

Case study

Epileptic falling spells after epidural haematoma in adult Down's syndrome

M. YOSHIDA[†], Y. KURODA[§] and
T. YOSHIDA[‡]

[†]Department of Neurology; and [‡]Department of Radiology, Division of Visually Impaired, Tsukuba College of Technology, City of Tsukuba, Japan

[§]Department of Circulation, Mito General Hospital, Miyamachi, Japan

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A 35-year-old man with Down's syndrome showed epileptic falling spells. He had suffered from a traumatic right-sided epidural haematoma 3 years before. It had been neurosurgically treated, but MRI taken 5 days later had revealed a small contracoup contusion at the left temporal lobe. His falling spell was a brief tonic seizure without disturbance of consciousness. Background activities of EEG consisted of slow α waves interspersed with sporadic θ waves and the amplitude at the left temporal area was lower than the opposite one. Interictal EEG showed sharp waves or sharp and slow wave complexes predominantly at the right temporo-centro-parietal area as well as diffuse, though predominantly at frontal areas, bursts of slow waves with high amplitude. The EEG suggested focal epileptic activities evolving into secondary generalization. SPECT of the brain showed the hypoperfusion at the left temporal area and at the right posterotemporo-parietal area, where the hypoperfusion was somewhat reduced after the improvement of seizures. Seizures were well controlled with phenytoin combined with phenobarbital. The incidence of epilepsy in the Down's syndrome has been reported to increase after the middle age in association with the development of Alzheimer's neuropathology. When those people would sustain head injuries, it was necessary to follow carefully using SPECT and EEG.

Introduction

Prevalence rates of epilepsy in Down's syndrome (DS) are relatively small as compared with that of individuals with other mental retardations, where prevalence rates of seizures are from 20 to 50% [1]. However, the incidence of seizures in cases with DS has been reported to increase after the age of 30 or 40 years.

Epileptic falling spells are induced by multiple seizure types [2]. However, cases who showed only falling spells have been rarely reported in adult cases of DS with epilepsy. This study presents an adult case with DS who developed falling seizures after he had suffered from a traumatic epidural haematoma.

Case report

A 35-year-old man was referred to the neurological department for falling spells in October 1997. He showed falling spells five times without disturbance of consciousness in September 1996. At that time, EEG revealed sporadic sharp waves at the

Correspondence to: M. Yoshida, M.D., Department of Neurology, Division of Visually Impaired, Tsukuba College of Technology, 4-12-7, Kasuga, City of Tsukuba, 305-0044, Japan.

right centro-parietal region. The spells disappeared for a while without treatments. However, they occurred again a few times monthly since May 1997.

His past history revealed that he was suffering from DS. He had a congenital heart disease as well. He suffered from an attack of gout in 1989. He was staying in an institution for mentally-handicapped people. The right-sided acute epidural haematoma was caused after his head had been struck by a colleague in September 1994. He was surgically treated on the same day without neurological sequelae. However, a small contracoup contusion at the left temporal lobe was revealed by MRI taken 5 days after the operation (figure 1). He was prophylactically treated with valproate for 1 year. Thereafter, it was ceased since EEG showed no epileptic abnormality.

His family history was unrevealing.

His falling spells occurred when standing, walking or working in the sitting position. He usually fell backward rather than forward without myoclonic jerks or atonia. It was followed usually by no symptoms though, on rare occasions, followed by a mildly rigid phase for up to 10 seconds or an arrest for approximately 10 seconds. After falling, he responded to calling at once or after a delay of up to 10 seconds and stood up by himself. His facial colour was not altered or occasionally slightly livid.

He was 145 cm in height and 37 kg in weight. His physical features were correspondent with those of DS. His pulse was regular, but a murmur was audible

