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Clinical Quiz

A patient with Guillain-Barré syndrome and late recovery after 1 year

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Case

A 39 year old male, presented to our department with rapid onset flaccid paralysis (upper extremity muscle weakness that was followed by lower extremity paralysis within hours). No sensory disturbances, respiratory failure or cardiovascular dysfunction was noted. 9 days before admission, the patient reported high fever with acute non bloody diarrhea. Past medical history was negative for comorbidities.

Motor strength upon presentation was more severely affected in lower than upper extremities: deltoid/ elbow strength 3/5, wrist strength 1/5, hand strength 1/5 (extension) and 2/5 (flexion), iliopsoas 1/5, maximus gluteus 1/5, quadriceps 1/5, foot dorsiflexion/ plantarflexion 1/5.

Electroconductive studies along with cerebrospinal fluid paracentesis were performed; the laboratory tests along with the clinical presentation led to the diagnosis of post-gastroenteritis Guillain-Barré syndrome (GBS).

Since the patient was nonambulatory within 4 weeks of symptoms presentation, treatment with plasmapheresis (5 sessions) and intravenous immune globulin (2 sessions) was undertaken. Thromboprophylaxis with compression stockings and low molecular weight heparin was simultaneously initiated.

Rehabilitation program was aggressively initiated that consisted of physiotherapy, hydrotherapy, psychological support and proper nutrition. An individualized program with gait training, strengthening exercises involving isometric, isotonic, isokinetic, and resistive exercises was fostered. Rehabilitation program also focused on proper limb positioning, posture, contracture prevention and utilization of orthotics. Occupational therapist was also employed to improve activities of daily living (ADLs).

Electroconductive studies were repeated 9 months upon therapy initiation that showed axonal degeneration in ulnar nerve and severe axonal degeneration in L2, L3, L4, L5 and S1 nerve roots, especially in the right side. However, there was evidence of muscle regeneration on the left lower extremity. Dual energy xray absorptiometry (DXA scan) was twice performed during treatment course in left hip and lumbar spine. On both occasions T-score values were within normal limits.

The patient did little improvement until 14 months post treatment; however, thereafter an impressive motor recovery was noted; Barthel index (a 0-100 scale with ten categories of ADLs) was recorded at various points during treatment course. In the beginning the Barthel index was 20 and upon discharge a 60 score was recorded, essentially reflecting the improvement after 1 year of therapy. Motor strength was restored to 4/5 in shoulder/ elbow area, 3/5 in the wrist and 3+/5 in finger flexion/ extension. In the lower limbs a similar improvement was noted: iliopsoas 2/5, gluteus max. 3/5, quadriceps 3/5, foot plantarflexion 3/5, and foot dorsiflexion 2/5 (similar values in both extremities).

Commentary

GBS is an acute demyelinating inflammatory polyradiculoneuropathy, named after the authors who first described it. Acute inflammatory demyelinating polyneuropathy (AIDP) is the typical and first recognized entity; a disease variant (acute motor axonal neuropathy-AMAN) with absence of demyelinating findings in

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electrophysiological studies was recognized in the 1990s in China, post campylobacter jejuni infection. However, we consider our case a typical AIDP with axonal degeneration. Although EMG and CSF exam may confirm the diagnosis, this still remains largely a descriptive one. Annual incidence is about 1-2 per 100.000 persons. The syndrome is frequently post infectious (especially after Campylobacter Jejuni enteritis) and the main symptoms are flaccid paralysis of the extremities with symmetrical weakness and sensory disturbances, usually within 2-4 weeks upon presentation. In the most severe cases, respiratory failure or cardiovascular complications may ensue, necessitating intubation and mechanical support in the ICU[1,2].

Electrodiagnostic studies are of prognostic significance and may aid the diagnosis: distal compound muscle action potential (CMAP) amplitude less than 20% of lower normal values and profuse fibrillation potentials on needle examination implies axonal degeneration and is considered to be a poor prognostic factor. Same applies for inexcitable nerves and severe conduction block of common peroneal nerve (more than 44%)[2,3].

Other dismal factors are rapid onset of symptoms, older age (more than 40 or 50 years), severe weakness on admission and/ or need for intubation and preceding infection with diarrhea[2]. In our case, there were some adverse factors such as older age (close to the cutoff of 39 years), diarrheal infectious disease, axonal degeneration EMG pattern, and significant weakness.

Treatment options include plasmapheresis and intravenous immune globulin. Although beyond the scope of this article, in general, both modalities yield equivalent results and they should be given early in the course of disease for nonambulatory patients, since they hasten recovery[2,4].

