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### Tobacco-alcohol amblyopia: magnetic resonance imaging findings

Sir: Tobacco-alcohol amblyopia (TAA) is a condition occurring in smokers which causes progressive bilateral visual deterioration. It is commonest in middle-aged and elderly men and malnutrition and excessive alcohol consumption tend to increase the incidence and severity of the condition.<sup>1</sup> The characteristic visual deficit is a centro-caecal scotoma, with loss of colour vision being an early feature.<sup>1</sup>

It has been suggested that TAA may be a consequence of the toxic effect of the cyanide constituent of tobacco smoke.<sup>1</sup> Cessation of smoking and hydroxocobalamin replacement usually result in visual improvement.<sup>2</sup> Pathological studies in TAA have failed to establish whether the primary location of the defect is retinal or in the optic nerve itself.<sup>3</sup>

Recent advances in magnetic resonance imaging (MRI) of the optic nerve have shown great sensitivity in the detection of optic nerve lesions. For example, in optic neuritis, lesions are found in 84% of clinically affected nerves.<sup>4</sup> We recently reported abnormal MRI signal in 12 of 15 affected optic nerves in Leber's optic neuropathy (LON),<sup>5</sup> a maternally inherited condition characterised by the acute or sub-acute onset of bilateral visual loss.<sup>6</sup> To search for evidence of optic nerve involvement in TAA we have performed optic nerve and brain MRI.

Five patients with TAA were studied. All had typical bilateral centro-caecal scotoma, high tobacco and alcohol consumption,

negative findings for other causes of visual loss and improvement in visual acuities after reduction in smoking. In two patients some improvement in vision had already occurred by the time of MRI (table). Scanning was performed on a Picker 0.5T Superconducting MRI, with axial SE<sub>2000/60</sub> brain images (5 mm contiguous slices), and coronal STIR (IR<sub>-1500/40/150</sub>, 5 mm contiguous slices) optic nerve images, as in the Leber's study.<sup>5</sup>

Scans were examined by an experienced neuro-radiologist (BEK). All optic nerve images were normal, in contrast to the findings in LON where most optic nerve images showed either definite or equivocal abnormalities.<sup>5</sup> One patient (Case 4) had small multiple high signal areas in the white matter, a non-specific finding in this age group.<sup>7</sup>

The results suggest that the optic nerve is not damaged in TAA, or that such damage that occurs is unlikely to produce abnormal MRI signal. However, given the sensitivity of MRI in other conditions, our findings do not provide evidence to support the suggestion that the primary insult in TAA is the optic nerve itself, and are consistent with a proposal from electrophysiological studies that retinal damage makes a significant contribution to the visual loss in TAA.<sup>8</sup> From the point of view of differential diagnosis we are able to conclude that MRI in TAA is usually normal and therefore may be useful to exclude other causes of visual loss when indicated.

It is interesting to compare these results with those mentioned above in LON.<sup>5</sup> Recently a specific point mutation in mitochondrial DNA has been proposed as the cause of LON.<sup>9</sup> This would result in an energy deficit similar to that caused by chronic poisoning with respiratory inhibitors, such as the cyanides<sup>9</sup> which are found in tobacco smoke.

There are significant clinical differences between TAA and LON. The results reported here provide further evidence against a common aetiology for the two disorders.

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### Opsoclonus-myooclonus following the intranasal usage of cocaine

Sir: A wide variety of neurological complications have been reported with cocaine usage. Opsoclonus-myooclonus is an uncommon syndrome thought to be of cerebellar-brain-

Table Visual acuities (VA) and colour vision on Ishihara plates refer to the worst affected eye.

At presentation	VA	Correct plates	VA	Duration of symptoms	Final
Case	Age	Ishihara	At scan	At scan	VA
1 M	57	1/60	0/14	6/36	6/36
2 M	50	6/60	3/12	6/60	6/6
3 F	42	6/12	6/14	6/5	6/5
4 M	54	6/36	0/14	6/36	6/6
5 M	39	6/24	9/14	6/12	6/12

stem origin. A case is presented of acute onset of opsoclonus-myoclonus following the intranasal usage of cocaine.

A 26 year old woman was admitted for uncontrollable shaking of her entire body, associated with nausea, vomiting, and an inability to keep her eyes still. The symptoms began when she awoke after having "snorted" cocaine late the previous night. The cocaine was the first from its batch and was provided by her usual supplier. It was not known whether the batch was adulterated, but it was established that the patient was the only person who reported an adverse reaction. There was no history of ingestion of alcohol, medications, or other drugs. The patient had never used intravenous drugs. Past medical history was notable only for a single seizure, associated with cocaine use, some eight years previously. Family history and review of systems were entirely negative. There was no fever, arthralgia, recent upper respiratory infection, focal weakness or weight loss. The patient was a college student, living with her parents, and had been "snorting" cocaine once a week for many years.

