Benign recurrent multiple mononeuropathy in Wegener's granulomatosis

Sir: Neurological involvement in Wegener’s granulomatosis is common, occurring in 26% of patients in a review of 374 cases. In the more localised form, midline granuloma, 12% of 125 patients had neurological involvement but only one had a peripheral neuropathy. We wish to report a patient with a 5 year history of recurrent multiple mononeuropathy prior to histological diagnosis of Wegener’s granulomatosis.

A 62 year old woman presented in March 1983 with a two month history of severe right frontal and temporal headache. Sinus radiographs were consistent with sinustitis. She did not improve with antibiotics and ENT washout in the University ENT Department was normal; she was referred to the neurology department and treated with prednisolone with resolution of her headache.

Four months later on prednisolone 20 mg per day, she developed double vision due to an almost complete right external ophthalmoplegia without ptosis. ESR was 46 mm in 1 hour; and her signs resolved after 5 days of prednisolone 100 mg per day which subsequently was gradually reduced.

In November 1983, while on prednisolone 12.5 mg per day, she developed a left vocal cord paralysis and a chest radiograph showed elevation of the left hemidiaphragm. ESR was 20 mm in 1 hour: mediastinal tomography, CT thorax and neck, bronchoscopy, sputum cytology, thyroid and bone isotope scans were all normal. The gradual reduction in prednisolone dosage was continued and her voice was much better one year later. She remained on 2.5 mg prednisolone daily until March 1986 when she developed numbness and nagging pain in the left side of her face and forehead. Sensation over the left infraorbital nerve was impaired. ESR was 40 mm in 1 hour and sinus radiographs showed an opaque left antrum. Left maxillary antral examination revealed an absent medial wall which was attributed to previous surgery; antral washings contained polymorphs only with no malignant cells. Her symptoms improved with prednisolone 15 mg per day. CT of her naso-pharynx revealed no other abnormality. In January 1987 on 7.5 mg of prednisolone she developed sudden visual impairment of the left eye with pain around the orbit. Examination revealed constricted peripheral vision in the left eye with visual acuity of N48, and a left relative afferent pupillary defect. There was full recovery after one week on 30 mg prednisolone per day, and the dose was gradually reduced.

In June 1987 because of osteoporosis the steroids were gradually reduced and discontinued six months later. In August 1988 she complained of left maxillary pain and an oro-antral fistula with erosion of all the surfaces of the maxillary antrum was found. Histology of the fistula lining showed multinucleated giant cells and necrosis surrounded by pallisading of histiocytes with necrotising vasculitis of small arteries, characteristic of Wegener’s granulomatosis. She was started on cyclophosphamide 75 mg per day and reducing doses of prednisolone, and remains well. At no stage has there been any renal or respiratory system disorder.

This woman had a 5 year history of recurrent cranial and peripheral nerve lesions including the left optic, left infraorbital, left phrenic and left recurrent laryngeal nerves before the diagnosis of Wegener’s granulomatosis was made when an oro-antral fistula developed. The initial diagnosis based on the severe headache, high ESR and response to steroids was giant cell arteritis and the subsequent external ophthalmoplegia was attributed to this disorder also. The correct diagnosis probably could have been made by biopsy of antral mucosa in March 1986. Any cranial nerve may be affected by Wegener’s granulomatosis but ocular involvement is frequent; the optic neuropathy was unusual in the rapid response to a modest dose of steroids and was presumably due to compression by contiguous inflammatory granulomas. The phrenic and recurrent laryngeal nerves were presumably affected by a vasculitis of the vasa nervorum and the external ophthalmoplegia may be explained by orbital muscle rather than third nerve involvement since ptosis and pupillary abnormality were absent.

This case report serves to emphasise that Wegener’s granulomatosis may be a rather indolent process and that, in the presence of cranial or peripheral neuropathy, symptoms of sinus disease should be fully investigated, including biopsy.

