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.\(^1\) 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, polyvinylidifluoride (PVDF; Millipore).\(^2\) 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 dysarthria. During this period chlorazepoxide, clonazepam, benzhexol, tetrabenazine, pimozide, sodium valproate, carbamazepine, bromocriptine, and levodopa/carbidopa, have all been tried without definite benefit and the patient currently takes no medication.

This patient exhibits a torsion dystonia in which \(\ldots\) sustained and forceful muscle contractions twist the body and limbs into characteristic postures \(\ldots\).\(^3\) Onset in childhood, progression from involvement of legs to arms and increasing disability with walking identify this as the syndrome termed dystonia musculorum deformans by Oppenheim and idiopathic generalised dystonia by later authors.\(^4\) Absence of ocular dysmobility, spasticity, ataxia, identifiable biochemical abnormalities or structural brain defects makes a diagnosis of secondary dystonia unlikely.

The presence of oligoclonal IgG bands in the CSF of patients with idiopathic torsion dystonia has not been reported previously. Kjellin and Stibler found increased cathodal protein fractions on isoelectric focusing of CSF from patients with spasmocodic torticollis although non-specific staining techniques did not allow identification of the exact nature of these abnormal proteins.\(^5\) 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.\(^6\) Identification of oligoclonal IgG in the CSF of patients with encephalitis lethargica has been taken as supportive evidence for a viral aetiology in this condition.\(^7\) 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.\(^8\) 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

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Tarsal tunnel syndrome secondary to intra-neural 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 hypoesthesia 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: 2·7-5·1 ms) with a small compound muscle action potential on the left. The left medial plantar sensory action potential (SAP) was absent; the right medial SAP was 0·7 μV of amplitude (normal values: 0·1-3·5 μV),\(^1\) with a sensory conduction velocity of 42 m/s (normal values: 56-90 m/s).\(^1\) 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 1·5 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 intraaneurmal tumours filled with gelatinous material and referred to as "ganglia" are rare. Their pathogenesis and pathology have been discussed by Sherman et al.\(^8\)
Neopterin: bioppterin ratios in Down's syndrome

Sir: Urinary neopterin:bioppterin ratios are elevated in senile dementia of the Alzheimer type (SDAT) compared with age matched controls, due to a reduced conversion of dihydronorbioppterin triphosphate to tetrahydrobioppterin. There is a similar elevated neopterin:bioppterin ratio in Down's syndrome, a condition producing a similar dementia to SDAT.

Morning urines were collected into ascorbic acid (2% final concentration) from a group of 53 Down's sufferers of mixed sex aged between 1 to 70 years without any other disease and, following storage at −20°C, the neopterin and bioppterin levels were determined by HPLC after acid iodine oxidation. Compared with a group of 35 healthy controls of mixed sex and aged 23 to 93 years, the neopterin:bioppterin ratio was significantly elevated 3:44, 1.73 vs 1:12, 0.55 (mean, SD) p < 0.2% (Table). Against a creatinine baseline bioppterin levels were unaffected but the urinary neopterin levels were significantly increased (Table). No age trend was observed suggesting that these metabolic effects precode dementia onset. The elevated neopterin:bioppterin ratio could be due to a disease related immune response causing stimulation of neopterin biosynthesis, although all disease states known to produce such were absent and the rise is less pronounced that in such conditions. It is known that there are disturbances in the immune system of Down's patients, and it could be suggested that these disturbances are causing the rise in urinary neopterin in a similar manner to viral and malignant disorders. However, it has been found that interferon gamma (IFNg) production is lower in Down's patients than in normal patients with infection. It is IFNg which stimulates the macrophages to produce neopterin in infectious disorders, so any decrease in IFNg levels should result in similar or reduced levels of urinary neopterin and not the increased levels observed. A second possibility is the presence of greater levels of guanosine triphosphate in the urine pool from an increased biosynthesis of purines as a result of the extra chromosome 21, which has the locus for some of the enzymes for purine biosynthesis.

The levels of neopterin and bioppterin are measured against a creatinine baseline. In the Downs subjects creatinine clearance is reportedly lower, reflecting a reduced glomerular filtration rate, and this could be causing a distortion in the results. However, creatinine clearance has a positive linear correlation with both neopterin and bioppterin so any

References
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