SHORT REPORT

Selective epileptic gait disorder

B G R Neville, S G Boyd

Abstract

Two children with an unusual gait disorder, one combined with acquired aphasia, in association with focal epilepsy are reported. Both children also showed paroxysmal "dystonic" phenomena, and a clear therapeutic response to corticosteroids. This newly described condition widens the range of discrete, recoverable defects of cerebral function that are associated with epilepsy in the developing nervous system and suggests that the site of action is at a functionally combined bilateral motor/sensory level of the cerebral cortex.

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The Landau-Kleffner syndrome comprises selective acquired loss of speech and language comprehension in childhood in association with epilepsy.^{1,2} Although antiepileptic drugs are often ineffective, good results from the early use of corticosteroids and surgical treatment by multiple subpial transection have been reported.^{3,4}

Patients**CASE 1**

This child was normal until 5.6 years old when partial seizures began. She reported nausea, said "here's another one", and held her hand beneath her chin to catch saliva; she stared, her tongue quivered, and after 30-60 seconds she recovered but with disordered speech—for example, "Mummy I can't proper talk". As the attacks increased in frequency to a maximum of several attacks an hour speaking became more difficult and attempts to speak seemed to provoke tongue jerking. After three weeks her speech was limited to "yes" and "no", but considerable understanding was preserved. By eight weeks she had no speech or demonstrable comprehension but she later reported understanding much of what was said but being unable to respond. Right sided motor seizures continued at a lower rate and were associated with transient right limb disuse and a curious posture of the right arm for three days (the arm

was held above the head with a flexed elbow, and the hand partly open). Sometimes the right foot would collapse when put to the ground. Her gait deteriorated, becoming slow, broad based, and high stepping, and she was unable to run. There were no other neurological signs. She also briefly had problems with moving food in her mouth and in recognising that her bladder was full. At 5.9 years she was highly distractible with total lack of language functions but with preservation of non-verbal/performance skills. High dose prednisolone was followed by recovery of all motor and most language skills by six weeks, with non-fluent aphasia at one stage. Early EEG showed a left frontotemporal slow wave abnormality with sharp components in the left posterior temporal region. An attempted sleep record only recorded during drowsiness and showed repetitive frontal sharp waves, more left than right. Later EEGs showed slow asymmetric rhythmic activity on eye closure and some continuing posterior temporal sharp waves after recovery. Magnetic resonance imaging was normal.

CASE 2

This girl could pull herself to a standing position at 8 months, and first tried, unsuccessfully, to walk at 11 months. From 6 months of age showers of myoclonic jerks of her left leg and occasionally of the right appeared 10 to 30 times daily. On attempting to walk sometimes her right leg and big toe extended with jerky inversion of the left foot and flexion of the left knee. These attacks resembled those of paroxysmal kinesogenic choreoathetosis,⁵ but they were unresponsive to carbamazepine. An interictal EEG in the waking state showed irregular spikes around the vertex (figure (A)), and tactile stimulation of the fingers and feet occasionally provoked runs of spike wave complexes at around 3-4/s, focal at the vertex (figure (B)). These were not associated with clinical signs. Similar changes were seen around the midparietal region, but these were always of lower amplitude. Clinical attacks, consisting of the left leg being held in flexion and shaking were associated with the child putting her foot to the floor, especially if this had a slapping quality. These attacks were associated with similar runs of discharges around the vertex (figure (C)). An EEG during sleep did not show any additional abnormalities. She walked independently at 23

Neurosciences Unit,
Institute of Child
Health and Great
Ormond Street
Hospital for Children
NHS Trust, The
Wolfson Centre,
Mecklenburgh Square,
London WC1N 2AP
B G R Neville
S G Boyd

Correspondence to:
Professor B G R Neville,
Neurosciences Unit, Institute
of Child Health, The
Wolfson Centre,
Mecklenburgh Square,
London WC1N 2AP, UK.

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(A) Small spikes mixed with slow components, seen spontaneously around the vertex. (B) Tapping of the fingers of the left hand (arrow) sometimes evoked a run of spikes and slow waves, again maximal at the vertex, in addition to a larger (150 mV) spike in the centroparietal region. (C) A typical clinical attack consisting of flexion and myoclonic jerking of the leg was again associated with a run of spikes and slow components.

months with corticosteroids given for asthma. At 25 months her gait was stiff legged and broad based, her feet were slapped down, she tended to back-knee, and occasional sudden collapses occurred. The gait became virtually normal with a further course of prednisolone. Neurological examination was otherwise normal as was general development. Magnetic resonance imaging was normal.

