protacted disease course and a failure to improve.[5] 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 progressive and the patients died in a few weeks without haematological remission.[1-3] This case is remarkable because the associated neurological syndrome apparently did 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 contributing factor in triggering Guillain-Barré syndrome in some patients.[3] The findings described in our case tend to support this view.

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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 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 adrenomyeloneuropathy, hyperpipecolic acidaemia, isolated absence of an enzyme of the β-oxidation sequence, and several variants described clinically as "Zellweger-like" and "neonatal adrenoleukodystrophy-like".[1] In all these syndromes very long chain fatty acids (C-24, C-26) accumulate in plasma. Medical treatment at present succeeds in normalising their level, by suppressing endogenous synthesis by glycerol trioleate and glycerol trierucate 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 22:6ω3 (DHA), a component which normally rises in the brain and retina during prenatal and postnatal development.[2] Depletion of DHA in the rhesus monkey as well as in children causes disturbances of photoreceptor function (ERG, visual acuity) and peripheral neuropathy.[3,4] Martinez[5] orally administered 250 mg of DHA ethylester daily for three months to a child with an NALD-like syndrome. DHA erythrocyte levels normalised but in addition there was improvement of psychomotor development and visually evoked potentials.

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.*[6] These authors have shown that in the rat synthesis of C 22:6ω3 proceeds by β-oxidation of C 24:6ω3. 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 phenothiazines.[7] These drugs are in vivo inhibitors of peroxisomal β-oxidation. The patients' thrombocytes became deficient in PUFA, especially arachidonic acid and DHA; while the ratio C 26:0/22:0 increased.[7]

We propose that: a) all 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 necessarily 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[2] found the DHA in the brain of a single case to be below those of extreme nutritional deprivation; the brain of a patient with adrenomyeloneuropathy had normal PUFA; b) patients with a lowered DHA should be treated by DHA supplementation. Such treatments have been initiated by Dr M Martinez, under a European project "Pathogenesis, prevention and treatment of peroxisomal leukodystrophy", in which more than 30 centres in 13 countries take part. For a correct evaluation it is necessary that objective tests (MRI, neurophysiological and other tests) be carried out before treatment starts. The evolution of the PUFA level should be monitored.

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Herpes simplex encephalitis in a patient with complex partial epilepsy: confirmation by the polymerase chain reaction with necropsy studies

Human herpes simplex virus 1 (HSV) is a relatively common and eminently treatable cause of encephalitis. Despite its moderately distinctive clinical presentation,[1] the clinical 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; antigen detection 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 episodic depresonalisation, micropsia, formed visual hallucinations and, rarely, loss of consciousness. Successive EEGs revealed only moderate bitemporal abnormalities, but a CT scan demonstrated a right petrous meningioma with associated intracerebral oedema. The possibility of surgery was considered but rejected. Her epilepsy was controlled by anticonvulsant therapy although she became prone to periods of prolonged postical 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, belligerent and possibly dysphasic. Five days after admission she had a flurry of complex partial seizures, became drowsy and febrile, and was treated with intravenous antibiotics. 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 rhyth-