

Rapidly Progressive Paralytic Syndrome: Review Questions

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QUESTIONS

Choose the single best answer for each question.

Questions 1 through 5 refer to the following case study.

A 37-year-old man comes to the emergency department for evaluation because of paresthesias in both feet radiating to the ankles. He reports that he woke up yesterday with a tingling sensation in both feet, had some unsteadiness in walking and a feeling of heaviness in his feet that afternoon, and noted a wobbly sensation in his knees that evening; he began to have dull, aching pain in the lower back without radiation but had no difficulty with bladder or bowel function. Earlier this morning, the patient had increased difficulty ambulating and required assistance to walk. He reports no speech difficulty, swallowing disturbance, vertigo, diplopia, or upper-extremity symptoms and no urinary hesitancy or incontinence.

The patient has no significant medical history. He had a viral upper respiratory infection 3 weeks ago. He does not take medication, smoke cigarettes, or use alcohol in excess. He reports no use of recreational drugs. Review of systems is otherwise unrevealing.

General physical examination is unremarkable, except for mild lower lumbar tenderness to percussion. Neurologic examination reveals normal mental status. Cranial nerve examination shows no abnormalities, but there is mild weakness of eye closure. Motor testing reveals grade 4/5 weakness of the intrinsic hand muscles (ie, dorsal interossei, flexor pollicis brevis, flexor digitorum profundi) bilaterally. Lower extremity strength is 3/5 bilaterally in the iliopsoas, quadriceps, hamstrings, tibialis anterior, and gastrocnemius. The patient has diminished sensation to vibration in the legs bilaterally, to the midcalves. His gait is paraparetic, and he requires assistance to take more than a few steps. Results of cerebellar testing involving the upper extremities are normal. Deep tendon reflexes are diffusely absent; plantar responses are flexor.

- 1. This patient's disorder is most likely localized to which of the following structures?**
 - A) Cerebellum
 - B) Muscle
 - C) Neuromuscular junction
 - D) Peripheral nerves
 - E) Spinal cord
- 2. Which of the following is the most likely diagnosis?**
 - A) Cerebellar hemorrhage
 - B) Guillain-Barré syndrome
 - C) Myasthenia gravis
 - D) Polymyositis
 - E) Transverse myelitis
- 3. Which of the following tests would be most useful to diagnose the patient's condition?**
 - A) Computed tomography scan of the brain
 - B) Magnetic resonance imaging of the spine
 - C) Lumbar puncture
 - D) Measurement of the serum creatine kinase level
 - E) Tensilon test
- 4. Which of the following parameters should be most closely monitored in this patient?**
 - A) Arterial blood gases
 - B) Blood pressure
 - C) Forced vital capacity
 - D) Mental status
 - E) Pupillary size and reactivity

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5. Administration of which of the following substances is most appropriate?

- A) Antibiotic agents
- B) Corticosteroids
- C) Immunoglobulin, intravenously
- D) Interferon beta
- E) Pyridostigmine

EXPLANATION OF ANSWERS

1. **(D) Peripheral nerves.** This patient has an acute sensory-motor syndrome affecting the legs and arms, with a distal predominance. Reflexes are diffusely absent, with loss of vibration in a stocking pattern. The presence of areflexia and a distally predominant sensorimotor deficit suggests a diffuse large-fiber polyneuropathy of motor and sensory nerves. The patient has no ataxia or nystagmus suggesting a cerebellar disorder. The presence of sensory symptoms and distally predominant weakness argue against a disorder of the neuromuscular junction or muscle. A spinal cord disorder would be expected to produce a sensory deficit to pain and temperature below a segmental spinal level and upper motor neuron signs (eg, hyperreflexia, Babinski's sign).
2. **(B) Guillain-Barré syndrome.** This patient has a rapidly progressing ascending motor disorder, characterized by weakness in the legs more than in the arms, mild facial weakness, areflexia, and vibration loss in a stocking distribution. An antecedent viral infection occurred 3 weeks ago. Acute inflammatory demyelinating polyneuropathy, also known as *Guillain-Barré syndrome* (GBS), is the most common cause of acute nontraumatic generalized paralysis. GBS has an annual incidence of 1.2 cases per 100,000 and affects every age group. A recent febrile illness can be identified in more than 60% of patients. The lower extremities are usually involved initially, followed by the upper extremities. More than 50% of patients with GBS have involvement of facial and other cranial muscles (eg, oropharyngeal, oculomotor). Although GBS primarily involves motor nerves, sensory symptoms and signs (eg, distal paresthesias, mild vibration, proprioception deficits) are not uncommon.
3. **(C) Lumbar puncture.** Most patients with GBS develop an abnormal spinal fluid profile within the first week of illness, with elevated cerebrospinal fluid protein in the absence of pleocytosis (albuminocytologic dissociation). Patients with GBS who have a lymphocytic pleocytosis should be evaluated for coexistent HIV infection, Lyme disease, or sarcoidosis. Electro-

myography and nerve conduction studies are useful to demonstrate the presence of a demyelinating polyneuropathy with reduced conduction velocities and temporally dispersed compound muscle action potentials. These abnormalities may be delayed for 10 to 14 days; however, prolonged long-latency F waves, a measure of spinal root integrity, may be found early in the disease course. Neuroimaging of the brain or spine is not usually warranted in patients with GBS. Serum creatine kinase and Tensilon (edrophonium) testing are useful in the diagnostic evaluation of myopathy and myasthenia gravis, respectively.

4. **(C) Forced vital capacity.** Patients with GBS or suspected GBS are best monitored in the intensive care unit. Approximately one third of patients with GBS develop respiratory muscle weakness requiring mechanical ventilation. The most reliable parameters of neuromuscular respiratory muscle (ventilatory) weakness are forced vital capacity (FVC) and negative inspiratory pressure. Intubation should be strongly considered if the FVC decreases to below 12 to 15 mL/kg body weight. Arterial blood gases and pulse oximetry are important but less sensitive measures of ventilation. The pupillary reflex is not affected by GBS, although autonomic nerve involvement may result in labile blood pressure, urinary retention, and cardiac arrhythmia. Changes in mental status are not seen in GBS unless associated with hypercapnia or hypoxia; however, acute intermittent porphyria may present as an acute polyneuropathy and encephalopathy.
5. **(C) Immunoglobulin, intravenously.** Both immunoglobulin administered intravenously (IVIg) and plasmapheresis have demonstrated benefits in treating patients with GBS. Both treatments improve time to clinical recovery, defined as either discontinuing mechanical ventilation or regaining independent ambulation. IVIg should be given at a dosage of 0.4 g/kg daily for 5 days, and 4 or 5 plasma exchanges of 3.5 to 4 L can be performed on alternate days over a period of 1 to 2 weeks. Complications are more common with plasmapheresis (eg, hypotension, line sepsis) than with IVIg (eg, headaches, myalgias, fevers, chills). Administration of corticosteroids, antibiotics, pyridostigmine, and interferon beta has no proven therapeutic benefit in patients with GBS.

SUGGESTED READING

Pascuzzi RM, Fleck JD. Acute peripheral neuropathy in adults. Guillain-Barré syndrome and related disorders. *Neurol Clin* 1997;15:529–47.