Discussion

These children showed a stable gait disorder that might be clinically described as apraxia in association with partial epileptic seizures. The apraxia was isolated in one and in the other combined with dense acquired aphasia suggesting a close analogy of cortical cognitive disturbances between the Landau-Kleffner syndrome and this type of motor disorder, and that both may also be responsive to corticosteroids. Transient changes of hand dominance,⁶ as seen in patient 1, and hand praxis have been reported in Landau-Kleffner syndrome,⁷ and also bulbar apraxia in association with benign Rolandic epilepsy.^{8,9} These highly selective deficits are rare but motor and cognitive regression attributed to coincident non-convulsive status occur in a number of childhood epilepsy syndromes, and may also be partially responsive to corticosteroids.

The combined and variable receptive and expressive language problems previously reported in Landau-Kleffner syndrome¹⁰ are well illustrated in patient 1. When epilepsy occurs it may combine focal motor and sensory shock like phenomena in a common distribution.¹¹ The "apraxia" reported here is also not separable into motor and sensory elements. The simplest explanation for these

deficits would be that the cortical system that is disrupted combines receptive and expressive elements. There is an obvious bilaterality to the pervasive deficits—that is, the gait disorder and dense aphasia—with no early tendency for relocation to the non-dominant hemisphere and often with a bilateral EEG abnormality, in the presence of focal epilepsy. Thus the functional unit that seems to be disrupted is bilateral, homologous, cortical, motor, and sensory.

The "dystonic" phenomena, although so designated by experienced observers, do not conform to the typical mobile spasms of classic dystonia, and in this context of two children with additional clear cortical phenomena, may represent cortical disconnection rather than a positive basal ganglia event. The attacks of patient 2 resembled those of familial paroxysmal kinesogenic choreoathetosis, particularly in the motor intentional provocation.¹² This phenomenon of cognitive intent, that seemed to provoke a seizure in both children, also points to a tight link between motor, sensory, and cognitive control at the cortical level.

The relation of paroxysmal dystonia to epilepsy has been debated.⁵ Epileptic dystonic postures are well described. For example, the bulbar tonic phenomena, including stiffening and twisting of the tongue with speech arrest described in benign Rolandic epilepsy¹³ closely resemble those seen in some cases of familial paroxysmal choreoathetosis (for example, case 3 of Lance⁵). Both conditions are characterised by normal imaging and the absence of neurological deficit. In both, sensory symptoms closely related to the motor site are reported—for example, tingling and prickling of the face, mouth, and tongue in benign Rolandic epilepsy—again suggesting a

common functional motor/sensory unit disturbance. Similarities between benign Rolandic epilepsy and Landau-Kleffner syndrome have been proposed.¹⁴ In this context the issue of brief cognitive arrest in some partial epilepsies also deserves further study.¹⁵

The phenomena designated "epileptic negative myoclonus", although associated with a gait disturbance, differs from the phenotype of patient 1 by the lack of dystonia, the range of types of seizures, and the ages of these patients.¹⁶ Although the EEG responses to foot tapping might suggest benign partial epilepsy with extreme somatosensory evoked potentials, the legs are characteristically spared both in paroxysmal and pervasive clinical effects.¹⁷

These two children broaden the range of acquired discrete higher cortical deficits that coexist with focal epilepsy. Perhaps more important is the suggestion that there is a group of disorders that occur in the immature but structurally normal nervous system that includes Landau-Kleffner syndrome, benign Rolandic epilepsy, and some paroxysmal dystonias. These would affect a bilateral joint sensory/motor functional cerebral cortical system and produce a range of phenomena such as selective cognitive deficits, isofocal sensory/motor phenomena including epilepsy, and paroxysmal dystonia, which are often provoked by motor intention or stimulus. It is possible that the selective functional cortical disconnection may occur as a deviant response to sensory or intentional input, and that corticosteroids and perhaps multiple subpial transections may be effective treatments.

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