Letters

do not have a satisfactory explanation for our patient's left cerebral hemisphere dysfunction. In the final stage of the disease, the clinical picture was dominated by radicular pain in the legs and faciocranial paraparesis, also indicating spinal subarachnoid seeding. Postmortem examination not only confirmed this, but also disclosed that the primary tumour was located in the pons. The incidence of leptomeningeal dissemination of brainstem gliomas is difficult to determine since only a few series of patients have been described. Moreover, the diagnosis is often made by postmortem examination. However, Packer et al were able to diagnose meningeal gliomatosis antemortem in five of 15 brainstem glioma patients (33%).

All previously reported patients were found to have extensive spinal subarachnoid seeding. To the best of our knowledge, there have been no well-documented reports on brainstem gliomas spreading to the supratentorial meninges.

We are grateful to Professor A J M van der Werf for performing the brain biopsy.

EP VRIES*  
M DE VISser*  
D TROOST†  
The Departments of Neurology* and Pathology,†  
Academic Medical Centre, Amsterdam, The Netherlands

Address for correspondence: E P Vries, MD, Department of Neurology, Academisch Medisch Centrum, Meibergdreef 9, 1105 AZ Amsterdam, The Netherlands

References


Accepted 7 March 1989

CSF oligoclonal IgG bands in a patient with torsion dystonia

Sir: The CNS changes underlying idiopathic torsion dystonia are unknown. Immunological changes have not been recognised in this condition but we report here a case in which oligoclonal IgG bands were identified repeatedly in the cerebrospinal fluid. A 38 year old male patient was referred for investigation of his abnormal gait in 1983. His walking was disturbed by gross abnormal movements. Sustained dystonic muscular contractions provoked inturning, twisting and high stepping of the right foot with a tendency to hold the left leg stiffly. An exaggerated lordosis, twisting of the trunk, and wide swinging of the left arm accompanied these movements.

Dystonic posturing of both legs, and particularly the left arm, were exaggerated by voluntary activity. Finger-nose testing on the left was disrupted by these movements although there was no ataxia, alteration in tone, weakness or sensory loss in the limbs. Dystonic tongue movements made the patient's speech dysarthric. Visual acuities were 6/60 right and 6/6 left. There were no Kayser-Fleischer rings and cranial nerve examination was otherwise normal. Limb reflexes were present symmetrically, and both plantar responses were flexor. General physical examination was otherwise normal.

The patient had been born at 38 weeks gestation by caesarean section, birth weight 2.97 kg, his mother having had hypertension in pregnancy. He was healthy at birth but developed pertussis at 6 weeks of age. This illness required hospital admission for 10 weeks and he was said to have had a number of seizures during this period. He could walk at 15 months, and at this time was noted to have a squint affecting the right eye.

His abnormal gait was first brought to medical attention by his mother at the age of 6 years, and gradually deteriorated throughout childhood. Clinical records suggest a "...jerky, clumsy gait, ... with normal balance, co-ordination and reflexes (6 years). ..." "...spastic muscular incoordination (7 years) ..." and "...a curious gait in running, a habit (8 years). Examination at 8 years was recorded as showing no evidence of organic disorder or spasticity. The condition progressed gradually to abnormal posturing of the legs and left arm at rest. Dystonic movements of the tongue, jaw and neck were noted when the patient was seen with back pain aged 36 years.

The patient's family were non-Jewish and there was no family history of any abnormal movement disorder. The patient did not smoke, drank alcohol only occasionally, and was taking no drugs when seen. He had never received neuroleptic or other drugs associated with abnormal involuntary movements.

Investigations showed a normal full blood count and film, urea and electrolyte estimation, plasma glucose, B12, folate, thyroid and liver function tests. Serum copper 16.2 μmol/l, (laboratory normal range 13-24 μmol/l), and caeruloplasmin 190 mg/l, (laboratory normal range 150-600 mg/l). Serum antinuclear factor, auto-antibody screen, VDRL, and urine amino acid chromatography were negative. Peripheral motor and sensory nerve conduction studies, somatosensory and visual evoked potentials, chest radiograph, CT and magnetic resonance imaging of the brain were normal. The presence of Kayser-Fleischer rings was excluded by slit lamp examination.

Cerebrospinal fluid (CSF) examination in 1983 showed a normal pressure and glucose level, less than 1 white cell/mm3, protein 0.650 g/l, IgG 0.089 g/l, IgG index 1.36 and multiple oligoclonal bands detected by isoelectric focusing. Repeat CSF examination in 1987 showed CSF protein 0.544 g/l, albumin 0.237 g/l, IgG 0.108 g/l, IgG index 1.78, oligoclonal bands strongly positive in

Fig Immunoblot showing multiple discrete IgG bands in unconcentrated CSF (C) but not serum (S).

