nor structural cause was found. The presence of biological markers of lead intoxication and the favourable response to chelation therapy allowed the diagnosis of acute lead encephalopathy. The search for recent occupational and environmental exposure was negative. The normal blood lead in the patient’s wife rules out some other environmental source which might be unnoticed. Therefore, the only apparent lead exposure was his past work. The previously reported cases of lead encephalopathy occurred shortly after recent lead exposure except for patients with retained bullets. On the other hand, it is known that lead can be stored in bone for decades and mobilised by an intermittent stress with increased bone turnover. The gastroenteritic process and bed rest in this patient could have been the conditions which caused lead mobilisation.

To our knowledge, this is the first case reported of acute encephalopathy as a likely delayed presentation of occupational lead exposure. This observation emphasises the possibility of severe late toxicity as the first manifestation of a remote lead exposure.

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It is unlikely that this patient suffers from Alzheimer’s disease. The presence of progressive and selective language disorder in the absence of other cognitive deficits is distinct from the characteristic pattern of Alzheimer dementia, in which linguistic impairment is combined with profound amnesia and visuo-spatial impairment. Moreover, the effortful, paraphasic speech is dissimilar to the fluent speech normally associated with Alzheimer’s disease. The normal electroencephalogram and strikingly asymmetric computed tomogram contrast with the significant slowing of wave forms on electroencephalography and either normal or generalised atrophic scan appearances reported in Alzheimer’s disease. Reduced uptake in the temporo-parietal regions in Alzheimer’s disease has consistently been demonstrated by positron emission tomography and single photon emission tomography. Whilst the left hemisphere may be predominantly affected in patients with disproportionate language disorder, abnormalities are not exclusively unilateral. In the present case, abnormalities appeared to involve virtually the entire left hemisphere, with the exception of the occipital areas, whilst right hemisphere uptake was normal.

This patient closely resembles cases of slowly progressive aphasia without generalised dementia, one of whom also developed a right hemi-paraesis. Left temporal and parietal hypometabolism found by positron emission tomography is consistent with the left hemisphere abnormality demonstrated in the present case. Although an association has previously been reported between progressive aphasia and extrapyramidal features, unlike the present case, this was in the context of more widespread cognitive changes, indicating bilateral affection. Occasional extrapyramidal disorder has occurred in combination with fronto-temporal atrophy, but most commonly with additional clinical features of motor neuron disease.

Common to forms of localised cerebral atrophy are the pathological findings of cell loss, gliosis and spongiform change and a minority of these cases have additional neuronal swellings and argyrophilic inclusions. There appears to be a spectrum of topographical distribution of pathological change. In most cases both frontal and temporal lobes are involved but purely frontal, purely temporal and predominantly subcortical presentations have been described. In other cases with clinical features of motor neuron disease there has been additional involvement of the corticospinal tracts and anterior horn cells. Cases of progressive aphasia without dementia have demonstrated pathological change in the dominant left perisylvian region. The case described in this letter is an example of a predominantly left hemisphere degeneration presumably involving primarily the perisylvian regions and subcortex.

It is suggested that the clinical picture of our patient of progressive aphasia and progressive right-sided extrapyramidal signs represents one of a spectrum of possible presentations of localised cerebral atrophy all of which may share a similar pathological basis.

References


Fig On all images the left hemisphere is on the right. (a) SPET image from patient with progressive aphasia, taken at the level shown on the lateral view. There is a striking reduction of uptake in the left cerebral hemisphere. (b) Computed tomogram in patient with progressive aphasia. There is marked dilatation of the left lateral ventricle and enlargement of the left sylvian fissure.
Severe aggravation of blepharospasm in Fisher’s syndrome

Sir: Essential blepharospasm has been considered a cranial dystonia caused by a biochemical imbalance of the extrapyramidal system1 with no relevant peripheral nervous system contribution.2,3 Fisher syndrome is characterised by external ophthalmoplegia, ataxia and areflexia and represents a limited form of acute idiopathic polyneuropathy (Guillain-Barré) syndrome. Both processes coincided in a 67 year old male. At 20 years of age he began to suffer occasional involuntarily lid closure, more prominent in the right eye, but without disability. These spasms had increased slightly in recent years, often triggered by bright light. After an episode of acute febrile rhinopharyngitis 15 days earlier, over a week he developed severe progressive ataxia, complete external and internal ophthalmoplegia, the eyes remaining in neutral position, ptotic but without diplopia, and general areflexia. Consciousness was normal. CSF showed 0 cells, glucose 0.68 g/l and protein 0.63 g/l. In the next few days, transitory breathing and swallowing difficulties developed, as well as mild weakness of the facial musculature. In this situation of complete oral paralysis, the patient made constant gesticulation due to frequent, occasionally sustained, bilateral blepharospasm attacks. This picture regressed to the previous situation after the ophthalmoplegia resolved some months later.

The severe aggravation of facial spasms in our case was striking, well in excess of what could have been expected from the emotional stress of hospitalisation or appearance of new symptoms. A coincidental relation to an improbable midbrain lesion is purely speculative. A lesion located in the midbrain tegmentum was discovered in only one case of Fisher’s syndrome and had not been confirmed in other necropsy cases. On the other hand, in only one case of blepharospasm was a well-localised upper brain stem lesion found.4 In our patient, the futility of efforts to counter ocular paralysis and palpebral weakness (m. elevator palpebrae) may have accentuated the actions of antagonist muscles (m. orbicularis oculi). The excess of frustrated central excitation and lack of reciprocal inhibition is considered the EMG pattern of dystonia5 and could explain our case.

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Orthostatic tremor: diagnostic entity or variant of essential tremor?

Sir: Heimlan1 described three patients with orthostatic tremor, that is, tremor of the legs and trunk which commenced shortly after standing but disappeared when walking. Heiman considered orthostatic tremor to be a distinct neurological entity related to the maintenance of static posture.

Certain features distinguish orthostatic tremor from classical essential tremor. The oscillation frequency of orthostatic leg tremor has been reported to be between 14 and 18 Hz,2,3 far higher than the frequency range of classical essential tremor. Furthermore, it has been reported that propranolol, the drug of choice in essential tremor, is ineffective in orthostatic tremor.4 Clonazepam (2–4 mg/day) has been found to be beneficial in orthostatic tremor4,5 but to be of little benefit in essential tremor.6

There is, however, some overlap in the phenomenology of orthostatic tremor and essential tremor. One of Heiman’s orthostatic tremor patients had a concurrent postural tremor of the hands and a family history of essential tremor.7 Wee et al.8 described a family in which some members had a 7–8 Hz tremor of the hands which was responsive to propranolol, and others had a 6–7 Hz tremor of the legs on standing which was responsive to clonazepam but not to beta-blockers.

We describe a patient with a diagnosis of essential tremor of the hands and legs. Leg tremor, but not hand tremor was successfully treated with clonazepam.

A 53 year old women had a 15 year history of trembling of the legs when standing or unsupported. The tremor disappeared on walking or when leaning against a support. There was no tremor at rest or when seated with the legs held outstretched against gravity. For the last 3 years a postural tremor of the hands had been also present. On examination, there were no general or neurological abnormalities other than tremor (see below). In particular there were no cerebellar or Parkinsonian signs. Her father had a 6 year history of hand tremor. Objective (accelerometric) measures showed a 7–8 Hz tremor of the distal muscles of the upper limbs when held outstretched inpronated posture. A 6–4 Hz tremor was present in the proximal muscles of the lower limbs when standing but was absent during sitting and walking. Diazepam had been tried without success. Propranolol (30 mg/day) was ineffective but higher doses were not tried. Primidone initially produced a marked