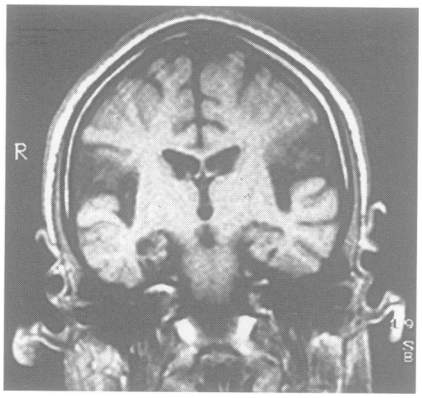

Patient with acquired bilateral opercular syndrome and eating epilepsy: coronal T1 weighted MRI showing focal atrophy in both rolandic opercula.

ground activity with focal interictal slow spikes and waves over the right centrotemporal region, not increased during sleep. No ictal recording was obtained.

Voluntary control of facial muscles is dependent on the normal function of the motor cortex and of the corresponding pyramidal tract. Subcortical structures, particularly the basal ganglia, thalamus, and subthalamic nuclei, are involved in automatic or emotional movement. A bilateral lesion involving the anterior opercular region is manifested by facial diplegia with dissociation of voluntary and involuntary movements.[1] Pyramidal signs, cheiro-oral paresthaesiae, and severe speech disturbance or even mutism with unimpaired language comprehension may be due to or associated with bilateral opercular lesions depending on their symmetry and extent.[1] Symmetric lesions of the posterior limbs of both internal capsules may produce a picture similar to AOS because the projections from the anterior opercular cortex are represented there. The differential diagnosis of AOS, therefore, is based not only on bulbar dysfunction but also on other clinical features of pseudobulbar palsy, such as those due to striate or pontine lesions.[3] The absence of automatic-voluntary dissociation and affective lability point to the presence of AOS.[1] Bilateral stroke is the main aetiology of AOS and the two events may be separated in time. A unilateral lesion may lead to AOS in a patient who already has contralateral opercular dysfunction on the other side.

The occurrence of AOS in childhood is exceptional. In most cases the syndrome is of developmental origin with polymicrogyria most likely due to prenatal vascular lesions. Infections of the CNS are also an important cause of the syndrome in children.[2] There are well documented cases with transient bilateral rolandic dysfunction and features resembling AOS due to an epileptic disturbance.[4]

In this patient, reflex seizures were triggered by a specific stimulus, the act of eating. Eating epilepsies have been subclassified into two distinct groups with either temporal or opercular onset, according to a recent anatomoclinical review.[5] In the opercular subgroup electroclinical studies pointed to the presence of an epileptogenic focus in the postcentral gyrus, frequently with evidence of a cortical structural lesion at that level. The physical characteristics and the type of food seem to be important factors in activating this form of eating epilepsy. Attacks may recur even when temporal structures and those anterior or posterior to the sensory motor strip have been resected. In addition to reflex seizures, spontaneous attacks may occur. Thalamic sensory afferents to the damaged cortex seem to play a predominant part in the genesis of seizures in these patients. By contrast, eating epilepsy of temporal origin is dependent more on the context of a meal and there are often additional clinical features suggesting temporal localisation. Such patients have no features of a postcentral lesion and a temporal resection usually leads to cessation.

Our patient clearly showed an AOS. He had no spontaneous seizures and we were not able to confirm that the characteristics of the food played a part in triggering the attacks. He is exceptional because of the association of two infrequent but related neurological syndromes. We stress the value of careful imaging and of inquiring about the circumstances of occurrence of attacks in diagnosing this syndrome and the associated reflex epilepsy.

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HTLV 1 associated myelopathy and adult T cell leukaemia-lymphoma in the same patient: report of a case

Tropical spastic paraplegia (TSP) is a chronic progressive myelopathy occurring in endemic areas around the world with particularly high prevalence in Japan, the Caribbean, parts of Africa, and South America. In 1985, Gessain and colleagues working on the island of Martinique identified the presence of anti-HTLV 1 antibodies in patients with TSP.[1] This finding was later confirmed in other tropical regions where this condition is endemic. In Japan, Osame *et al* called it HTLV 1 associated myelopathy (HAM).[2] The HTLV 1 retrovirus had previously been isolated and identified in 1980 by Poiesz *et al* as the aetiological agent of adult T cell leukaemia-lymphoma (ATLL).[3] Epidemiological studies have since shown the high prevalence of both ATLL and HAM/TSP in areas with a high HTLV 1 carrier rate. The virus has been isolated from patients with both the conditions and has been shown to be the same strain of the retrovirus. It is not known why some carriers develop chronic progressive myelopathy whereas others develop the haematological malignancy. The coincidence of both conditions occurring together as in our patient is extremely rare.

