Letters

neck-abdominal reflex, and seems to indicate severe brainstem damage, and hence a poor prognosis.

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References

Accepted 10 January 1984

Spinal neck-tongue syndrome

Sir: The neck-tongue syndrome was first described by Lance and Anthony. Their patients experienced pain in the neck with or without numbness and numbness in the tongue on sudden turning of the head. The authors attributed the symptoms to compression of the C2 roots in the atlanto-axial space; the numbness of the tongue was caused by compression of proprioceptive fibres from the tongue through the ansa hypoglossi, the cervical plexus and finally the C2 roots. We wish to report a patient who developed unilateral numbness in the neck and tongue of spinal origin.

A 35-year-old woman developed symptoms of acute transverse myelopathy in the upper thoracic spinal cord at the age of 23 years. After recovery, she had eight similar exacerbations and remissions during the following ten years. Myelography at the age of 28 years was normal. At the age of 33, she became unable to walk owing to paraplegia and was confined to a wheelchair. At the age of 35, weakness of the left upper limb developed acutely. Neurological examination on admission revealed positive Lhermitte sign and paraesthesia on the left between C2 and C3 segment. Marked weakness with hyperreflexia was noted in the left upper limb. Joint position sense of the left upper limb was severely impaired. There were motor and sensory deficits due to transverse thoracic myelopathy which persisted since the age of 33 years. Laboratory examinations were normal except for a mild pleocytosis in the spinal fluid. An EEG and a CT scan were negative.

Tonic seizures appeared after the signs and symptoms of the neck and left upper limb had subsided considerably. Voluntary movement of the left arm provoked tingling sensation in the left hand, which radiated to the shoulder. Simultaneously the spasm appeared in the left upper limb, with fingers flexed, wrist and elbow flexed and the arm abducted. The tonic spasm lasted about 60 seconds. About 20 seconds after beginning, the neck and occipital region on the left side became numb, accompanied by simultaneous numbness of the left half of the tongue. Other parts supplied by trigeminal nerves were not involved. The numbness lasted from 30 to 60 seconds. The seizure occurred in a stereotyped fashion about once an hour. Carbamazepine, 400 mg daily, suppressed the attacks.

This patient developed unilateral numbness in the neck and tongue during the tonic seizures following the upper cervical lesion. The neurologic signs on admission suggested involvement of the left posterior funiculus and left corticospinal tract. The cause of the myelopathy was not clear. The exacerbating-remitting course may indicate demyelination. Tonic seizures are one of paroxysmal neurological disturbances in multiple sclerosis and also have been reported in traumatic injury of the spinal cord. Although their pathophysiology is unknown, it seems probable that in some cases such tonic seizures have a spinal origin. Lance and Anthony reviewed the available data on the course of afferent fibres from the tongue and concluded that they pass via the ansa hypoglossi to the C2 dorsal roots. The pathway of the afferents from the tongue after entering the central nervous system was little discussed. Bowman and Combs showed that the hypoglossal afferents in the rhesus monkey project rostrally in the ipsilateral dorsal funiculus via the dorsal root ganglia of the C2 and/or C3 nerve. It seems reasonable to postulate that a similar situation obtains in humans. The numbness in the neck and tongue may be explained by involvement of the dorsal funiculus during the tonic seizures. However, an alternative explanation may be possible. Sensory fibres of the mandibular branch are distributed to the tongue and descend in the brainstem as the spinal trigeminal tract. They terminate at the medullary level or extend into upper cervical spinal segments. Disturbance of this tract may cause the numbness in the tongue.

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References

Accepted 10 January 1984

HLA antigens and multiple sclerosis in Greeks

Sir: While the relationship between the major histocompatibility system (HLA) and susceptibility to multiple sclerosis is well documented in northern European populations, there has been controversy over the relationship for ethnic groups of Mediterranean origin. Most workers have noted an increased frequency of HLA-A2 and B, antigens in multiple sclerosis,

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patients in central and north Europe, \(^1\) while an analogous evidence is lacking in Italian, \(^3\) and Israeli patients. \(^4\) Such an association between HLA-A, and B, antigens and multiple sclerosis also has not been proven in Japanese \(^5\) and Indians. \(^6\)

In view of the unique immunogenetic profile of the Greek population \(^7\) we studied the HLA A and B specificities in 81 Greek patients with a well established diagnosis of multiple sclerosis. Fifty specific antisera were used, determining 24 HLA A and B antigens (9 and 15 respectively) according to the standard microlymphocytotoxicity test.

The results were compared to those of 280 healthy unrelated controls. For statistical evaluation, the X\(^2\) analysis was applied after Yates correction and multiplication of the p values by the number of antigens tested (P corrected). As it can be seen, (table) the frequency of HLA A, and B, antigens in the patients group (44-46% and 32-33% respectively) was significantly greater than that in the controls (26-07% and 13-21% respectively). The relative risks (RR) for the antigens A, and B, were 2.23 and 3.23 respectively.

Our results thus show that Greeks, unlike Caucasian ethnic groups of the Mediterranean area, there is an increased frequency of HLA A, and B, antigens in multiple sclerosis patients, analogous to that observed in populations of central and northern Europe. We think that this observation, should be reported, as contributing to understanding of the complex immunogenetic profile of this disorder.

\(^2\) RR 2.23 X\(^2\) = 10.46 P corrected 0.001

\(^3\) RR 3.33 X\(^2\) = 24.83 P corrected 0.001


Accepted 30 January 1984

Papilloedema with extravascular erythropoiesis and Cushing's syndrome

Sir: Extravascular erythropoiesis may cause a variety of clinical problems depending on the site or sites at which it occurs. Various locations are well recognised but central nervous system involvement, although documented, is relatively uncommon. We describe a case where there was cranial involvement presenting in very unusual fashion, the clinical features arising because of the particular location of the erythropoietic tissue in the pituitary fossa, in a patient who concomitantly and coincidentally had a pituitary adenoma.

A 55-year-old Greek Cypriot lady presented in 1977 with a fracture of her humerus following a fall. A blood count showed her to be anaemic. On direct questioning she gave a history of listlessness for at least six years and a family history of anaemia. Examination revealed a moderately enlarged spleen, first noted at the age of 10 years. Investigations showed: haemoglobin 8.4 g/dl; red cell appearances of thalassaemia intermedia; starch gel electrophoresis showed mainly HbA with HbA\(_{2}\), 6-4% and HbF 8.6%, confirming the diagnosis of beta thalassaemia intermedia. Routine chest radiographs showed a posterior mediastinal mass which proved to be extravascular erythropoiesis on \(^{57}\)Fe scanning. This also demonstrated increased uptake by the spine, pelvis and liver.

In 1980 her face became puffy and her changed appearance clinically suggested Cushing's syndrome. This was confirmed by the following investigations: 9:00 am cortisol 755 nmol/l; midnight cortisol 775 nmol/l; 9:00 am cortisol following 1 mg dexamethasone at 11 pm, 945 nmol/l; urinary free cortisol 6485 nmol/24 hrs; plasma ACTH 17 ng/l. Her plasma prolactin was above 5000 mU/l on three occasions. It was noted from her history that she had no children and had had secondary amenorrhoea since the age of 33, but had no galactorrhoea.

Skull radiographs showed erosion of the posterior clinoid processes, but computed tomography did not show expansion of the sella turcica or any abnormality in the pituitary fossa. At this stage she was considered disabled by a proximal myopathy.
HLA antigens and multiple sclerosis in Greeks.

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*J Neurol Neurosurg Psychiatry* 1984 47: 751-752
doi: 10.1136/jnnp.47.7.751-a

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