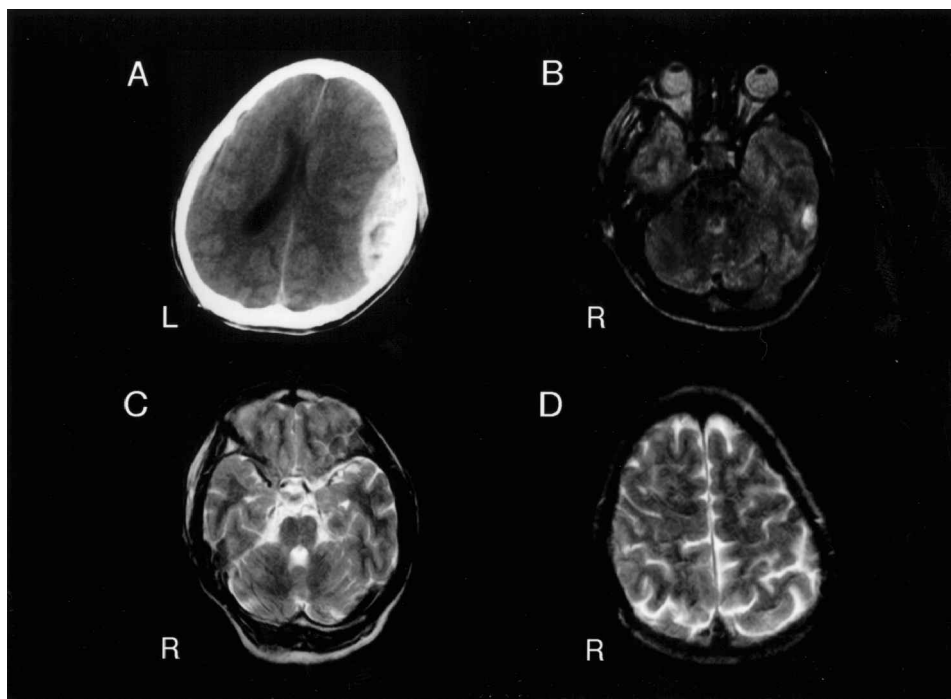


Figure 1. CT and MRI of brain. (A) Epidural haematoma was shown in CT taken in September 1994. (b) Contracoup contusion at the left temporal region was revealed by T2-weighted MRI taken 5 days after (A) had been taken. (C) and (D) T2-weighted MRI taken in October 1997 showed no localized brain atrophy.

over the whole heart. Blood pressure was 118/40 mmHg. There was no orthostatic hypotension.

Neurological examinations revealed mild rigidity in the right arm as well as the neck. Myerson's sign, snout reflex and palmo-mental reflex were positive. There was neither constitutional apraxia nor dressing apraxia. There was neither left-right agnosia nor finger agnosia. The mental development was severely retarded. He could not tell the date, and could neither retain three names of flowers nor make simple calculations. However, this mental ability did not deteriorate after the appearance of falling spells. He could carry out simple daily work such as folding towels or packing face cloths and tooth brushes into handy bags. Care-takers did not notice the recent deterioration of ability of such daily works. His ADL was perfect. His personality did not change.

The analysis of chromosome showed a standard 21 trisomy. Urinalysis, blood analysis and routine blood chemistries were normal. T3, T4 and TSH levels in serum were normal. Ultrasonic cardiography showed a ventricular septal defect.

Head CT and MRI revealed biputaminal calcifications, but there was neither cortical atrophy nor ventricular dilatation (figure 1).

Head ^{123}I -IMP SPECT revealed the obvious hypoperfusion at the right postero-temporo-parietal region and the slight one at the left anterotemporal region (figure 2).

Background activities of EEG consisted of 7 Hz fast θ to 8 Hz slow α waves, interspersed with sporadic 5 Hz θ waves diffusely. The amplitude at the left temporal area was lower, as compared with the right one.

Sporadically sharp waves and occasionally sharp and slow wave complexes appeared predominantly at the right temporo-centro-parietal areas. Bursts of 4 Hz slow waves with high amplitude appeared diffusely at the drowsy state or by photic stimulation (figure 3).

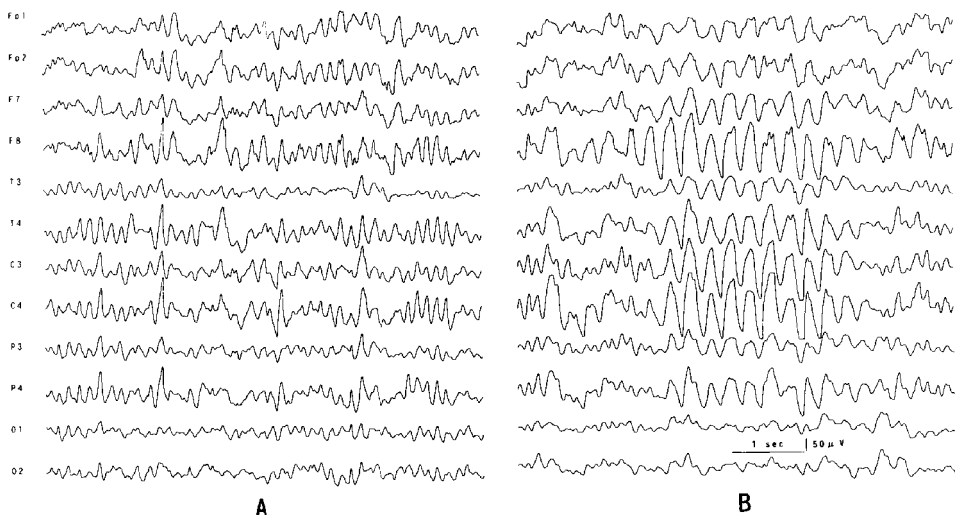


Figure 2. EEG. (A) Sharp waves appeared at the temporo-centro-parietal region of the right hemisphere and background activities were slow. (B) Generalized high amplitude slow wave bursts appeared at the drowsy stage and by flicker stimulation.

