PAROXYSMAL TONIC SEIZURES IN DEMYELINATING DISEASE

C T Tan
E T Low

Department of Medicine
University of Malaya
Kuala Lumpur
Malaysia

C T Tan
Associate Professor

E T Low

SYNOPSIS

Paroxysmal symptom is a well known feature of disseminated sclerosis. Paroxysmal tonic seizure, in particular, is frequently seen in disseminated sclerosis among patients from Japan and Taiwan. Three patients with paroxysmal tonic seizure from demyelinating disease seen in the University Hospital, Kuala Lumpur, were discussed in this paper. The clinical pattern of the seizure seemed closer to the Japanese patients, with bilateral symptom in two of the cases. The seizure was probably spinal cord in origin and responded promptly to carbamazepine.

INTRODUCTION

Multiple sclerosis patient has been known to present with several forms of paroxysmal symptom. They are: paroxysmal tonic seizure (P.T.S.), paroxysmal ataxia and dysarthria, paroxysmal paraesthesia, itch and pain, and trigeminal neuralgia. The exact incidence of the paroxysmal symptom is not clear. Shibasaki and Kuroiwa (1) reported 17% of 64 Japanese multiple sclerosis patients developed P.T.S. sometime during the course of their illness. They also commented that other paroxysmal symptom were less commonly seen. Espir and Millac (2) on the other hand,

reported that 32 out of 600 patients (5.3%) with definite or suspected multiple sclerosis had paroxysmal symptoms of various types. Hung et al. (3) reported 28% of their multiple sclerosis patients in Taiwan had P.T.S., a relatively high incidence. In this paper, we like to present three patients with demyelinating disease seen in the University Hospital, Kuala Lumpur, in 1983 and 1984 with P.T.S.

CASE 1

LSE was a 43-year-old Chinese housewife from Penang who was previously in good health. She was first seen in U.H., K.L., for progressive weakness and numbness of both lower limbs of one week's duration. Physical examination then showed spastic paraparesis with sensory level at T5. Myelography and CSF examination were normal. Diagnosis of transverse myelitis was made. The patient responded to a short course of dexamethasone with only minimal residual signs.

P.T.S.

It started a month after onset of illness when the patient was recovering from the paraplegia. The seizure commenced as a tight sensation from left side of the trunk to the left foot, this was accompanied by flapping of the left foot, then inversion of the same foot and plantar flexion of the left big toe. This was followed by plantar flexion of the right foot with extension of the right big toe and splaying of the other toes. The right hip became flexed and adducted. Concomitant with the motor spasm was tightness and numbness from T5 left side radiating distally to both lower limbs as well as proximally to the left ear lobe. The whole episode lasted about two minutes. It may abort without going through all the sequence. The attack was precipitated by sitting up from lying position, turning the trunk, putting the feet on the ground to walk and rubbing firmly on the area of hyperaesthesia at the left lower chest. Numerous attacks occurred in a day. EEG done during the attack was normal. Carbamazepina at 200 mg q.i.d. promptly reduced the number and intensity of the seizure. One and a half year after the initial illness, there was still relapse of the seizure if she missed the medication; although the seizure was less intense and frequent.

CASE 2

L.L. was a 54-year-old Chinese lady who developed progressive tetraparesis, sphincteric incontinence and right-sided Horner's Syndrome over one month. The CSF and CT scan of the head were normal. She was given a short course of prednisolone. Subsequently she made progressive recovery until only mild reflex changes were noted six months later. 18 months later, a relapse of tetraparesis occurred which recovered over 4 months. Cervical myelography, visual and brainstem auditory evoked responses were normal. Somatosensory evoked response showed prolonged N9-N20 segment on stimulation of right median nerve.

P.T.S.

It started during the first episode of tetraparesis with hot sensation of left foot spreading up to the same buttock. Then the right thumb became adducted and fingers of right hand flexed at metacarpal phalangeal joints and extended at interphalangeal joints. The right elbow was extended and the right foot plantarflexed and inverted. The whole episode lasted about 30 seconds with up to 20 attacks per day. The seizure occurred spontaneously or precipitated by sudden movement of the trunk. She responded to carbamazepine 200 mg. t.d.s. The P.T.S. persisted for a year after the initial illness. There was a recurrence of the same seizure when the patient has a relapse of the tetraparesis.

CASE 3

O.B.H. was a 20-year-old salesgirl from Penang. She first presented in August 1984 for a month's history of vertigo. Physical examination did not show any abnormality. Further investigation with EEG, CT scan were normal. CSF study showed raised protein of 57 mg% and pleocytosis with 8 lymphocytes/μl. Electronystagmography done showed left directional preponderance in the calorie test. Clinical diagnosis of an episode of brain stem demyelination was made.

She improved symptomatically until 2 months later, when she developed numbness in both hands, right leg and weakness of right hand. Physical examination showed global weakness of right upper limb and hyperreflexia of the right upper and lower limb. CSF repeated showed raised CSF protein of 67 mg% and 8 lymphocytes/μl. She was given two weeks' course of ACTH with which she improved. Visual and Brain stem Auditory Evoked Response were normal. Somatosensory evoked response showed delayed central conduction time (N13-N20) segment.

P.T.S.