On examination, she was a young, thin, white woman in marked distress. She was unable to sit upright in bed without severe myoclonic jerking involving trunk and extremities. Her pulse was 110 and regular. Respirations were 20 per minute and not laboured. The blood pressure was 118/68 mm Hg and the oral temperature was 36.7°C. There were no rashes or petechiae. The general examination was normal except for ocular abnormalities and periodic myoclonic jerking. Neurological examination revealed that the patient was alert and fully oriented. The optic fundi were poorly visualised due to opsoclonus; visual fields were intact to confrontation. The pupils were equal and reactive to light and accommodation. Extraocular movements were full. Marked continuous opsoclonus was present; spontaneous conjugate jerking was observed in all directions, as well as in a clockwise rotatory direction. The remaining cranial nerves were normal, and there was no palatal myoclonus. Motor strength, tone, and sensory modalities were all normal. Deep tendon reflexes were 2+ and symmetrical. Plantar reflexes were flexor. There was severe myoclonus involving all extremities; titubation and prominent truncal ataxia were present. Myoclonus increased with arm movement, on finger to nose and finger to finger testing. The patient was unable to sit upright without assistance; efforts to do so elicited vertigo, nausea and, on one occasion, vomiting.

The patient was admitted to the neurology

unit and sedated with lorazepam. Attempts to control the myoclonus with valproic acid and clonazepam met with moderate success. CT, MRI brainstem auditory evoked potentials, electroencephalogram, cultures, serologies and the remainder of an extensive workup failed to yield a definite aetiology. The syndrome proved self-limiting over a period of four weeks. All medications were discontinued two weeks later. At follow up one year later, a careful neurological examination failed to yield any abnormalities. There have been no further episodes of opsoclonus or myoclonus and the patient reports excellent health.

Opsoclonus is defined as periodic, jerking, ataxic conjugate eye movements occurring in all planes and sometimes in a rotary fashion. It is usually part of a syndrome seen with myoclonus of the trunk and limbs along with cerebellar dysfunction. Opsoclonus-myoclonus has also been called "dancing eyes, dancing feet" and the syndrome of "lightning eyes", among other descriptive terms.

Despite reported clinico-pathological studies, there is no consistently found structural lesion in opsoclonus or opsoclonus-myoclonus. Opsoclonus is thought to occur when there is a loss of inhibitory control over the system that produces ocular saccades. The loci of control have been variously placed in the pontine paramedian reticular formation, cerebellum, superior colliculus, midbrain tegmentum, pretectal region of the rostral dorsal midbrain, thalami, and parts of the cerebral hemispheres.<sup>1,2</sup> Thus, there are probably several sites in the CNS that, when disrupted structurally or functionally, can result in the clinical picture of opsoclonus-myoclonus.

The differential diagnosis of opsoclonus-myoclonus includes a wide variety of disorders. It has been associated with demyelinating disease, namely brainstem involvement in multiple sclerosis; structural lesions such as glioma; hydrocephalus in a child; trauma; degenerative diseases; metabolic disturbances such as hyperosmolar non-ketotic coma; and in vascular events such as thalamic haemorrhage, and vertebral-basilar insufficiency. Opsoclonus-myoclonus has been reported in a variety of encephalitides, both viral and bacterial. Classically, opsoclonus-myoclonus was seen as a paraneoplastic manifestation of neuroblastoma. It has also been reported as a paraneoplastic syndrome of a number of other malignancies.

Finally, a variety of drugs and toxins have been associated with the syndrome of opsoclonus-myoclonus. These include chloroquine poisoning, the combination of lithium and haloperidol lactate, amitriptyline hydro-

chloride, thallium, phenytoin, and toluene. Despite the long list, the exact pathogenesis of these insults, be it autoimmune, directly toxic or compressive, remains obscure.<sup>1-5</sup>

To the best of our knowledge, this is the first reported case of opsoclonus-myoclonus in association with cocaine usage. The self limiting nature of the syndrome in this case argues against a structural lesion or a paraneoplastic syndrome. It is tempting to postulate a direct toxic effect of cocaine, or a toxic effect of one of the many substances that are used to "cut" cocaine. Alternatively, one might postulate that the opsoclonus-myoclonus was the result of a mild encephalitis or an immune complex-mediated phenomenon. Extensive investigation failed to support any of these hypotheses.

In summary, cocaine usage should be added to the list of toxic substances thought to induce the syndrome of opsoclonus-myoclonus. This striking neurological finding has a long list of suggested aetiologies, while its pathogenesis remains elusive.

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#### Autosomal dominant cerebellar ataxia with deafness, myoclonus and amyotrophy

Sir: Autosomal dominant cerebellar ataxia (ADCA) can be associated with ophthalmoplegia, optic atrophy, retinal degeneration, dementia, extrapyramidal signs, deafness, amyotrophy and bulbar signs.<sup>1</sup> Baraitser<sup>2</sup> described a family with a syndrome of