Hypothermia due to hypothalamic involvement in multiple sclerosis

Hypothermia can be caused by several different mechanisms, the most common being environment, neglect, drug overdose, and endocrine disturbance such as hypoglycaemia, hypo- and hyperthyroidism. Disease affecting the hypothalamic regulatory centre can rarely cause hypothermia. We describe the first case of hypothermia in association with pathologically established hypothalamic damage due to multiple sclerosis.

A 53 year old woman with longstanding, severe, clinically definite multiple sclerosis presented with a five day history of dysphagia and lethargy. She was well cared for, in an adapted and centrally heated flat. Her skin was warm and well perfused. Although she was dysarthric with a severe spastic tetraparesis, examination of eyes, there were no new neurological signs. A relapse of multiple sclerosis was provisionally diagnosed and arrangements were made to admit her to the ward the next day.

Over the next 24 hours her conscious level deteriorated to a Glasgow coma scale score of 9. She developed a sinus bradycardia of 45/min with a blood pressure of 80/50 mm Hg. Her oral temperature was 35.9°C and her rectal temperature was 29°C. She was appropriately dressed for the ambient temperature of 18°C. Her skin did not feel abnormally cool and there was no shivering. General examination disclosed a few coarse crepitations scattered throughout both lungs. There was no meningois. The pupils were constricted and the corneal reflexes depressed. The jaw was tightly clenched. There were no tetraplegic signs with extensor plantars but the reflexes were now depressed.

Haematological investigations showed a picture consistent with mild disseminated intravascular coagulation (platelets 79; activated partial thromboplastin time 45 s (control 32 s); TCT 17 s (control 14 s); INR 1-0). Electrolytes and glucose were normal but the amylase concentration was slightly raised at 321 IU/l, suggesting mild pancreatitis. An ECG showed widespread J waves, the presence of which had led to the taking of the core temperature and thus the diagnosis. Chest radiography showed mild bilateral basal shadowing. Blood and urine toxin screens were negative.

The patient was slowly warmed and treated with intravenous atropine and antibiotics. However, she had recurrent dysrhythmias, predominantly bradycardia in nature, and bronchopneumonia. She died three days later.

Postmortem examination confirmed the presence of bronchopneumonia and pancreatitis. The brain and spinal cord contained numerous plaques of demyelination typical of chronic multiple sclerosis. There was a large plaque involving the hypothalamus (figure). This had the grey, translucent appearance typical of a mature gliotic plaque. However, at the microscopic level there was evidence of current activity in a proportion of the plaques, including the one involving the hypothalamus. In addition to gliosis, lymphocytic cuffing of vessels and occasional macrophages containing lipid debris were seen, providing evidence of continuing demyelination. The corpus callosum appeared unremarkable.

The centre for temperature regulation is situated in the hypothalamus and commonly disorders in this region can cause hypothermia or hyperthermia, which can be continuous or paroxysmal. Disrupted thermoregulation, secondary to hypothalamic damage, has been described with a wide range of pathological processes including tumour, trauma, infarct, haemorrhage, sarcoidosis, Wernicke's encephalopathy, ventricular shunt embolism, and fat embolism. A combination of hypothermia, hyperhidrosis, and other autonomic abnormalities are occasionally seen in cases of concomitant hypothalamic damage with corpus callosum dysgenesis.

The precise anatomical lesion within the hypothalamus leading to hypothermia is unclear but may involve areas of the anterior hypothalamus (as in episodic hypothermia) or the posterior hypothalamus (as in Wernicke's encephalopathy). There are few reported cases of hypothermia in multiple sclerosis. The disease process primarily affects white matter and, until now, hypothalamic disease has never been pathologically established. Sullivan et al described two patients with clinically definite multiple sclerosis who presented with acute hypothermia and on recovery developed chronic hypothermia. No pathological lesion was proved, but thermoregulatory studies suggested a central hypothalamic defect, presumably secondary to demyelination.

Lammans et al described three patients with multiple sclerosis who presented with coma and hypothermia. Pathological examination in one case showed no abnormality. Finally, Ghaswhe et al reported a woman with multiple sclerosis who presented with three episodes of hypothermia. Hypothalamic disease was considered to be the likely cause, but repeated enhanced MRI failed to show a lesion in this region.
The physiological response to hypothermia is controlled by the hypothalamus, involving vasconstriction and shivering. In hypthalamic hypothermia these systems fail with loss of reactive peripheral vasconstriction to reduce heat loss and loss of the shivering response to produce heat. It is the failure of these systems that contributes to the hypothermia and also produces diagnostic difficulty, with the patient feeling warm to the touch and not shivering. The ECG showing the pathognomonic J waves, with absence of shiver waves mirrored the hypothalamic cause of the hypothermia.

This is the first description of hypothermia associated with multiple sclerosis with a proved hypthalamic plaque and no other identifiable cause for hypothermia.

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Lyme neuroborreliosis presenting with proprispinal myoclonus

A 60 year old white woman presented with an arthralgia myalgia and a tick bite on the right thigh on 11 July 1995. On 2 October 1995, she complained of a lumbar pain which radiated to the right thigh. She received dextropropoxyphene, paracetamol, thiocholchicoside, and tetrazenam and then was additionally treated with codeine, chlorzemanone, and tenoxicam. Despite this, the pains, which prevented sleep, rapidly radiated bilaterally to the low back and abdominal region. On 11 October 1995 she presented with flexor non-rhythmic symmetric jerks of the trunk, the abdomen, both hips, and knees evident both sitting and standing. The increasing when lying, without suppression by an effort of will or during voluntary movements. Although the pains and jerks were atypical, the patient was diagnosed as having a herniated disc; an epidural infiltration of dexamethasone (10 ml) gave a transient relief of the pains and jerks. A second infiltration was not effective. On 13 October the patient was admitted to hospital.

