury or blood on the floor. At other times, he recalled being frightened by images of parts of people sticking out of walls or by the belief that the bedposts were his room mates restraining him. The patient subsequently became depressed and had aggressive outbursts and paranoid beliefs. Before his tumour, he did not have a history of neurological or psychiatric difficulties, and there was no family history of a sleep disorder.

At the age of 24, polysomnography documented his night terrors. Spontaneous arousals punctuated all stages of sleep (12 a night), particularly stage three and four sleep. The patient's arousals from slow wave sleep were typically sudden and included restlessness, vocalisation, and looking accompanied by interspersed alpha and delta activity on electroencephalography (EEG). After starting clonazepam (0·5 mg at bedtime), the episodes of night terrors decreased, but he developed enuresis.

There are few documented cases of pavor nocturnus from neurological disturbances.[1] Reports suggest that night terrors may occur as a consequence of a right temporal lobe seizure focus.[2] This report documents pavor nocturnus associated with a fourth ventricular brainstem lesion.

Pavor nocturnus is usually not associated with any psychiatric or neurological disturbance when it occurs in children.[1 3] Its natural course is to disappear by adulthood,[3] suggesting a disorder of maturation of the nervous system. When night terrors begin in adolescence or adulthood, there is a likelihood of psychological difficulties.[4] Patients with night terrors may repress their anger or aggression and show elements of obsessive-compulsive and phobic behaviour as well as depression.[4] Our patient has no difficulty expressing anger and does not have obsessive-compulsive behaviours. Rather, he has a brainstem lesion disrupting non-rapid eye movement (REM) sleep and resulting in night terrors. Furthermore, clonazepam which suppresses slow wave sleep has decreased his night terrors while unmasking enuresis.

Pavor nocturnus is a consequence of partial arousal during non-REM slow wave sleep. During deep sleep, the brain continues to process subcortical sensory information such as respiratory awareness.[5] With arousal from non-REM sleep, these sensory experiences may intrude into consciousness as single, brief, frightening experiences. Patients are terrified and may even injure themselves trying to escape the frightening image or feeling. In this patient with a brainstem lesion, the mechanism of slow wave arousal may be disruption of the nucleus of the solitary tract in the dorsum of the brainstem near the wall of the fourth ventricle. Activation of this nucleus promotes sleep and may modulate the arousal properties of the ascending reticular activating system.[6] Polysomnographic studies of patients with comparable brainstem lesions would help elucidate the pathophysiology of pavor nocturnus and the proposed role of the nucleus of the solitary tract.

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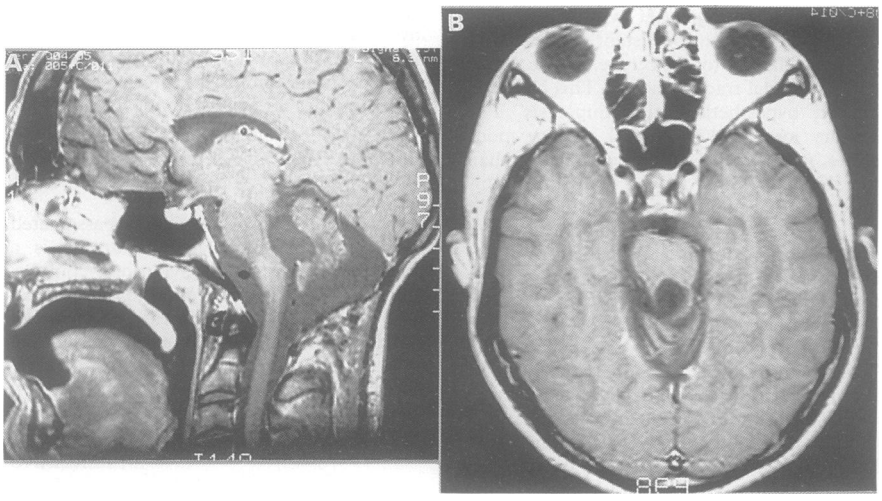

Figure Cerebellar astrocytoma adherent to brainstem.

Octreotide—a new treatment for diarrhoea in familial amyloidotic polyneuropathy

Familial amyloidotic polyneuropathy (FAP) is a well known hereditary polyneuropathy which was reported for the first time by Corino de Andrade in Portugal.[1]

This disease generally begins with sensory symptoms and signs (50%) and sexual impotence in man (30%). A less significant number of subjects have constipation (20%), loss of weight (10%) or diarrhoea (10%) as an initial symptom. After two to five years of other dysautonomic symptoms are very common, namely orthostatic hypotension and severe diarrhoea.

In a group of 60 patients followed at the neurology outpatients, we found that half had regular diarrhoea which was particularly refractory in six. To control the diarrhoea we used a low fibre diet, antibiotics (tetraciclines and metronidazole), metoclopramide and loperamide.