In conditions that lead to acute deterioration of mobility such as GBS or acute spinal cord injury thromboprophylaxis is strongly recommended as thromboembolic events are extremely common; time to develop deep venous thrombosis or pulmonary embolism varies from 4 to 67 days after presentation. Duration of anticoagulation, methods of thromboprophylaxis or monitoring of high-risk patients are less straightforward as clinical studies are missing and most of these patients remain immobile for long periods[4,5].

Regarding prognosis, most patients have disease progression for up to two weeks (8 days on average to disease nadir) and then reach a plateau for another two weeks before recovery starts. About 84% of patients will be fully ambulatory at 1 year. However, only 60% will achieve full motor strength at this point. On average, 5-15% of patients will have a protracted illness course with incomplete recovery and/ or significant musculoskeletal problems. Most of the improvement appears early in the course of the disease; it is estimated that up to 80% will be able to walk independently at 6 months with only modest improvement thereafter (as stated above this figure only rises to 84% at 1 year). There are unusual cases though that will report improvement up to 6 years after presentation[2]. Our case scenario is worth reporting since there was negligible progress during the first year and then against all odds (including electrodiagnostic studies showing axonal degeneration) astonishing recovery was noted, starting from the upper and few months later the lower extremities. It also highlights the fact that patients should not be discouraged neither by early therapy failure nor by axonal degeneration in electromyogram/nerve conduction studies. Late recovery is feasible with strict adherence to the rehabilitation protocol.

Approximately 40% will need inpatient rehabilitation program, since up to 20-54% will suffer from some sort of plegia (ranging from mono to tetraplegia). A multidisciplinary team of physical therapists, occupational therapists, speech-language pathologists, social workers, psychologists, nurses and other allied health professionals working under the supervision of a neurologist or rehabilitation physician should focus on improving ADLs. Although bibliographic data are missing it is most likely that intensive rehabilitation improves long-term symptoms. Physiotherapy interventions include strength, endurance and gait training with gradual increase in mobility, maintenance of posture and alignment as well as joint function. Along with muscle weakness, contractures and muscle shortening are encountered, that should be prevented with daily range of motion exercises and joint mobilization. Proper positioning is crucial and orthotics can aid in this effort and also maximize residual motor function. In terms of muscle weakness and wasting, a tailored program should be structured consisting of gentle strengthening exercises in the beginning (isometric, isotonic, isokinetic and manual-resistive), moving to progressive resistive exercises; however, overfatiguing the affected muscles should be avoided, since it has been shown to cause paradoxical weakening and negatively affect recovery[4,5].

Occupational therapy aims to improve everyday function with domestic and community tasks as well as driving and work. Home modifications, gait aids, orthotics and splints may be provided. Speech-language therapists may be required in those with speech and swallowing problems, as well as to support communication in those who require ongoing breathing support (often through a tracheostomy). Nutritional support may be provided by the team and by dietitians. Psychological counseling may also be required for anxiety, fear and depression[4,5].

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Questions

1. What is the 1-year outcome in GBS patients?
A. More than half of the patients will be nonambulatory.
B. No motor weakness is generally anticipated after 1 year.
C. Around 10% of patients will have more severe course with various sensory-motor sequela.
D. Half of the patients will die from disease progression.

Critique

Around 14% of patients will have severe disability 1 year after presentation, whereas 84% will be able to walk independently. Mortality rate is around 3-7% despite intensive care.
The correct answer is C.

2. What is the optimal treatment of GBS?
A. Only supportive care
B. Plasmapheresis
C. Intravenous Immune Globulin
D. B & C

Critique

Both modalities have yielded similar results and they hasten recovery especially when given early in the course of the disease. Preference over Plasma exchange therapy or iv immune globulin depends on local availability and should be individualized. Steroids are not recommended in general.
The correct answer is D.

3. Which are bad prognostic factors?
A. Young age (less than 40 years).
B. Upper respiratory preceding infection.
C. Severe weakness upon presentation and/ or need for mechanical ventilation.
D. Inexitable nerves and distal compound muscle action potential (CMAP) amplitude more than 20% of lower normal values in electrophysiology.

Critique

Except for severe presenting symptoms and need for ventilation, other risk factors are older age (after 40 or 50 years in other studies), preceding diarrheal infection with rapid onset of symptoms and CMAP less than 20% of normal values along with profuse fibrillation potentials that suggest axonal degeneration.
The correct answer is C.

4. When is recovery anticipated?
A. Mainly up to 6 months.
B. Between 6 months and 1 year.
C. After one year no further improvement is possible.
D. Up to 15 years post presentation.

Critique

Most patients (80%) recover during this timeframe. Another 4% will improve at 1 year. Few patients will continue to improve (up to 6 years in the literature) and therefore as also suggested by this article patients should not be overwhelmed after early treatment failure.
The correct answer is A.