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References

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Progressive systemic sclerosis presenting as a case of trigeminal neuropathy

Sir: A 58 year old shipyard welder first noticed numbness over the right lower lip in
1981. When he was first seen in 1986, the numbness had spread to both sides of the face and inside the mouth. His speech was slurred and he constantly dribbled saliva from the corners of his mouth. To eat or drink, he had to observe himself in a shaving mirror in order to avoid loss of fluids from his mouth, and because he claimed that this manoeuvre reduced his tendency to bite his lip or tongue, which otherwise he did unknowingly. Examination revealed bilateral trigeminal sensory loss to pinprick in all three divisions, with loss of corneal sensation. His ESR was persistently raised (103, 65, 92 mm/1h), and calcification of the pleura, consistent with his previous asbestos exposure, was seen on his chest radiographs.

A thorough and successfully performed ENT examination revealed no abnormality. CT of the brain, nasopharynx and sinuses, visually evoked potentials and all routine blood tests, including autoantibodies, were normal.

By 1987 he had developed mild bilateral facial weakness, loss of taste over the whole tongue, with slight wasting and fasciculation; sensory loss in the oropharynx and mild weakness of both sternomastoids. All tests were repeated with the same results. CSF analysis revealed no abnormality. MRI demonstrated non-specific areas of abnormal high signal in the periventricular white matter and subcortically in both cerebral hemispheres. Mild denervation to the right nasolabialis was confirmed and complete absence of blink reflexes, indicating a bilateral trigeminal afferent pathway lesion. Abnormalities in VEPs, or brainstem evoked potentials were absent.

It was now clear that the patient did not have idopathic trigeminal neuropathy. Neurologically he remained stable until late 1987, when, over a few weeks he became generally unwell, with generalised limb pains and tenderness, with swelling and stiffness affecting most joints. Bilateral sensory impairment in C2 and C3 dermatomes was present and he clearly demonstrated clinical, radiological and serological evidence of systemic sclerosis. Tight sclerotic skin was present in two areas of the body, and over the hands, and radiographs revealed an erosive arthritis. An anti-nuclear antibody titre of less than 1/25 was present; a polyclonal increase in IgG of 17-9 g/l; C reactive protein of 22 mg/l; but all pulmonary function tests were within predicted limits.

Treatment with naproxen, 500 mg bd and 500 mg of penicillamine daily was commenced and later increased to 750 mg daily, when his skin condition deteriorated in 1988. However, no subjective, or objective progress of his neurological disorders could be observed. Sadly, his practical problems regarding eating and drinking remain unchanged and he still eats with the aid of a shaving mirror.

Trigeminal neuropathy is an infrequent, but important condition. The onset of facial numbness requires careful investigation and long-term follow-up, with repeated clinical observation to exclude any possible underlying condition and before describing the disorder as an isolated and thus idopathic phenomenon. Sensory loss, only within the distribution of the trigeminal nerve, should alert the physician to consider the possibility of carcinoma of either the nasopharynx,10 the cerebellar pontine angle, or the base of the skull.2 Half of such neoplasms cause neurological symptoms before the diagnosis is established, and it may be many months before evidence for the neoplasm is discovered.2,4 In some cases the cause remains obscure, although a small subgroup of patients in Harris's original description, demonstrated a relatively benign condition that resolved within weeks or years.3 Facial numbness, as an isolated symptom, has been reported in a variety of disorders such as multiple sclerosis,6 infection of the paranasal sinuses, hypertension and diabetes.5 It may also follow exposure to certain toxins such as vinyl chloride.6 Some patients may have an underlying collagen vascular disorder such as systemic lupus erythematosus,11 mixed connective tissue disease,12 or progressive systemic sclerosis.12,13

In systemic sclerosis, Raynaud's phenomenon is followed by changes of atrophy and sclerosis, associated with widespread vasculitis in the skin and internal organs.14 Neurological involvement is rare and includes trigeminal neuropathy, cranial nerve palsy,15 as well as indirect involvement from subacute degeneration of the spinal cord from B12 deficiency secondary to malabsorption when the small intestine is involved.16 Neurological deficit may also arise secondary to treatment, as in steroid myopathy, or from damage to other organs, such as seizures from uremia with renal involvement. The features may also be totally coincidental.