Accepted 7 March 1989
CSF and absent from serum. (Serum protein 73 g/l, albumin 45 g/l, IgG 11·5 g/l). Oligoclonal bands were detected by a modification of the method of Walker. In brief, unconcentrated CSF and serum diluted 1:100 in 0·9% saline (to produce an IgG concentration equivalent to that in the CSF) were resolved by isoelectric focusing on agarose, followed by blotting on to a newly introduced support, polyvinylidendifluoride (PVDF; Millipore). Staining was by a double-antibody immunoperoxidase technique specific for IgG.

Follow up over a 5 year period has shown a slow progression of dystonic movements involving all four limbs and the tongue with dystarthis. During this period chlor Diazepoxide, clonazepam, dysarthria. During levodopa/carbidopa, have been taken in this patient with idiopathic dystonia. The presence of oligoclonal IgG bands in the CSF of patients with idiopathic dystonia has not been reported previously. Kjellin and Stibler found increased cathodal protein fractions on isoelectric focusing of CSF from patients with spasmotic torticollis although non-specific staining techniques did not allow identification of the exact nature of these abnormal proteins. In the present case protein bands found on CSF isoelectric focusing were specifically identified as IgG by an immunochemical method. Oligoclonal IgG bands present exclusively in the CSF suggest antibody production within the central nervous system and the elevation of the IgG index in our case is consistent with this. Identification of CSF oligoclonal IgG bands has proved useful in the diagnosis of multiple sclerosis and they are present in various other disorders, especially CNS infections and autoimmune conditions. Identification of oligoclonal IgG in the CSF of patients with encephalitis lathrigica has been taken as supportive evidence for a viral aetiology in this condition. Symptomatic torsion dystonias have been reported in association with a wide variety of hereditary and degenerative neurological syndromes as well as vascular, traumatic, and toxic brain disorders. The significance of oligoclonal CSF IgG bands in this case is uncertain but if confirmed in other patients would suggest either immune or virally mediated CNS damage as the pathophysiological mechanism in at least some cases of torsion dystonia.

References


Accepted 22 March 1988

Tarsal tunnel syndrome secondary to intraneural ganglion

SIR: A 61 year old healthy man complained of burning pains and paraesthesiae, which had been present for 7 months in the toes and along the sole of his left foot. Discomfort was worse at night, and sometimes radiated proximally along the medial aspect of the calf. There was no history of trauma about the ankle. On examination, there was a decrease in two-point discrimination and hypoaesthesia to pin-prick in the distribution of the medial and lateral plantar nerves. There was no obvious wasting or weakness of the small foot muscles. Tinel’s sign was positive in the posterior tibial nerve at the ankle. No mass was palpable along the medial aspect of the ankle. General neurological examination was normal. Radiographs of the left ankle and foot were normal. Electrophysiological tests were performed bilaterally.

EMG studies did not show any spontaneous activity in the small foot muscles. Distal motor latency (ankle to abductor hallucis) was 7·28 ms on the left and 4·21 ms on the right (normal values: 2·7–5·13 ms) with a small compound muscle action potential on the left. The left medial plantar sensory action potential (SAP) was absent; the right SAP was 0·7 μV of amplitude (normal values: 0·1–3·5 μV), with a sensory conduction velocity of 42 m/s (normal values: 36–50 m/s). The patient underwent surgery. A curved incision was made superior, posterior, and inferior to the medial malleolus. The neurovascular bundle was isolated just proximal to the tarsal tunnel, which was entered by dividing the flexor retinaculum. A calcaneal branch, and the medial and lateral plantar nerves were dissected free. Within the tarsal tunnel, the posterior tibial nerve showed an eccentric, translucent, rubbery, bean-sized swelling. The epineurium was incised longitudinally, and clear gelatinous material evacuated (fig). As the swelling collapsed, a small fusiform cavity, about 2 cm long, with a firm, whitish wall was found within the nerve. Nerve bundles could not be dissected away from the cyst wall. No pedicle was found connecting the cyst to nearby tendon sheaths or joints. The cavity was gently irrigated. A fragment of the cyst wall was taken for biopsy. Microscopically it was composed of lamellar connective tissue with scanty cells. The cyst contained mucoid material.

Symptoms and signs rapidly disappeared following surgery. When seen two years later, the patient did not complain of any symptom, and the neurological examination did not show any abnormality.

Cystic intraneural tumours filled with gelatinous material and referred to as ‘ganglia’ are rare. Their pathogenesis and pathology have been discussed by Sherman et al.
CSF oligoclonal IgG bands in a patient with torsion dystonia.

S J Wroe, P D Gills and R E Cull

J Neurol Neurosurg Psychiatry 1989 52: 1013-1014
doi: 10.1136/jnnp.52.8.1013

Updated information and services can be found at: http://jnnp.bmj.com/content/52/8/1013.citation

These include:
Email alerting service
Receive free email alerts when new articles cite this article. Sign up in the box at the top right corner of the online article.

Notes

To request permissions go to: http://group.bmj.com/group/rights-licensing/permissions

To order reprints go to: http://journals.bmj.com/cgi/reprintform

To subscribe to BMJ go to: http://group.bmj.com/subscribe/