The myoclonic jerks had reinforced, occurring sometimes in bursts, occasionally involving the neck and the shoulders but never the face. The patient was agitated and exhausted, and cried on account of the continuous distressing pains. She was free of headache and fever. Walking was difficult, although she felt better when standing. She had a mild to severe proximal paralysis of the lower limbs, patellar tendon reflexes abolished on the right, diminished on the left, normal ankle reflexes, plantar reflexes flexor, and axial muscles and neck were not rigid. Respiratory and auditory investigatons were normal. Ketoprofene, haloperidol, clorazepate, and then tiaprid, paracetamol, and buprenorphine were tried with negligible effects. On 28 October, she was transferred to the intensive care unit.

The painful jerks were flexor, simultaneous in all the muscles, and spontaneous or induced consistently by flexion of the neck, without initial acceleration and superior limbs. The intervals between the jerks became so short that the paroxysms gave the impression of being attacks of sustained truncal flexion. An EEG during jerking was unremarkable. Finally, the patient was anesthetised and ventilated artificially. The treatment was propofol, fentanyl, and muscle relaxant pancuronium. Ceftriaxone (2 g intravenously daily) was given for 14 days.

The CSF contained 398 mononuclear cells/µl, numerous atypical cytological features, normal glucose and chloride ratios, increased protein content (1:2 g/l), intrathecal synthesis of IgG and IgM, and three oligoclonal bands were detected. The titre of antibodies to Borrelia burgdorferi was raised in the CSF [1/64: normal <1/4] by indirect immunofluorescence, both for IgM (1/16) and IgG (1/16), 1:352 (normal <0.16) by enzyme linked immunosorbent assay (ELISA) (Immunowell borrelia Lyme—BMD); their detection in serum was negative three weeks later. On 28 October the patient was extubated. The jerks had totally disappeared and the pains dramatically improved. At this time, EMG failed to detect any myoclonic jerks. Recording of peroneal nerve somatosensory evoked potentials and MRI of the spine were unremarkable. On 24 October, the patient was free of pain and then recovered full strength and normal tendon reflexes.

The clinical features of pain resistant to analgesic agents, meningoradiculitis with a history of tick bite, and erythema migrans strongly evokes a Lyme neuroborreliosis confirmed by the detection of antibodies to Borrelia burgdorferi.1 However, the most dramatic feature was the myoclonic jerks which support the clinical diagnosis of proprispinal myoclonus characterised by repetitive, non-rhythmic jerks of the neck, trunk, both hips, and knees.1 Sometimes attacks of sustained truncal flexion are generated by paroxysmal bouts of abdominal jerks.1 In this type of myoclonus, the discharge arises from a limited segment of the spinal cord and then spreads slowly up and down by the involvement of the long propriospinal pathways.2 The jerks had appeared at the time of the EMG investigation in our patient. Accordingly, we could not ascertain the possible origin in the thacacic segment of the spinal cord, corresponding to the abdominal and lumbar muscles, which were painful throughout the course of the disease and constantly affected by the jerks. To our knowledge, no case of Lyme neuroborreliosis has been associated with a proprispinal myoclonus.

Another patient had stiffness, painful cramps, and spasmodic jerks confined to the left leg,3 which suggests a localised myelitis of the spinal interneurones: their own strongly evokes the involvement of many spinal segments. Apart from the myoclonus, no other evidence of spinal cord disease was apparent. The patient in question had relieved the pain and dramatically suppressed the myoclonus.

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Metamorphosis and visual hallucinations restricted to the right visual hemifield after a left putaminal haemorrhage

Metamorphopsia is a rare neurological phenomenon which objects appear distorted in form. Many reports have attributed the responsible lesion to the occipitoparietal cortex and its related structures. We reported a case of left putaminal haemorrhage followed by metamorphopsia and visual hallucinations restricted to the right visual hemifield. The origin of this patient's symptoms was considered to be the left optic radiation.

A 63 year old right handed man with a previous history of hypertension was admitted to the hospital with acute right hemiparesis. On admission, his visual field examination showed a right homonymous hemianopia and a right homonymous hemianopia without sensory involvement. The right homonymous hemianopia disappeared on the third day. On the fourth day, he complained that the doctor's left cheek seemed to have been scraped, that the doctor's left hand seemed tortuous, and that some of the fingers of the hand seemed to be missing. He drew a picture of what he saw (fig1A). Visual field examination by confrontation was immediately performed but no abnormalities were found, later confirmed by using Goldmann's perimeter. On the next day, he complained, "The right half of the curtain in front of me suddenly transforms into an animal's face. It rotates there for a while and finally flows to the right, and then disappears. At the next moment, another face springs up at the very portion and . . . ." He then drew a picture to illustrate his experience (fig1B). These phenomena lasted three to four days and then disappeared. One month later, he was able to walk without assistance and was discharged from hospital.

The laboratory analysis of blood and urine was within the normal range. Cranial CT on admission showed a left putaminal haemorrhage without ventricular extension.