Short report

Comprehension as the effective trigger in a case of primary reading epilepsy

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SUMMARY Reading epilepsy is associated with a variety of perceptual, motor and/or high level cognitive factors. A case of primary reading epilepsy is reported in which comprehension was the effective stimulus in provoking epileptiform activity.

Primary reading epilepsy is an uncommon type of reflex epilepsy. In particular, primary reading epilepsy cases in which seizures are evoked by higher cognitive functions are extremely rare. The present study systematically documents such a case in which text comprehension appears to be a sufficient trigger for primary reading epilepsy.

Case report

KM was a 29 year old right-handed non-identical twin. During the past 9 years he had suffered nine major seizures, always following reading. He had been prescribed phenytoin and more recently sodium valproate but continued to experience a tingling sensation in the tongue and myoclonic jerking of the jaw when reading even relatively short passages.

His past medical history was unremarkable. Physical, neurological examinations and a CT scan were all normal. His resting EEG or during photic stimulation and hyperventilation was also normal but during reading paroxysmal generalised EEG abnormalities were recorded. Pattern visual evoked response testing did not show unequivocal abnormalities. Also, there was no suggestion of sensitivity to intermittent photic stimulation when KM was required to observe stationary patterns of striped lines of epileptogenic spatial frequencies on a TV screen.

His verbal IQ was 128, his performance IQ 111 and the discrepancy between them was likely to be long-standing. On a comprehensive series of focal cognitive tests he performed satisfactorily.

As reading is a complex, mullable skill, a number of additional tests were administered in an attempt to identify the specific type of stimulation that provoked KM’s seizures. These tests extended over the longest period necessary to evoke a seizure by reading alone and included the following: (1) eye movements as if reading; (2) letter cancellation tests (Willison, personal communication); (3) silhouette cancellation tests: as in (2) but instead of letters small size silhouettes of objects had to be cancelled out; (4) singing and humming; (5) written calculations from The Graded Difficulty Arithmetic Test; (6) reading aloud long series of numbers typed as text and in single or double space; (7) scanning but not reading coloured magazines; (8) reading a list of nonsense words; (9) reading a passage comprising of nonsense words; (10) reading long lists of regularly and irregularly spelled words; (11) reading relatively short passages from children’s stories; (12) reading short news items from the popular press; (13) writing to dictation a long and complex technical text. None of these tasks appeared to provoke jaw jerking and the patient confirmed that he had felt comfortable throughout.

In contrast, the following experiments did elicit observable jerking of the right side of the jaw: (1) fairly lengthy news items (about 200 words) from the popular press; (2) reading shorter and relatively complex news items (typed and hand written) from a “quality” newspaper (150 words); (3) reading short technical passages from the Neuropsychologia journal and Brain’s Neurology (less than 100 words). Jerking was observed in all instances regardless of whether KM read aloud, subvocally, monocularly or through “reading masks” which are known to reduce pattern sensitivity. On the other hand, instructions to the patient to memorise a complex technical passage (200 words) resulted in the elimination of seizures after its third consecutive reading.

The above preliminary investigations were followed up by further experiments. Two types of reading material of differing complexity were typed on A4 white sheets. There
were six “easy” and six “difficult” passages, each consisting of 200 words. The easy passages were obtained from a Children’s Encyclopaedia and the difficult ones from Neuro-psychology (three passages) and human physiology textbooks (three passages). Easy and difficult texts were presented in a random order with 1 minute intervals between the individual passages. The patient was instructed to read them aloud and to attempt to understand their content as much as possible. Throughout the session an EEG was taken and KM was closely observed by the author who recorded the jerks as they occurred. Subsequently the number of paroxysmal abnormalities on the EEG was independently counted by a consultant neurophysiologist (WAK) who scored the record “blind”. There was a statistically significant correlation between these two types of scoring (Spearman ρ = 0·82, p < 0·01), albeit the observational method proved somewhat less sensitive. There was no significant difference between the mean number of jerks provoked by the two kinds of difficult text. Between reading the 6th and 7th passages, KM was asked to read an emotional text with sexual content. The number of jerks provoked by this kind of material was approximately halfway between the mean numbers of jerks provoked by easy and difficult texts (table).