A 61 year old black Afro-Caribbean woman who had migrated to the United Kingdom from the Dominican Republic was seen with a four year history of progressive difficulty in walking and backache. She had also noticed a progressive deterioration in her vision and difficulty in micturation. Two and a half years later she was found to have a positive syphilis serology. A CSF examination was normal.

Neurological examination on presentation disclosed diminution of visual acuity in both eyes (N9 near vision) and the fundus examination showed bilateral optic atrophy. The other cranial nerves were normal. She had evidence of a spastic paraparesis, with a pyramidal distribution of weakness in the legs, exaggerated reflexes, and extensor plantar responses.

The tone and power in her arms were normal, although the reflexes were brisk. Sensory examination was normal to all modalities and there were no cerebellar signs. She was able to walk only with the help of a stick and she had a spastic gait.

Haemoglobin concentration was 10 g/dl and the white cell count was $10{\cdot}2 \times 10^9$/l. Magnetic resonance imaging of the cervical and dorsal spine was normal but MRI of the brain showed hyperintense signals in the periventricular white matter on T2 weighted images. Visual evoked responses were delayed bilaterally and the somatosensory evoked responses showed delay of N19 on stimulation of the median nerves at the wrists bilaterally. Antibodies to HTLV 1 were detected in the serum and a diagnosis of HTLV 1 associated myelopathy (HAM) was made.

Four months after the diagnosis of HAM, the patient had a rapid deterioration of power in her legs. She was febrile and delirious. She became progressively obtunded and her level of consciousness rapidly worsened. There was considerable tenderness in the long bones. The liver and spleen could not be palpated. Haemoglobin was 10 g/dl, and the white cell count was 66×10^9/l. The peripheral smear showed a pronounced lymphocytosis and presence of abnormal lymphocytes with multilobulated nucleii with clover and cleaved forms. About 20% of the lymphocytes showed abnormal multilobulated nucleii. Leukaemia typing confirmed that the abnormal cells were all T lymphocytes (CD-2 count 98%, CD-3 count 97%, CD-4 count 76%, and CD-5 count 0%) and a diagnosis of adult T cell leukaemia was made. Examination of CSF, skeletal survey, and serum calcium concentrations were normal. The patient received supportive treatment but she soon deteriorated and died.

The occurrence of ATLL in a patient with HAM is extremely uncommon and has been described in only three other patients.[4-6]

It has been shown that the viruses isolated from patients with HAM and ATLL are identical in their genomic composition. The HTLV 1 carrier rate has been estimated to be 15% in the general population in Japan, and 5% in the Caribbean. The lifetime risk of developing ATLL if infected with HTLV 1 is 2% to 5% with an interval of about 30 years between acquiring the infection and developing symptoms. On the other hand the lifetime risk of developing HAM/TSP has been estimated to be 0·25%. Familial clustering of both ATLL and HAM is well recognised but the occurrence of both the conditions in the same family is extremely uncommon. Shoji *et al.*[7] describe the case of a 37 year old Japanese female patient with HAM, whose father had previously died of ATLL.

Why the two diseases, although caused by the same virus, do not occur in the same person, or for that matter in the same family, is not clear. The magnitude of the immune response in patients with HAM tends to be higher, as evidenced by the higher titres of the anti-HTLV 1 antibodies in the serum as well as the CSF. By contrast the titres tend to be lower in controls as well as in patients with ATLL. The degree of immune responsiveness is related to host genetic influences and the existence of HAM associated haloptypes and ATLL associated haplotypes has been suggested. Moreover, in vitro studies in patients with HAM have shown a high lymphocyte proliferation rate, spontaneously as well in response to stimulation by mitogens and HTLV 1 viral antigens, compared with asymptomatic carriers or patients with ATLL. Also, the virus integration site into the host genome in HAM is random, whereas it integrates at a very specific locus in ATLL. The monoclonal integration of proviral DNA in ATLL consists of the long terminal repeat 5'3' "tax" gene, the product of which induces interleukin 2 receptor expression and T cell proliferation.

