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CASE REPORT

Guillian–Barré syndrome in high tetraplegia following acute respiratory illness

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Study design: A case report of a Guillain–Barré syndrome (GBS) variant presenting in a patient with a high cervical spinal cord injury (SCI).
Objectives: To illustrate a clinical presentation of GBS in an individual with chronic SCI.
Setting: Vancouver General Hospital, Vancouver, BC, Canada.
Methods/Results: A 31-year-old man with chronic C2 AIS B (American Spinal Injury Association Impairment Scale) SCI and diaphragmatic pacing presented with respiratory failure with sepsis. His sepsis resolved with antibiotic therapy, but he continued to have autonomic instability and was unable to be weaned off his ventilator. Concurrently he developed flaccidity and facial diplegia. Investigations including nerve conduction studies and cerebrospinal fluid analysis prompted a diagnosis of acute motor-sensory axonal neuropathy, a variant of Guillian–Barré syndrome. Owing to ongoing autonomic instability, he was treated with intravenous immunoglobulin. His autonomic dysfunction resolved and he regained some facial muscle function, but 6 months post injury he remained dysphagic and required 24-h ventilator support.
Conclusion: Careful neurological reassessment prompted the diagnosis of acute polyradiculoneuropathy following respiratory sepsis as the root cause of diaphragmatic pacer failure and autonomic instability.
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Keywords: diaphragmatic pacer; respiratory failure; Guillian–Barré syndrome; cervical spinal cord

Introduction

Guillain–Barré syndrome (GBS) is an acute inflammatory polyradiculoneuropathy. The hallmark features of GBS are bilateral, symmetric and ascending weakness with reduced deep tendon reflexes. GBS can affect the phrenic nerve and cause respiratory dysfunction. Autonomic involvement is common, ranging from blood pressure lability to urinary retention to cardiac arrhythmias.[1]

We report an interesting case of GBS presenting in a patient with a high spinal cord injury (SCI). The hallmark features of bilateral, symmetric and ascending weakness were obscured by pre-existing complete motor paralysis. Complications of autonomic instability and respiratory failure that frequently present in GBS were more diagnostically challenging, as this patient was ventilated at the time, was dependent on a diaphragmatic pacer and had pre-existing autonomic dysreflexia.

Case report

At age 26, this patient sustained a complete motor cervical SCI (C2 AIS B).[2] After extended inpatient rehabilitation he was discharged home. On discharge, he required 24-h ventilation but later had a diaphragmatic pacemaker implanted. He lived independently and managed well in the community, enjoyed adapted sailing, travelled internationally and accepted public speaking engagements.

Five years after his SCI, he was found by a care aid at home to be unresponsive and febrile. He was brought to the hospital, diagnosed with respiratory sepsis and admitted to the intensive care unit.

With treatment his sepsis resolved, but a week later he continued to fail attempts to wean him off his ventilator. He also developed autonomic instability with episodes of paroxysmal tachycardia, arrhythmias and blood pressure lability. Although not initially recognized in the intensive

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care unit setting, his paralysis changed from a spastic to a flaccid pattern. He became areflexic. He developed bilateral facial diplegia, gaze-evoked nystagmus and lower motor neuron weakness of his tongue.

Neuroimaging and cerebrospinal fluid (CSF) studies completed on admission to hospital were repeated. Computed tomography imaging of his head was unchanged from scans at admission. CSF analysis at admission was mildly abnormal, with elevated white cell count (31, normal <5) and moderately elevated protein level ($948\,mg\,l^{-1}$, normal-$400\,mg\,l^{-1}$). His repeat CSF, however, showed marked albuminocytological dissociation, with a white cell count of 1 and protein level of $6835\,mg\,l^{-1}$. A paraspinal abscess was considered given the high protein level, but was ruled out owing to contrast images being obtained on computed tomography of the spine. His diaphragmatic pacing leads precluded investigation by magnetic resonance imaging.

Electrodiagnostic studies showed decreased compound motor action potential amplitudes with preserved velocities in the nerves that could be tested; however, most sensory and motor nerve conduction responses as well as F waves were unobtainable. Needle electromyographic studies revealed fibrillations and positive sharp waves, in keeping with active denervation.

The patient was diagnosed with acute motor-sensory axonal neuropathy, a variant of GBS. Critical illness polyneuropathy could not be completely excluded given that his neuropathy was mainly axonal; however, marked albuminocytological dissociation and profound facial diplegia supported the diagnosis of GBS.[3] He was treated with intravenous immunoglobulin $2\,g\,kg^{-1}\,day^{-1}$ for 3 days and his autonomic instability resolved. He recovered some facial muscle function but remained dysphagic.

Six months later, the patient remained in hospital with incomplete recovery. Although there was improvement in facial muscle strength, he still could not fully close his mouth or completely manage his oral secretions. A percutaneous endoscopic gastrostomy tube was placed to facilitate feeding. Owing to increased care needs, he was discharged to an extended care facility. Today, he remains on a ventilator and has periodic diaphragmatic pacer trials.

Discussion

As GBS progresses, the diagnosis is often relatively straightforward. This case is interesting because many of the hallmark signs and symptoms that lead a clinician to consider GBS, such as sensory paresthesia with ascending weakness, were obscured by his SCI. The patient had preexisting tetraplegia, respiratory failure and autonomic dysfunction. A change from spastic to flaccid paralysis, although not a subtle sign, is easily missed in an intensive care unit setting. The onset of facial diplegia was an unmistakable sign that a process other than chronic SCI with sepsis was taking place.

Conclusion

This case highlights the importance of early neurological assessment of chronic SCI patients in an acute setting. It illustrates how, in this population, an otherwise straightforward diagnosis can be quite nuanced. Facial weakness along with changes in tone, reflexes and spasticity were the key in understanding why this patient could not be weaned off his ventilator. More importantly, these signs led to an understanding of the cause and the necessity for a subsequent therapeutic intervention for potentially life-threatening autonomic instability.

Conflict of interest

The authors declare no conflict of interest.

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