It developed when the ACTH was being tailed off. The attack consisted of numbness in the right hand and right leg followed by tonic spasm of the right upper limb and sometimes right lower limb. The attack lasted about 1-2 minutes. The hand was in carpal spasm posture, and the right foot when involved was in plantar flexion. There may be associated generalized sweating. The attacks usually came on spontaneously but sometimes precipitated by foreceful usage of the right hand or getting up from the bed quickly. The attacks occurred up to 20-30 times/day. It responded promptly to Carbamazepine 200 mg. t.d.s. No further seizure was seen after 6 months.

DISCUSSION

To date, more than 60 patients with P.T.S. have been reported in the literature (4). The largest series of 15 patients were reported by W.B. Matthews (5) from Oxford, England. The seizures were all unilateral. The arms were invariably involved, and the ipsilateral legs affected in nine patients. In nine patients, the hand adopted tetanic posture. The seizure was extremely painful in seven patients, this being the same distribution as the spasm. In two patients, the pain preceded the muscle contraction. Thirteen patients noted precipitation of the seizure by movement or sensory stimulation. Putting the foot to the ground, on standing up or turning over in bed were the motions most frequently noted. Seizures were always brief, the longest observed being 90 seconds. Up to 30 seizures per day were common. The seizure persisted up to two months, remitting either spontaneously or to treatment. Tonic seizures were not accompanied by disturbance in conscious state. The good response to carbamazepine was first noted by Kuroiwa and Shibasaki (6).

It is interesting to note that in contrast, nine out of eleven patients reported by Shibasaki and Kuroiwa (7) from Japan had bilateral symptom. The author attributed the symptom to phatology in the spinal cord and noted that Japanese multiple sclerosis patients have

frequent pathological finding of severe necrotic demyelinating lesion involving the white matters as well as grey matters of the spinal cord. Two of our three patients have bilateral symptons. In this way, the clinical manifestations of our patients are closer to the Japanese cases. Perhaps, the pathology of our patients, all being ethnic Chinese, is also more similar to that of the Japanese.

Shibasaki and Kuroiwa (1) pointed out that none of their patients showed paroxysmal EEG abnormality in association with P.T.S. None of them had impairment in consciousness in spite of frequent bilateral involvement. Therefore, it seemed unlikely that P.T.S. involved the cerebrum. The authors considered spinal cord to be the origin of the P.T.S. in their patients. This was supported by the clinical evidence of severe spinal cord involvement during the course of illness in all the eleven patients reported. Seven of the eleven patients in addition had Lhermitte's sign which give further support to the spinal cord origin of P.T.S. However, it is likely that P.T.S. can also originate from other site, as evidenced by the simultaneous occurrence of tonic seizure and dysarthria in two of the patients reported by Matthews (5). We consider spinal cord to be the origin of our patient's P.T.S. Our Case 1 and 2 clearly have thoracic and cervical cord lesion. Case 3 has demyelination plaque probably in cervical cord as well as the brain stem. However, there was no bulbar feature in her P.T.S.

The main differential diagnosis of P.T.S. are flexor spasms and tetany. Sensory symptom may precede the spasm in P.T.S. whereas the pain in tetany and flexor spasm correspond to the muscle contraction. There is a peculiar spread of painful spasm in P.T.S. the pattern of which only bears superficial resemblance to carpopedal spasm of tetany and the flexor spasm. The P.T.S. attack is often more prolonged than that of flexor spasm. Furthermore, there is no correlation between P.T.S. and the intensity of paralysis or the Babinski toe sign. Whereas flexor spasm usually occur in association with spastic paraplegia with Babinski sign.

As for the pathophysiology of the P.T.S., Ekbom et al. (7) postulated a transversely spreading activation of axons in the lateral funiculus at the affected level in the spinal cord. This would explain the Brown-Sequad Syndrome in reverse where the motor spasm is seen in one side and the sensory paroxysm in the other side as in Case 2. The stream of impulses may also travel contripetally along the sensory path explaining the tight sensation in the cephalic region experienced by Case 1. This abnormal activation in a demyelinated spinal cord is sometimes triggered by afferent input explaining the observed precipitation of the attack by sensory stimulation (5). Bilateral motor and sensory attack as seen in Case 1 can be explained by the transverse spread of activation across the spinal cord; or the triggering of the contralateral cord by the peripheral sensory input. No other example of clonic movement in the P.T.S. as was seen in our Case 1 has been reported in the literature.

REFERENCES

1. Shibasaki H, Kuroiwa Y: Painful tonic seizures in Multiple Sclerosis. Arch Neurol 1974; 30: 47-51.
2. Espir MLE, Millac P: Treatment of paroxysmal disorders in multiple sclerosis with carbamazepine. J Neurol Neurosurg Psychiat 1970; 33: 528-31.
3. Hung TP, Landsborough D, Hsi MS: Multiple sclerosis among Chinese J Neurol Sci 1976; 27: 459-84.
4. Twomey JA, Espir ML: Paroxysmal symptoms as the first manifestations of multiple sclerosis. J Neurol Neurosurg Psychiat 1980; 43: 296-304.
5. Matthews WB: Paroxysmal symptoms in multiple sclerosis. J Neurol Neurosurg Psychiat 1975; 38: 617-23.
6. Kuroiwa Y, Shibasaki H: Carbamazepine for tonic seizure in multiple sclerosis. Lancet 1967; 1: 116.
7. Ekbom KA, Westerberg CE, Osterman PO: Focal sensorymotor seizures of spinal origin. Lancet 1968; 1: 67.