KM was relatively fluent in Russian and was able to understand a text of moderate complexity. He was instructed to read aloud two 200-word passages in Russian, one easy and one difficult. On this occasion no EEG was recorded. The difficult text was read first and it was taken from a scientific paper of substantial complexity. The easy passage was from a textbook for learners of the Russian language. Between reading these two passages, KM was asked to read a 200-word passage in Greek which was copied from a newspaper using small size capital letters. There is a substantial overlap between the Greek and Russian alphabets and the patient was instructed to read the Greek text ignoring the letters which were unfamiliar to him. KM could not understand Greek at all. There showed only limited seizures with the easy Russian passage (2 jerks) and an increased incidence of seizures with the difficult passage (6 jerks). The Greek text did not provoke any jerks.

**Discussion**

In this case the diagnosis of primary reading epilepsy is supported by a variety of data. In so far as KM was unaffected by scanning different types of material or naming digits or observing stripes of epileptogenic frequencies, his jaw jerking could not be explained by pattern sensitivity. Also, in contrast to the way most patients with pattern epilepsy respond, KM continued to have seizures when viewing text monocularly or through reading masks. On the other hand, proprioceptive input from the muscles of the eye and mouth or throat did not appear to induce epileptiform activity. Finally, other more general factors such as high levels of concentration during rather laborious cognitive tasks were irrelevant to his seizures.

However, reading per se was not a sufficient trigger. For example, reading nonlexical material (nonsense words/text) or other verbal material devoid of meaning (words in isolation) did not induce seizures. In contrast, reading meaningful material over a relatively short length of time consistently produced seizures. It was also found that the greater the complexity of the text, the higher the frequency of the seizures. Reducing the complexity of text by re-reading the same passage extinguished jaw jerking. These observations leave little doubt that comprehension associated with reading was the necessary and sufficient factor for the provocation of seizures. Additional evidence for this view was provided by the absence of jaw jerking during reading in a foreign language he did not understand; by contrast, jaw jerking was observed during reading in a foreign language which he understood.

More recent models of reading have distinguished between at least two separate processes in reading. One has close links with semantic knowledge or comprehension (lexical/semantic) the other works by some sort of print-sound conversion system at a more mechanical subword level (phonological). Whereas Stella et al documented a primary reading epilepsy case in which phonological reading appeared to be the effective stimulus, for KM only lexical reading triggered primary reading epilepsy.

Both Forster and Ramani reported that the seizures in their patients were associated with comprehension during reading. On the surface KM appears to be very similar to their cases. However, there are some important differences amongst these different cases. Forster reported that seizures were evoked in almost all his patients (nine out of 11) when reading in foreign languages which they failed to understand. Unlike KM, 6/11 of Forster’s patients also had seizures when reading digits. These discrepancies may be explained by the possibility that comprehension was not the only trigger in the cases reported by Forster. Unlike the nine cases described by Forster and the case reported by Ramani, in the present case there

**Table**  Mean number of jerks as a function of the complexity of text

<table>
<thead>
<tr>
<th>Paroxysmal abnormalities on EEG</th>
<th>Observed jerking of the jaw</th>
</tr>
</thead>
<tbody>
<tr>
<td>Easy text</td>
<td>Difficult text</td>
</tr>
<tr>
<td>15</td>
<td>83</td>
</tr>
</tbody>
</table>
was no known family history of epilepsy. Although a number of authors have proposed a dominant gene mode of inheritance in reading epilepsy\(^{10,11}\) other authors have argued that a positive family history is rather infrequent.\(^{12}\)

Another difference between the present case and those reported by Forster and Ramani concerns the EEG patterns. During reading Ramani's patient showed discharges which were consistently lateralised to the dominant hemisphere. In the large majority of the cases reported by Forster there were similar abnormalities with a preponderance of focal EEG dysrhythmias in the fronto-temporal region. In KM the discharges were generalised and affected both hemispheres. Geschwind and Sherwin\(^{13}\) described a case of a patient who also showed bilateral generalised EEG dysrhythmias albeit their patient suffered seizures not only when reading but also during speaking and writing.

Despite striking similarities amongst the reported primary reading epilepsy cases in which seizures appear to be triggered by complex higher cerebral functions, nevertheless they comprise a heterogeneous group. In view of the high specificity which is seen in reflex epilepsy this conclusion may not be surprising and only future research can provide further clarification.

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