Letters

Long-term survival of a case of subacute sclerosing panencephalitis

Sir: Subacute sclerosing panencephalitis (SSPE) carries a grave prognosis, death usually occurring within less than two years of the onset of the disease. I report a patient who has not only survived for nine years to date, but who has also shown spontaneous clinical, electrophysiological and immunological improvement sustained over the past five years.

A 15-year-old schoolboy was referred to the Midland Centre for Neurosurgery and Neurology in May 1975, having had a tonic-clonic seizure. A similar seizure had occurred two years earlier, and since that first episode his parents had witnessed several typical absence attacks, and a gradual deterioration in his concentration, short-term memory and school performance. His birth history and developmental milestones had been normal. He had suffered measles when 15 months old. On examination his responses were slow, but the only formal sign was a right upper motor neuron facial weakness. An electroencephalogram (EEG) showed repetitive generalised complex discharges every four seconds (fig.). Serum measles antibody titre by complement fixation was 1:1024 on three occasions, and the spinal fluid titre was 1:128. The protein content of the spinal fluid was normal at 0.52 g/l, with no excess of cells. A right parietal brain biopsy showed gliosis of white matter with hypertrophied astrocytes and scattered small "glial stars" with activated microglia at the centre. In addition, occasional single or grouped round cells appeared in perivascular spaces, but only a single vessel showed classical cuffing. The cortex showed some neuronal loss and satellitosis of other neurons. No intranuclear inclusions were seen, but the absence of these is not unusual in the sclerotic phase of SSPE, and the histological findings described above are otherwise typical of that condition.

Eight injections of transfer factor were given over the following three months, but clinical deterioration continued. Four months after presentation he was dyslexic, disoriented in time, and had a mixed expressive and receptive dysphasia, with a spastic quadriaparesis. Myoclonic jerks of the right limbs had developed, and were synchronous with the EEG complexes. Spinal fluid protein was now elevated at 0.80 g/l, electrophoresis showing a gamma globulin content of 24.3% of the total, and three abnormal bands. Measles antibody titre in the spinal fluid had risen to 1:1024. His condition continued to deteriorate so that two years after presentation he was unable to communicate, walk, feed or dress himself, was incontinent of urine and faeces, and had bilateral myoclonic jerks every four seconds. Since then, over the past five years, no further deterioration has occurred, and there have been signs of clinical improvement. He has become continent of urine and faeces during the day, and walks with minimal support. He helps with dressing, and obeys simple commands, but remains aphasic, with a spastic quadriaparesis. Myoclonic jerks are now rarely seen, and repetitive complexes are absent from the EEG. Serum measles antibody titre has fallen to 1:32, but it has not been possible to obtain parental consent for a repeat examination of the spinal fluid.

The patient's clinical presentation and course over the following two years are typical of SSPE. This diagnosis is supported by the increased spinal fluid gamma globulin, the typical EEG, the findings at brain biopsy and the high serum and rising spinal fluid measles antibody titres. The disease usually progresses relentlessly, death frequently occurring within less than two years. It has been suggested that 10–20% of patients live for more than four years, and it may be that the disease can remit for several years before relapsing and progressing to death.

There are several reports of spontaneous remission with apparent clinical improvement and even recovery, but some are difficult to assess because of incomplete laboratory evidence that the disease was truly SSPE rather than some other form of encephalitis. The case described by Kurtzke, in which there was incomplete recovery after four months, had the unusual feature of an acute onset of pleocytic cerebrospinal fluid, and the EEG findings are not recorded. A patient with complete recovery after seven months has been reported by Simpson, but the EEG was atypical, and cerebral biopsy not performed. Pearce and Barwick recorded a patient with a clinical history compatible with SSPE, and EEG findings which were highly suggestive of that disorder, who recovered completely nine months after the onset of symptoms. No cerebral biopsy had been performed, and the authors admit that the illness could have been another variety of subacute encephalitis. Two patients reported by Cobb and Morgan-Hughes had a slowly progressive illness which was almost certainly SSPE; one deteriorated for 15 months, and the other for two years, following which a clinical improvement occurred, and had lasted eight years and two years respectively by the time of their publication. Resnick et al...
record a proven case of SSPE in a 15-year-old boy who deteriorated for 20 months and then made an impressive spontaneous improvement which had lasted for 3½ years at the time of their report. Vandvik et al. suggested treatment with transfer factor, but the results have not been encouraging. The patient reported here received this treatment but continued to deteriorate for 21 months afterwards, and it seems unlikely to have contributed to the long-term survival. There have been reports of remission occurring following treatment with isoprinosine and with amantadine. Huttenlocker et al. reported sustained remission with clinical improvement for up to six years in six patients treated with isoprinosine, and Robertson et al. reported remission and mild improvement for one to seven years in four out of eight patients treated with amantadine.

There is no doubt that this patient will remain permanently severely disabled. However, the mild but sustained clinical improvement over the past five years, the disappearance of generalised repetitive complexes from the EEG, and the dramatic fall in serum measles antibody titre all suggest that the disease is not just in remission, but that the pathological process has ceased. The possibility of this event occurring spontaneously must be borne in mind when assessing the therapeutic effects of any new treatment for this disorder.

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References

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Thrombosis of cerebral venous sinuses due to a catheter for parenteral nutrition

SIR: Venous thrombosis in patients with an indwelling venous catheter is a well known iatrogenic condition. We report a case of cerebral venous sinuses thrombosis in a patient receiving parenteral nutrition.

In March 1979, after a subtotal intestinal resection for mesenteric infarction, a 41-year-old patient was fed by chronic parenteral nutrition using a silicone catheter implanted into the right external jugular vein and entering into the superior vena cava; the other end of the catheter was inserted under the skin along a distance of 10 cm. In October, the catheter was removed after inflammatory signs had appeared suggesting a right external jugular vein thrombosis. The patient successfully underwent treatment for eleven days with heparin and ampicillin. Intra-venous heparin subsequently was replaced by sub-cutaneous calcium heparinate (5000 units every 12 hours). On November 3rd, the patient was re-admitted to the emergency ward with violent headache, behavioural changes and progressive somnolence. He soon developed a left hemiparesis and hemic-hypoesthesia including the face. He then developed partial motor status epilepticus involving the face and the left upper limb. Intra-venous diazepam and phenytoin therapy was unsuccessful. Lumbar puncture at the admission was normal. EEG showed a slowed background rhythm with a high voltage delta rolandic focus in the right hemisphere. A right carotid arteriography showed, on the late films, the absence of opacity of the superior longitudinal sinus and of the right lateral sinus. There was no sign of collateral venous pathways. He was treated with pentothal infusion and fibrinolysis with streptokinase for 36 hours. Subsequently he recovered and a repeat angiography was normal.

Venous thrombosis may be an iatrogenic condition in patients with indwelling catheters. Subclavian vein thrombosis were reported in three patients in a series of 771 cardiac stimulator implantations1 and in one patient of another series of 298 implantations.2 Cerebral venous thrombosis is much less frequent, since it was reported in only two patients who underwent cardiac pacing.3 Chronic parenteral nutrition which requires indwelling catheters for several months may constitute a similar potential risk. To our knowledge, however, the case we report is the first instance of cerebral venous thrombosis complicating chronic parenteral nutrition.

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