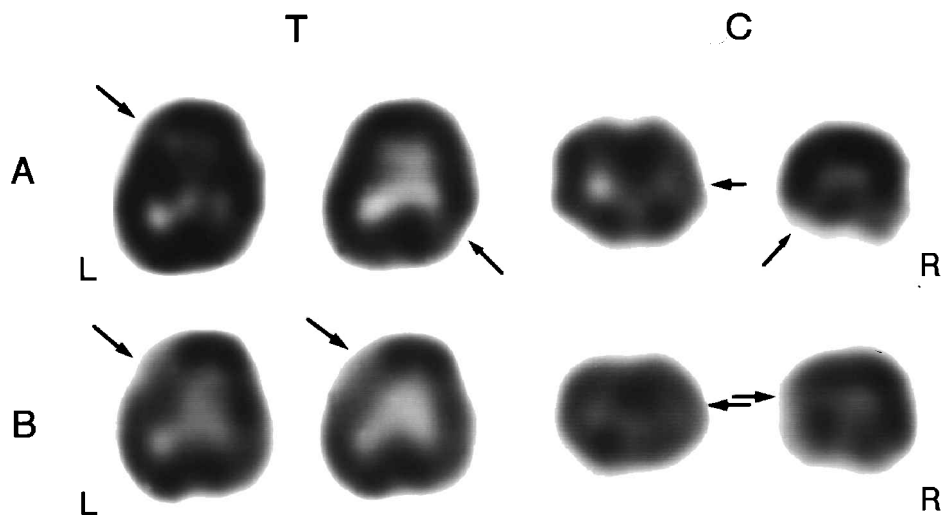


Figure 3. SPECT of brain. (T) transaxial section. (C): coronal section. (A) ^{123}I -IMP SPECT of brain showed the uptake defect of radio-pharmaceuticals clearly at the right posterotemporo-parietal region and mildly at the left anterotemporal region. (B) $^{99\text{m}}\text{Tc}$ -HMPAO SPECT taken 5 months after A had been taken showed that the hypoperfusion at the right posterotemporo-parietal region was reduced. On the other hand, the hypoperfusion at the left temporal region became extended.

Subsequent courses

The patient was initially treated with valporate 200 mg b.i.d. However, falling spells were not decreased. When valporate was increased to 200 mg t.i.d., seizures were rather increased. After phenytoin 50 mg combined with phenobarbital 17 mg t.i.d. was substituted, seizures almost completely disappeared.

EEG and SPECT were taken 5 months later. The EEG showed the reduction of sharp waves or spikes. Lower amplitudes at the left temporal region were not altered. $^{99\text{m}}\text{Tc}$ -HMPAO SPECT showed some reduction of hypoperfusion at the right posterotemporo-parietal region (figure 2). On the other hand, the hypoperfusion at the left temporal region became more extended.

Discussion

The patient fell backward on his buttock. He showed neither myoclonus nor atonia in the seizure. A brief tonic phase with arrest was observed on rare occasions after falling. Those manifestations indicated that his falling spell was the seizure associated with a brief tonic component.

The epileptic focus was localized at the right temporo-centro-parietal region, since sharp waves predominantly appeared there. There were also generalized bursts of slow waves with high amplitude. Those findings of EEG suggested that epileptic activities originated focally and evolved into the secondary generalization.

Egli *et al.* [3] reported that the predominant type of falling seizure in patients with secondary generalized epilepsy was pure tonic drop seizure, where patients fell since axial spasms involved hip joint muscles.

Epileptic falling attacks may appear in a number of different seizures types [2]. Most patients with falling spells have concurrently one or more types of seizures. Patients complaining of falling spells alone, as in this case, were relatively rare [2].

Patients with DS may have epileptic seizures of all types [4]. In adult patients with DS, partial seizures evolving into secondary generalized tono-clonic seizures or primary tono-clonic seizures are most frequent [5–8]. Adult patients with DS who had only falling seizures have rarely been reported. McVicker *et al.* [7] reported only one case with sudden fall out of 18 adult patients with DS. Stafstrom *et al.* [1] reported one case of pure atonic seizure and three cases of atonic plus tono-clonic seizure out of 47 DS cases with epilepsy. However their patients were less than 22 years old.