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MATTERS ARISING

Magnetic resonance spectroscopic study of parkinsonism related to boxing

We read with interest the paper by Davie *et al* reporting a study of proton magnetic resonance spectroscopy (MRS) in three exboxers with parkinsonism.[1] They report a significant reduction in the absolute concentration of N-acetyl-aspartate (NAA) in the putamen and globus pallidus in the boxers with a parkinsonian syndrome compared with patients with idiopathic Parkinson's disease and controls. They speculate that the reduced NAA may result from neuronal loss in the corpus striatum secondary to head trauma. In support of this hypothesis reference is made to their previous study in which it was reported that NAA is reduced in the lentiform nucleus in patients with striatonigral and olivopontocerebellar variants of multiple system atrophy compared with patients with idiopathic Parkinson's disease and controls.[2]

This interpretation may be too simplistic. We have carried out a pilot study using MRS in 10 patients with idiopathic Parkinson's disease with motor response fluctuations on chronic levodopa treatment (satisfying the United Kingdom Brain Bank criteria for diagnosis of idiopathic Parkinson's disease) and seven healthy age matched controls using a voxel size of 4 ml centred on the putamen and one cerebellar hemisphere.[3,4] We found a consistent and striking reduction in NAA/creatine and NAA/choline ratios in the putamen in patients with idiopathic Parkinson's disease but not in controls. The choline/creatine ratios between controls and idiopathic Parkinson's disease in the putamen and cerebellum were unchanged suggesting that the changes seen were due to changes in MR-visible NAA itself. Repeat studies in two patients three months later, with regions of interest centred on the putamen bilaterally, showed similar reductions in the observed NAA signal.

These findings contrast with the results reported by Davie *et al*[1] and raise several questions about the importance of localised changes in brain NAA in idiopathic Parkinson's disease and related disorders.[2] Firstly, the exact positioning of the region of interest and voxel size are both likely to be crucial. The spectra analysed by Davie *et al*[1,2] were obtained from a voxel centred on the globus pallidus and striatum, whereas ours was restricted to the putamen. Striatopallidal degeneration is a feature of multiple system atrophy, but not (so far as is known) of idiopathic Parkinson's disease.[5] The findings of Davie *et al* thus may reflect the pathological changes in the pallidum rather than in the putamen. Certainly, it is not possible to conclude from the study of Davie *et al*[1] that striatal NAA concentration is unchanged in idiopathic Parkinson's disease compared with multiple system atrophy and other related disorders.

Similarly, in the study of Holshouser *et al* in which there were no significant differences in "striatal" NAA/creatine ratios between patients with idiopathic Parkinson's disease and normal controls (but a significant reduction in NAA/choline ratios in patients with idiopathic Parkinson's disease between 51 and 70 years of age compared with controls), the region of interest was centred wholly on the globus pallidus, not in the putamen and a much larger voxel size (8 ml) was used.[6] Furthermore, Holshouser *et al*[6] reported that choline/creatine ratios in idiopathic Parkinson's disease and controls were in the normal range and it is surprising, therefore, to note that they found significant reduction in NAA/choline and not NAA/creatine ratios in idiopathic Parkinson's disease. Thus at present conclusions on the relevance of changes in NAA concentration or NAA/creatine ratios in the "striatum" in idiopathic Parkinson's disease, multiple system atrophy, and other neurodegenerative disorders such as progressive supranuclear palsy or parkinsonism in boxers are premature.[1,7] Our finding of reduced NAA/creatine and NAA/choline ratios in the putamen could reflect a functional change, loss of nigrostriatal dopamine terminals, or loss of intrinsic striatal neurons, or a combination of these factors. Diagnostic error is another possibility as the present diagnostic criteria of idiopathic Parkinson's disease has an accuracy of 82% but the reduction in NAA/creatine ratios were consistent in most of our patients diagnosed with the disease.[8]

Further work is needed to establish the best paradigms for acquiring spectra in idiopathic Parkinson's disease and related disorders to decide whether striatal (putaminal) NAA really is reduced, and to understand what this means diagnostically and in terms of neuronal dysfunction and pathology.

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