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. 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 epilepsies in which 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. 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 "ecurocrat", using lies to evoke the truth in adversarial EEC negotiations, it is thanks to carbamazepine that he has been able to work again.

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.1 Symptomatic peripheral 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 paresthesiae and painful dysesthesiae of the 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 centre. Admission to the unit was alert, well oriented and pale, and febrile. Blood pressure was 130/60, pulse rate 72/min and respiratory rate 19/min. Purpura, lymphadenopathy and hepatosplenomegaly were absent. There was generalised flaccid motor weakness of grade III/IV (MRC) in the upper limbs and of grade I/IV (MRC) in the lower limbs. Deep tendon reflexes were absent or very much diminished in the arms and were absent in the legs. Pinprick sensation was symmetrically impaired in the lower limbs in a stocking distribution and was preserved in the arms. Joint position and vibratory sensations were impaired in the legs and preserved in the arms. Signs of meningeal irritation, cor-ticospinal tracts, cranial nerves and autonomic involvement were absent. Higher mental function remained normal. The haemoglobin was 6.7 g/dl without reticulocytes. Leucocyte count was 49 000/mm³ with 6% polymorphonuclear neutrophils, 5% lymphocytes, 5% monocytes and 84% blast cells. The platelet count was 81 000/mm³. Bone marrow aspirate specimens showed increased cellularity due to infiltration (67%) by blasts, Peroxidase + and of promyelocytes. The haematological diagnosis was classified as acute non-lymphoblastic leukemia (FAB classification M2). Chemotherapy was immediately started and included daunorubicin 40 mg/m² intravenously for three days and cytosine arabinoside 200 mg/m² in continuous perfusion for seven days. A remission of the haematological measures was obtained with one therapeutic cycle, after a 22 day period of aplasia complicated with infection and gastrointestinal hemorrhage. Consolidation of the haematological remission was carried out 38 days after the induction, according to a similar protocol. As remission therapy, cycles of cytosine arabinoside associated with VP16, 6-thioguanine and daunorubicin have been performed each month to date.

Considering the severity of the haematological condition on admission and the emergency chemotherapy, lumbar puncture was delayed until nine weeks after the onset of the symptoms. CSF was clear and colourless, with normal opened pressure, cell count (one lymphocyte) and protein concentration. A pure transudate was found with a protein content of 132 mg/dl (normal <50 mg/dl) and an albumin quotient (CSF/serum albumin ×10) of 38.2 (normal <9). Oligoclonal gamma globulins in serum and CSF protein electrophoresis were not observed. There was no evidence of intrathecal immunoglobulin synthesis. Electrophysiological studies were first performed eight weeks after the onset of symptoms. Motor nerve conduction of median nerve was 38.4 ms (normal >48 ms) with a distal motor latency of 4.1 ms (normal, up to 4.2). Distal sensory action potential was absent. Motor conduction velocity of peroneal nerve was 25 ms (normal >40 ms) with a distal motor latency of 8.7 ms (normal, up to six). Electromyography of the abductor pollicis brevis, tibialis anterior and extensor digitorum brevis muscles revealed a pattern of discrete activity with an increase of mean duration of individual motor unit potentials. Fibrillation potentials and positive waves were present in these muscles. A sural nerve biopsy showed a mild perivascular mononuclear cell reaction in the endoneurium, and marked demyelination in several foci of nerve fascicles was observed. However, there was no axonal degeneration. Leukemia cell infiltration or areas of necrosis were not present. A progressive improvement of the neurological picture was apparent nine weeks after the onset of symptoms. Thirteen weeks later, 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 paresis in his legs.

This patient had a neurological illness that fulfilled the criteria for the diagnosis of Guillain-Barré syndrome (GBS): he developed progressive areflexia and 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), improvement (40 weeks) was also typical of GBS. Cancer patients with GBS differ significantly from non-cancer patients by a more