The prevalence rate of epilepsy is smaller in DS as compared with other mental retardation [9]. However, the recent longevity of patients with DS increases the prevalence rate of epilepsy. Prasher [8] reported that the prevalence rate of epilepsy in adult DS was 15.9% with the bimodal distribution of seizure onset in childhood and in middle age. The latter was probably due to Alzheimer's dementia. Stafstrom [4] reported that cases with DS had distinct peaks of the incidence of seizures in infancy, in teenage years and in the 4th and 5th decades. The last peak was coincident with the onset of Alzheimer-type neuropathologic changes. Wisniewski *et al.* [10] reported that senile plaques and neurofibrillary tangles were observed in 100% of individuals with DS and 27% of them were clinically demented. Mann [11] reported that neuropathological changes of Alzheimer's disease in DS appeared in 7.5% of patients already in the 2nd decade, in 80% of those in the 4th decade and almost 100% of those after the age of 40 years. The onset time of later onset epilepsy has been reported to coincide with the onset time of Alzheimer's dementia in DS [7].

The standardized diagnostic criteria for dementia were useless in this case, since the mental retardation was so severe. However, the obvious mental deterioration was not noted, since no deterioration of both ADL and the ability of working was noticed. No change in personality was noticed. Lai and Williams [6] suggested that there was an incubation period of 20–30 years between neuropathological evidences and clinical expression of dementia. The slowing of background activity on EEG in this case suggested the deterioration of brain function.

Generalized slowing of background activity on EEG was most common in middle-aged DS patients with seizures, as well as without them [6, 7, 12, 13]. These abnormalities suggest the premature deterioration of brain function consistent with Alzheimer's neuropathology [13, 14]. Focal (temporal or parietal) or non-focal paroxysmal activities were also described [6, 7]. Bursts of slow waves at temporal areas similar to this case have been also reported [18].

The slowing of background activities and bursts of high amplitude slow waves in this case were consistent with those findings.

The traumatic injury in combination with the underlying vulnerability of brain characteristic of adult DS was supposed to induce such an epileptic seizure in this case. Conversely, it was supposed that the brain trauma accelerated Alzheimer's neuropathology, resulting in the induction of seizures.

Although MRI showed no localized brain atrophy as the post-traumatic sequelae, SPECT revealed the regional hypoperfusion. SPECT, taken initially, showed the hypoperfusion at the right temporo-parietal region. It was somewhat decreased when both falling spells and paroxysmal activities on EEG were

decreased. In contrast, the hypoperfusion at the left temporal region became extended in the follow-up study.

SPECT of intractable epilepsies, especially complex partial seizures, has been reported to show the decreased regional perfusion in the interictal period. In some cases, hypoperfused regions on SPECT were not always correspondent to epileptic foci on EEG [18–22].

It has been reported that SPECT or PET demonstrated hypoperfusion or hypometabolism in brains of demented patients with DS [15, 23–25]. The pattern was similar to that of Alzheimer's disease [26, 27]. The hypoperfusion at bifrontal lobes and the right temporal lobe was also shown in the early stage of dementia with DS [28]. Even non-demented young patients with DS showed unilateral perfusion defects in the temporo-parietal or parieto-occipital regions [29].

It was supposed that the right temporo-parietal hypoperfusion in this case was induced by the epileptic foci, due to the traumatic lesion which had a probable Alzheimer's neuropathology.

Lower amplitudes of EEG and the hypoperfusion at the left temporal area in this case should be attributed to the sequelae of contracoup contusion. However, it was equivocal that such a small contracoup lesion alone produced the long-lasting dysfunction of such a broad area. The contribution of the probable underlying Alzheimer's neuropathology should also be taken into consideration.

Epileptic falling spells are thought to be intractable [2]. Snead [2] described that valproate was effective against epileptic falling spells and virtually all types of epileptic falls resisted phenytoin and phenobarbital. In this case, valproate was ineffective and phenytoin combined with phenobarbital was effective. The easy controllability of seizures in patients with DS has been reported [4, 5, 30].

It was reported that 45.2% of epileptic seizures were associated with falling and 6.1% of falling spells were associated with head injury [31]. Falling stiffly, as shown in this case, was more at risk as compared to atonic seizures.

An adult patient with DS after sustaining a traumatic brain injury should be followed carefully. In that case, SPECT and EEG would be useful, since these functional studies would reveal the hypofunction of the brain before CT or MRI would do.

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