

Clinical Reports

Guillain-Barré syndrome mistaken for brain stem death

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Summary: A case of acute severe polyneuropathy in a previously healthy 45 year old male is presented. At the height of his illness the clinical picture was mistakenly interpreted as brain death. Spontaneous recovery occurred.

Introduction

The Guillain-Barré syndrome may present with a wide range of clinical pictures. Typically there is a rapid onset of progressive, ascending paralysis with loss of deep tendon reflexes and usually some associated paraesthesiae.¹ Many variants of the typical syndrome are recognized.² The severity of the condition also varies, and sporadic reports have described the occurrence of an acute severe inflammatory polyneuropathy leading to a complete 'locked-in' syndrome.^{3,4}

The present report describes a case of locked-in syndrome due to an acute inflammatory polyneuropathy, which, but for the preceding history, might have been misdiagnosed as an acute and possibly fatal brain stem event. During the worst phase of his illness the patient was mistakenly thought to be brain dead.

Case report

A previously fit 45 year old man presented to a district general hospital with a 2 day history of diplopia and increasing weakness of all four limbs. Ten days previously he had suffered a mild gastrointestinal upset.

Initial examination showed bilateral lateral rectus palsies with distal upper limb weakness, proximal lower limb weakness and areflexia. Over the next 24 hours the limb weakness became global and more marked, and he required ventilation for respiratory failure. At this stage he was able to respond appropriately to commands despite mild sedation. Routine blood tests were unremarkable. A lumbar puncture showed sterile, acellular cere-

brospinal fluid, with a protein content of 0.25 g/l (normal range 0.15–0.45 g/l). Screening tests for polio and porphyria were negative. Nerve conduction studies were not performed.

His clinical condition worsened, developing a complete external ophthalmoplegia with loss of all reflex ocular movements, loss of pupillary and corneal reflexes, absent gag reflex and total paralysis of all four limbs. There was concern that he had suffered a superimposed vascular brain stem event.

A neurological opinion was sought and he was transferred to the regional neurosciences unit. The initial clinical impression on arrival was that the patient appeared to be brain dead, though the preceding clinical history and the lack of a positive diagnosis precluded formal testing for the absence of brain stem function.

An electroencephalogram (EEG) was performed which showed a normal dominant alpha rhythm, wholly unresponsive to stimulation. The neurophysiological opinion was that the distribution and lack of responsiveness suggested a brain stem lesion leading to 'alpha coma', i.e. a reactive alpha rhythm seen on EEG which was consistent with consciousness, recorded from a paralysed patient who appeared clinically to be comatose. Computerized tomographic and magnetic resonance imaging scans of the cerebral hemispheres, mid brain and brain stem were normal. A repeat lumbar puncture gave unchanged results from the first.

A second EEG was performed some 48 hours after the initial recording. This showed a marked change, with arousal phenomena being induced easily with posterior rhythmical activity appearing at 7–8 Hz. The first clinical sign of improvement was a subtle movement of the eyes upwards and inwards on calling the patient's name and instructing him to close his eyes, i.e. a weakly positive 'Bell's phenomenon'. Over the next two weeks his ophthalmoplegia and facial weakness resolved.

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This enabled communication to be established using eye movements. His pupillary and corneal reflexes returned during this time. He continued to require assisted ventilation for 6 months after the onset of his illness.

Rehabilitation has now been transferred to his home, and progress seems to have reached a plateau. Fourteen months after the illness he is still wheelchair bound with minimal power in his arms and residual paralysis of his legs.

Discussion

The striking and alarming feature of this case was the initial clinical impression that the patient was brain dead at the time of transfer to the regional neurosciences unit. The criteria for the diagnosis of brain death produced by the United Kingdom Medical Royal Colleges comprise a set of preconditions which must be satisfied before formal tests for the absence of brain stem function may be performed.^{5,6} These preconditions dictate that a clear cause of irremediable brain damage has been identified, and a set of necessary exclusions specifically ensures that reversible causes of brain stem depression such as depressant drugs, neuromuscular blocking agents, hypothermia and metabolic imbalance have been excluded.^{7,8}

Clearly, in the present case, these preconditions could *not* be fulfilled as a consequence of the preceding history. However, at the height of his illness, neurological examination of the patient showed little to suggest an intact brain stem, though caloric testing, and examination for respiratory movements following disconnection from the ventilator were not deemed appropriate tests in view of the history. In the present case, the observed EEG pattern and its subsequent change abolished any concern about brain death.

Few reports of a complete locked-in phenomenon attributable to a post-infective cranial and peripheral polyneuropathy have appeared.^{3,4,9} The

majority of descriptions of the locked-in syndrome relate to patients who have preserved consciousness in the face of quadriplegia and paralysis of all lower motor cranial nerves, usually due to a destructive lesion in the ventral pons.¹⁰ One previous report has documented a total locked-in state due to polyneuropathy described as resembling brain death.⁴

In the present case deterioration was rapid with assisted ventilation starting soon after admission. Although the antecedent history and the initial pre-hospital deterioration made a progressive polyneuropathy the likely diagnosis, the differential diagnosis of an acute brain stem syndrome of central origin and therefore poor prognosis¹⁰ was considered due to the rapidity of the patient's deterioration and need for assisted ventilation.

Other reported examples of such an extreme polyneuropathy^{3,4} also showed a rapid progression to a locked-in state which improved gradually. Electroencephalographic patterns showed a similar unresponsive alpha rhythm in the acute state. The present case, along with those of Carroll *et al.*³ and Kotsoris *et al.*,⁴ is distinct in illustrating an extreme polyneuropathy to the point of being able to masquerade as brain stem death. EEGs in all three cases were consistent with previous descriptions of alpha coma.¹¹

This is an important, although relatively rare, variant of Guillain-Barré syndrome to consider for a number of reasons. Firstly the condition is reversible although undoubtedly associated with a significant morbidity.¹² Secondly, misdiagnosis without a good antecedent history is potentially easy and carries disastrous consequences. Thirdly, the present case illustrates the importance of electroencephalography in this group of brain stem syndromes without obvious cause.

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