It is unclear as to whether the primary pathological process causing the sclerosis causes the neuropathy.17 A peripheral lesion due to a combination of micro-angiopathy and fibrosis, was proposed by Teesdale et al.13 Recently, Lecky et al suggested that if the trigeminal motor pathway is spared and there is individual involvement of the sensory divisions of the nerve and a lack of dissociation of sensory loss, then lesions in the trigeminal ganglion or in the proximal part of the main division of the nerve are more likely. The circmoral numbness and taste disturbance is consistent with such a peripheral lesion.18

Systemic sclerosis rarely affects the nervous system. However, four percent of patients develop unilateral, or bilateral facial numbness.13 Although trigeminal neuropathy may be the first feature, systemic sclerosis usually becomes obvious at, or shortly after the onset of the neuropathy.19 Farrell and Medsger9 described two patients in their series and only a further five in the literature they reviewed, whose neuropathy was the first and only feature. However, in all cases the time from neuropathy to onset of clinical systemic sclerosis was measured in months and not years. The patient reported is unusual in that the initial presentation of trigeminal neuropathy, followed later by further cranial nerve involvement, preceded the onset of clinical systemic sclerosis by more than six years and did so in the absence of Raynaud's phenomenon.

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References
vivid nocturnal auditory and visual hallucinations. She showed almost continuous and severe generalised irregular myoclonic jerking. She was unable to stand and walk. Her tendon reflexes were exaggerated. Laboratory work-up of blood and CSF yielded normal results. The serum amitriptyline concentration was within the therapeutic range. CT revealed minimal frontocortical atrophy. The first EEG showed slow alpha-activity with periodic runs of jerk-locked polyspike/sharp waves. SSEP showed normal latencies and a markedly increased cortical N20/P25 amplitude (16 μV) after median nerve stimulation. One week after admission and discontinuation of amitriptyline the EEG showed regular alpha activity. The SSEP latencies and N20/P25 amplitudes were within normal limits (6 μV). By this time no muscle jerks except mild facial twitching could be observed. The delusional syndrome resolved completely after discontinuation of amitriptyline, parallelising the normalisation of the EEG and the SSEP.

The first EEG recording seemed to confirm Creutzfeldt-Jakob disease. The SSEP was initially reminiscent of “giant potentials” indicating cortical myoclonus or myoclonus epilepsy. These diseases had to be considered because of the patient’s professional and family history. After discontinuation of amitriptyline, however, psychosis, myoclonus, EEG and SSEP changes proved to be fully reversible.

Cyclic antidepressants frequently lead to an anti-cholinergic delusional syndrome and myoclonus. Our observations may perhaps contribute to the understanding of these side-effects. Jerk-locked polyspike/sharp waves and enlarged cortical SSEP amplitudes often reflect increased cortical excitability, which could account for the epileptogenic and possibly hallucinogenic properties of cyclic antidepressants. Increased SSEP amplitudes might therefore prove reliable indicators for patients at risk to develop seizures during antidepressant therapy.

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Psychiatric disturbance in mitochondrial encephalomyopathy

Sir: Luft et al. reported a case of muscle disorder which was considered as mitochondrial myopathy because of abnormal existence of muscular mitochondria revealed by the ultrastructural, histochemical and biochemical assays. The term mitochondrial encephalomyopathy was introduced by Shapira et al. to represent various types of neuromuscular disorders which showed defects in the oxidative metabolic system for energy production in the muscular mitochondria. There have been, however, no studies to our knowledge to investigate the psychiatric disturbance of mitochondrial encephalomyopathy. This case report describes psychiatric features of mitochondrial encephalomyopathy.

The patient was a 35 year old male. His short stature had been remarkable since the age of 10 years. At the age of 19 years he complained of severe abdominal pain with vomiting. General muscle atrophy had developed gradually since then. At the age of 25 years, he had a sudden abdominal pain and showed schizophrenia-like symptoms: auditory hallucination, delusion of reference and persecution, delusional mood, loosening of association and disorganised behaviour. He was admitted to a mental hospital and the symptoms disappeared after treatment with various antipsychotics for about three weeks. At the ages of 28 and 30 years, he was admitted to the same mental hospital with symptoms such as illogical thought, psychomotor excitement and impulsive behaviour. After 2 months in the hospital, these psychiatric disturbances were
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