LETTERS TO THE EDITOR

"Pinocchio syndrome": a peculiar form of reflex epilepsy?

Reflex epilepsies can be defined as epilepsies in which all or a significant part of the seizures can be regularly provoked by a given triggering factor, most often a sensory stimulus. Sometimes it may be unexpected, such as, reading or decision-making.1 We report a very unusual epilepsy in which seizures occurred when the patient lied.

A 51 year old patient had recently experienced three attacks with loss of consciousness and generalised convulsions. Afterwards he was confused for a few minutes. These attacks started with sensations the patient had regularly felt several times each day, in clusters of one or two weeks for five years. They consisted of epigastric constriction, a sensation of hot flush rising from his stomach to his head, and then auditory and visual illusions, for instance sensation of hearing in echo or metamorphopsia, with intense anxiety. More than a third of the attacks occurred while the patient was lying, for business reasons. The other episodes occurred without any obvious triggering factor. An EEG showed a right anterior and temporal slowing but no epileptogenic activity. Cerebral MRI showed a meningioma, 30 mm in diameter, located on the right cavernous sinus wall and the anterior clinoid process, near the sella turcica (fig). It compressed the medial part of the right temporal lobe with a mass effect. No further attack has been reported since the introduction of carbamazepine therapy and then meningioma ablation.

The attacks described fit with the diagnosis of partial vegetative seizures and the type of epilepsy fulfills the criteria of reflex epilepsy. Lying has not previously been described as a triggering factor of seizures but, according to certain authors, in some very rare cases seizures have been regularly provoked by emotion.1,2 Emotions are said to be linked to the limbic lobe, whose excitability was probably enhanced by its compression by the meningioma. When the patient was lying he probably felt a particular emotion and this limbic stimulation may have triggered epileptic discharges in the amygdala. As in Collodi's tale, in which lies made Pinocchio's nose grow, our patient's lies became obvious for his interlocutors. As he was an "eurocrat", using lies to evoke the truth in adversarial EEC negotiations, it is thanks to carbamazepine that he has been able to work again.

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Acute non-lymphoblastic leukaemia presenting as a Guillain-Barré syndrome

Peripheral nerve complications of acute leukaemia are uncommon. The neuropathy in these cases is frequently related to haemorrhage, into a nerve infarction or leukaemic infiltration.2 Symmetrical periphereral neuropathy in acute leukaemia is much less common and other pathogenic factors could be involved.2 We report a very unusual case of acute non-lymphoblastic leukaemia presented initially as Guillain-Barré syndrome.

This 69 year old man developed symmetrical parasthesiae and painful dysesthesiae of acute onset in the distal lower limbs. In the following days these symptoms increased in severity and he developed generalised progressive weakness, with greater involvement in the legs than in the arms. Four weeks after the appearance of the symptoms he went to a local hospital because he was unable to walk. Routine blood tests revealed severe abnormalities and the patient was immediately admitted to the haematological unit of a central hospital. Admission electrocardiogram showed atrial fibrillation, complete heart block, and abnormal T waves. The severity of the neurological disorder necessitated intensive care and he was transferred to the intensive care unit. Immediate investigations showed a marked leucocytosis with a differential count of 70% neutrophils and 30% monocytes.

Additional investigations included a raised alkaline phosphatase level, anorexia, and nausea. A lumbar puncture was performed and showed a pleocytosis of 143 cells/ml and a mildly raised protein level. The CSF morphology and electrophysiological studies were normal. The CSF protein was mildly raised at 38 4 mg/dl. The patient was diagnosed as having Guillain-Barré syndrome.

Figure T1 weighted MRI scan after gadolinium injection, showing the meningioma and its effect on temporal lobe.

On admission, he was unable to walk without assistance. Two months later, deep tendon reflexes were present though slightly depressed in the legs. Ten months after the onset of the symptoms, the neurological examination was normal and the patient now leads a normal social life and only complains of slight sporadic parasthesia in his left hand. He was able to walk without assistance. Two months later, deep tendon reflexes were present though slightly depressed in the legs. Ten months after the onset of the symptoms, the neurological examination was normal and the patient now leads a normal social life and only complains of slight sporadic parasthesia in his left hand. He was able to walk without assistance. Two months later, deep tendon reflexes were present though slightly depressed in the legs. Ten months after the onset of the symptoms, the neurological examination was normal and the patient now leads a normal social life and only complains of slight sporadic parasthesia in his left hand.

This patient had a neurological illness that fulfilled the criteria for the diagnosis of Guillain-Barré syndrome (GBS):2 he developed progressive areflexia, with weakness of all four limbs, and had characteristic CSF and electrophysiological features. The natural course of his neurological illness, with the phases of progression (four weeks), plateau (six weeks), and improvement (40 weeks) was also typical of GBS.3 Cancer patients with GBS differ significantly from non-cancer patients by a more
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protected disease course and a failure to improve. 3 Acute inflammatory demyelinating neuropathy in association with acute leukaemia has been reported in only a few instances. 1–3 However, in contrast to our patient, GBS in these cases was rapid progresssive and the patients died in a few weeks without haematological remission. 1–3 This case is remarkable because the associated neurological abnormalities apparently not adversely affect the therapeutic response and the prognosis of the myeloproliferative disorder. The reasons for this difference are not clear. The neuropathy in these cases is usually attributed to leukaemic infiltrations. 1–3 However, remote effects probably related to transient immunosuppression in acute leukaemia could serve as one contributory factor. 4,5

