Case report

Acute primary axonal Guillain-Barré syndrome

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Introduction

Classical Guillain-Barré syndrome (GBS) is an acute inflammatory demyelinating polyradicular neuropathy, with secondary axonal degeneration occuring only in more severe cases. In 1986 Feasby and others described 5 patients with neurophysiologically and histologically proven primarily axonal form of Guillain-Barré syndrome with normal or near normal myelin sheaths (1). The clinical presentation and other findings in these cases were identical to classical GBS but they had a poorer prognosis.

We describe two such cases of neurophysiologically and electron microscopically proven primary axonal GBS.

Case reports

Case 1

A 62-year old man, previously in good health, was admitted with weakness and numbness of all 4 limbs. His symptoms progressed rapidly over the next 3 to 4 days making him quadriplegic. There was no preceding history of viral infection or vaccinations. His vital capacity dropped and he was ventilated. Examination showed grade 0 power in all 4 limbs with global areflexia. Bilateral ptosis and weakness of the external ocular muscles were noted. His blood pressure fluctuated, with episodes of hypotension probably due to autonomic instability. Some sensory impairment was noted in the limb extremities.

None of the motor nerves were excitable at nerve conduction studies (NCS) even with supra-maximal stimulation. Compound motor action potentials (C-MAPs) were unrecordable with both proximal and distal stimulation. EMG was compatible with a denervation pattern. CSF analysis showed an elevated protein level (60 mg/dl) with no cells and negative results for both bacterial culture and viral studies. CSF for PCR-TB was also negative. Urine for porphyrins and HIV were negative. He developed investigatively proven severe syndrome of inappropriate ADH secretion (SIADH) with hyponatremia, a known complication of GBS. His liver and renal function were normal. CT head scan with brain stem cuts was also normal. A sural nerve biopsy was performed and the electron microscopic analysis revealed severely degenerated axons with normal myelin sheaths and no significant inflammatory response. The periaxonal space was occupied by macrophages (Figure). These pathological changes on electron microscopy are identical to the cases described earlier (1).

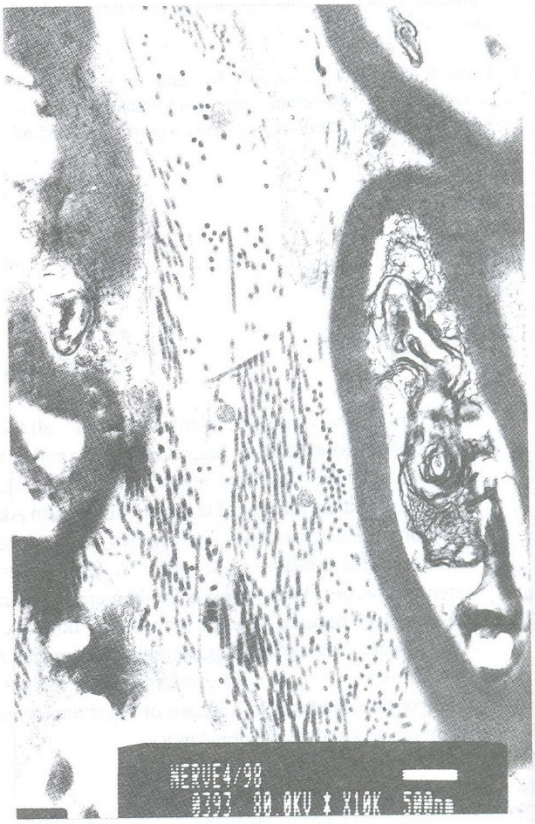

Figure. *Electron microscopic appearance of the sural nerve biopsy in case 1 showing a normal myelin sheath with severely degenerated axons. There is no inflammatory cellular reaction.*

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He was treated with repeated plasma exchanges, human immunoglobulin coupled with methyl prednisolone. His condition did not improve and the limb weakness remained unchanged. After a 4-month stay in the intensive care unit he succumbed to his illness. A biopsy of the ulnar nerve at post-mortem revealed the same picture as the sural nerve on electron microscopy.

Case 2

A 75-year old man was admitted with a 2-day history of weakness of all 4 limbs. His weakness worsened over the next few days making him quadriplegic. He had no preceding viral infection or vaccinations. He too was ventilated. He has grade 0 power in all 4 limbs, with global areflexia. His external ocular movements were normal and he had moderate facial weakness. Other cranial nerves were normal. Sensory system assessment was difficult. His NCS and EMG findings were identical to case 1. CSF studies showed cytoprotein dissociation. He too had a mild degree of SIADH. He also developed autonomic instability with episodes of hypotension. Other investigations including liver and renal function, HIV screening, viral antibody screening were normal. Sural nerve biopsy revealed severe axonal degeneration with minimal distortion in the myelin sheath on electron microscopy. There was no significant inflammatory response. This patient was treated with repeated plasma exchanges but he too succumbed to his illness after 2 months of intensive care.

Discussion

GBS is regarded as an immune mediated inflammatory demyelination affecting the myelin sheaths of peripheral nerves (2,3,4). The five cases described in 1986 had 4 major distinguishing features (1): inexcitable motor nerves and unrecordable C-MAPs with high voltage long duration electrical stimulation, reaching peak severity earlier (average 7.2 days versus 17 days in classical cases), comparatively poor prognosis, and severe axonal degeneration with no inflammation and with normal or near normal myelin sheath on histology.

Both our patients had primary axonal disease with normal myelin sheaths, reached peak severity with quadriplegia within 5 days and had to be ventilated. SIADH and autonomic instability developed in both. Both did not have antecedent viral infections. This condition is now termed acute motor and sensory axonal neuropathy (AMSAN), a primary axonal pathological variant of GBS. In more recent reports from China an acute motor axonal neuropathy (AMAN), similar in presentation to GBS, but less severe than AMSAN, is described with primary axonal degeneration but involving only the motor fibres (5,6,7). Hence, over the last 10 years it has become accepted that GBS includes at least 3 separate pathological entities (8):

(i) Acute inflammatory demyelinating polyneuropathy (AIDP)

(ii) Acute motor sensory axonal neuropathy (AMSAN)

(iii) Acute motor axonal neuropathy (AMAN)

The two cases we describe here are of the AMSAN type and both patients had a severe clinical course similar to the cases described in the western world. Few similar cases with a bad prognosis are also reported from India (9).

GBS carries a good prognosis with 75% patients making a complete recovery within 6 months (10). Mortality is higher for primary axonal AMSAN cases (10). A history of diarhoea with *Campylobacter jejuni* infection has been found to be more common in primary axonal cases of GBS. Stool culture for this organism was negative in both our cases.

Acknowledgements

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Measuring clinical outcome in asthma

In the last week/month

1. Have you had difficulty sleeping because of your asthma symptoms (including cough)?
2. Have you had your usual symptoms during the day (cough, wheeze, chest tightness, or breathlessness)?
3. Has your asthma interfered with your usual activities (eg housework, work/school etc)?

Each of the above questions should be answerable by a simple yes/no, but there could also be supplementary quantitative grades for answers.

The three questions above should be asked, and the answers recorded, at every asthma consultation both in primary and secondary care.

Pearson M, Bucknall C: eds Measuring clinical outcome in asthma: a patient focused approach. London: Royal College of Physicians, July 1999.