Transient neurological disturbances in disseminated sclerosis: a case report

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Transient neurological disturbances occurring in patients with disseminated sclerosis have recently been receiving increasing attention (Andermann, Cosgrove, Lloyd-Smith, and Walters, 1959; Espir, Watkins, and Smith, 1966). Espir et al. (1966) described six patients with frequent short-lived attacks of dysarthria with or without other symptoms of brain-stem dysfunction. The nature of the attacks was not clear, but by analogy with trigeminal neuralgia they were treated with phenytoin (Epanutin) and carbamazepine (Tegretol) (Espir et al., 1966; Kuroiwa and Shibasaki, 1967; Espir and Walker, 1967). Both these agents were effective in some of the patients but a tendency to spontaneous remission made assessment difficult. A further case of disseminated sclerosis with recurrent attacks of dysarthria and inco-ordination has recently been seen, and has made possible an assessment of therapeutic agents. The following is a case report of this patient.

**CASE REPORT**

R.M. is a 20-year-old man who had an episode of diplopia for two months in 1964. In 1965, for a similar period, he noted clumsiness of the left hand. In the spring of 1966 he felt some stiffness in his legs and began to notice that he was dragging his right foot. In May 1966 he had numbness over the left side of the trunk for one week. In an admission to hospital in December 1966 he was found to have lateral nystagmus, mild inco-ordination in the arms, and a spastic ataxic paraparesis. Investigations then revealed a high CSF protein (150 to 250 mg%,) with negative WR and Lange curve. Air encephalography and myelography were normal and a diagnosis of disseminated sclerosis was made. In November 1967 he had a retrobulbar neuritis in the right eye.

He was readmitted to hospital in March 1968 for the investigation of the occurrence of brief attacks of diplopia, dysarthria, and clumsiness of the right side of the body. These had developed suddenly three weeks previously and were occurring as frequently as once every three or four minutes. There were no abnormalities on general examination. Speech was mildly dysarthric. There was bilateral optic atrophy and first degree nystagmus to right and left and on vertical gaze. Tone was slightly reduced in the arms and increased in the legs. Power was normal. There was cerebellar ataxia of all four limbs. The tendon reflexes were pathologically brisk, with bilateral extensor plantar responses. Sensory testing revealed subjective diminution of vibration sensibility in the legs. His gait was spastic and ataxic.

Each attack lasted for 15 to 20 seconds and consisted of diplopia, a severe scanning and slurring dysarthria, and severe ataxia of the right arm and leg. If he were talking when an attack started he became almost unintelligible; if walking, he was forced to stop and seek support to prevent a fall. The attacks were of sudden onset and cessation and were not accompanied by any clouding of consciousness. They were sometimes precipitated by emotional stress, but no other factors appeared to influence their timing, frequency, or content.

As a test of co-ordination of the right hand the patient was requested to draw a regular sine curve on moving paper (Fig. 1), and to copy a paragraph from a journal (Fig. 2). The abrupt nature and short duration of the attacks is well seen. An EEG was normal and no change accompanied clinical attacks.

A course of ACTH was given in hospital without effect on the attacks. He was therefore treated with Tegretol, 100 mg t.d.s. The attacks stopped in three days. Unknown to the patient placebo Tegretol tablets were substituted two weeks later with recurrence of the attacks within 24 hours. Subsequently, phenobarbitone 50 mg t.d.s. and primidone, 250 mg t.d.s. were tried and found ineffective. Each time Tegretol was stopped the attacks recurred unchanged. Over the following three months their frequency (when untreated) fell to three or four a day.

**DISCUSSION**

The diagnosis of disseminated sclerosis is well established in this case, as in five of Espir's six cases (Espir et al., 1966). The transient attacks are also very similar, involving dysarthria and other symptoms localizing the responsible lesion to the brain-stem. The nature of the attacks is of great interest. Their rapidity of onset and offset is suggestive of an epileptic phenomenon, although
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The evidence of focal epileptic phenomena in the brain-stem is not universally accepted.

Temporary exacerbations of symptoms in disseminated sclerosis are well known after such stimuli as exercise and hot baths (McAlpine, Compston, and Lumsden, 1955). Espir et al. (1966) noted some relationship of the attacks of dysarthria to stress and overbreathing. The first but not the second factor seemed to influence our patient's attacks. The stereotyped nature of the attacks suggests that each is due to an epileptic discharge resulting from the changes around a brain-stem plaque.

It does not seem likely that they are due to a temporary exacerbation of pre-existing symptoms, as, in some of the patients described, the attacks were the first indication of a lesion in the brain-stem. Also, the attacks were strikingly similar between patients who otherwise had widely different clinical signs.

Because of the tendency to spontaneous remission in disseminated sclerosis, it has previously proved difficult to obtain convincing evidence of the efficacy of various therapeutic agents. In our case the attacks were sufficiently persistent for several drugs to be tried and their effects compared. Phenobarbitone, Mysoline, and placebo were all ineffective, only Tegretol influencing the attacks.

In the experimental animal it has been shown that both phenobarbitone and Tegretol affect electrical activity in the brain-stem. Phenobarbitone decreases the amplitude of evoked responses, whereas Tegretol has been shown to depress synaptic transmission (Hernandez-Peon, 1966; Fromm and Killian, 1967). Tegretol affects both monosynaptic and polysynaptic pathways (Theobald and Kunz, 1963). In view of this evidence of the action of Tegretol, and its efficacy in the control of the transient attacks, it is suggested that these are due to a build-up of post-synaptic facilitation from a discharge around a brain-stem plaque. Tegretol may depress such post-synaptic activity, thereby preventing an attack.

FIG. 1. Shows the patient's attempt to draw a regular sine wave on moving EEG paper. The short period of inco-ordination is clearly shown, with the limits of the attack marked by arrows. Time scale markers 1 second and 5 seconds.

FIG. 2. The patient was asked to copy a newspaper advertisement and to continue writing during an attack. This shows both the frequency and the short duration of the attacks of inco-ordination.
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SUMMARY
A patient with disseminated sclerosis is described who reported transient attacks of dysarthria and ataxia. Evidence is presented that Tegretol may have a relatively specific therapeutic effect on such attacks.

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REFERENCES


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