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Polyunsaturated fatty acids in peroxisomal disorders: a hypothesis and a proposal for treatment

Defects of peroxisomal β-oxidation of fatty acids are characteristic of a group of congenital disorders characterized with severe disturbances of brain function, or of the spinal cord, such as the cerebro-hepato-renal syndrome of Zellweger, infantile Refsum’s disease, neonatal adrenoleukodystrophy, X-linked adrenoleukodystrophy and adrenoleukodystrophy, hyperpipecolic acidemia, isolated absence of an enzyme of the β-oxidation sequence, and several variants described clinically as “Zellweger-like” and “neonatal adrenoleukodystrophy-like.” In all these syndromes very long chain fatty acids (C24–C26) accumulate in plasma. Medical treatment at present succeeds in normalizing their level, by suppressing endogenous synthesis by glycerol trioleate and glycerol trieruciate administration; but it is doubtful whether this leads to clinical improvement.

It has been shown recently that brain, retina, liver, kidney and erythrocytes of patients with several peroxisomal disorders are depleted of docosahexaenoic acid C 22:6(n-3) (DHA), a component which normally rises in the brain and retina during prenatal and postnatal development. 6,7 Deficiency of DHA presumably mimics the musk monkey as well as in children causes disturbances of photoelectric function (ERG, visual acuity) and peripheral neuropathy. 8,9 Martinez2 orally administered 250 mg of DHA ethyl ester daily for 10 months to a child with an NALD-like syndrome. DHA erythrocyte levels normalised but in addition there was improvement of psychomotor development and visually recorded daily movements.

We propose that the relationship between peroxisomal β-oxidation and DHA levels is not fortuitous, but is explained by the results of Voss et al.2 These authors have shown that in the rat the ratio of C22:6(n-3) proceeds by β-oxidation of C 24:6(n-3). Obviously, oxidation of C 24 is impaired in patients with a peroxisomal β-oxidation defect, and this must result in shortage of DHA. The existence of the latter mechanism is confirmed by most recent, and unexpected, data from patients with schizophrenia treated with high doses of pheno- thiazines. These drugs are in vivo inhibitors of peroxisomal β-oxidation. The patients’ thymocytes became deficient in PUFA, especially arachidonic acid and DHA; while the ratio C 26:0/22:0 increased.

We propose therefore that patients with an impaired peroxisomal function should be assayed for PUFA levels in erythrocytes or platelets, and in biopsied liver. Brain and spinal cord will be still more informative, as their content is not essentially reflected in blood cells. Data on X-ALD and AMN, which form the largest group of peroxisomal diseases, are still fragmentary. Although erythrocyte levels were normal in several patients, Martinez and others have found DHA in the brain of a single case to be below those of extreme nutritional deprivation; the brain of a patient with adrenoleukodystrophy had normal PUFA; b) patients with a lowered DHA should be treated by DHA supple- mentation. Such treatments have been initiated by Dr M Martinez, under a European project ‘Pathogenesis, prevention and treatment of peroxinsomal leukaemias’, in which more than 30 centres in 13 countries take part. For a correct evaluation it is necessary that objective tests (MRI, neuro- physiological and other tests) be carried out before treatment starts. There was a marked evolution of the PUFA level should be monitored.

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Hyperspetic encephalitis in a patient with complex partial epilepsy: confirmation by the polymerase chain reaction with necropys studies

Human herpesvirus 1 (HSV) is a relatively common and eminently treatable cause of encephalitis. Despite its moderately distinctive clinical presentation,1 the diagnosis of herpes simplex encephalitis (HSE) is unreliable,2 and until recently laboratory diagnosis of HSE has been unsatisfactory: isolation of the virus from CSF is rarely achieved; antiviral therapy has proved problematic, diagnosis by serology or immunoblotting must rely on the evolving immune response and is therefore delayed and indirect, and brain biopsy is highly invasive. The polymerase chain reaction (PCR)3 is a method for the amplification of DNA with exquisite sensitivity. In cases of suspected HSE it holds out the promise of early and accurate diagnosis.

We describe a case in which the eventual diagnosis of HSE by PCR was confirmed at necropsy. It illustrates the potential difficulty of making this diagnosis in a patient with pre-existing epilepsy and also exemplifies the occasional occurrence of a false negative result from PCR. This implies that negative results should be assessed critically and that repeated sampling may be worthwhile in difficult cases.

A 73 year old woman was admitted “drowsy but responsive” to her District General Hospital in 1991 following “multiple fits”. In 1982 she had been investigated because of a five year history of depersonalisation, micropsia, formed visual hallucinations and, rarely, loss of consciousness. Successive EEGs revealed only moderate bitemporal abnormalities, but a CT scan demonstrated a pitrus petrous meningioma with associated intracerebral oedema. The possibility of surgery was considered but rejected. Her epilepsy was controlled by anticonvulsants, and she became prone to periods of prolonged postictal confusion. On admission in 1991 she was drowsy but afebrile and there were no focal neurological signs. Her level of alertness improved over the following few days, although she remained confused, beligerent and possibly dysphasic. Five days after admission she had a flurry of complex partial seizures, became acutely confused and was treated with intravenous antibiot- ics. Three days later she was transferred to our care. On arrival she was cyanosed, febrile at 40°C, and deeply unconscious. She was severely hypoxic and was ventilated. A chest radiograph suggested left basal consolidation. An EEG showed rhythm...

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