The clinical scenario presented is that of Guillain-Barré syndrome. The patient experienced the typical prodromal symptoms of distal numbness, paresthesia and pain in her limbs, followed by relatively symmetrical progressive (“ascending”) weakness of the limbs. Physical findings revealed the common hyporeflexia/areflexia pattern. The CSF examination demonstrated the classic “albuminocytologic dissociation” in which the CSF protein level is elevated, often remarkably so, in the absence of a pleocytosis. In clinical practice, this combination of physical findings of acute peripheral neuropathy and typical CSF findings allows the presumptive diagnosis of Guillain-Barré syndrome. If nerve conduction studies are performed, they will demonstrate and confirm the symmetrical sensory and motor peripheral neuropathy.

EPIDEMIOLOGY

Ongoing research indicates an autoimmune cause for Guillain-Barré syndrome, very likely related to cross-reactive antibodies to components of a variety of microbial antigens.1 This fact correlates with the epidemiology of the syndrome. In two thirds of cases, a history of previous upper respiratory tract or diarrheal illness can be elicited. Meta-analysis has identified significant percentages of specific infections preceding a Guillain-Barré syndrome.

Which of the following statements about this case is correct?

A. Immunization against herpes simplex viruses after recovery is indicated to prevent recurrences.

B. The prognosis at 6 months after recovery from the acute illness is uniformly good.

C. The most common epidemiologic association for the patient’s illness is infection with Campylobacter jejuni.

D. The optimal therapy during the acute illness is a course of high-dose corticosteroids initiated within 5 days of onset.

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What’s The “Take Home”?  
Young Woman With Weakness in Her Arms and Legs

THE TAKE-HOME MESSAGE:

In the post-polio era, Guillain-Barré syndrome is the most common acute peripheral neuropathy. It has epidemiological links to Campylobacter jejuni, cytomegalovirus and, to a lesser extent, other infectious pathogens. Symmetrical, bilateral weakness of the limbs, albuminocytologic dissociation of CSF, and typical nerve conduction study results support the diagnosis. Therapy with plasmapheresis or IVIG, but not corticosteroids, hastens recovery and diminishes morbidity. Despite this, an appreciable proportion of patients remain unable to walk at 6 months.

TREATMENT

Therapeutics involve strict attention to general measures appropriate to neurologic disease and more specific immunotherapies. Decompensation of ventilation to the point of requiring mechanical ventilation is a major risk; thus, careful monitoring of vital capacity, hypercarbia, and hypoxemia is mandatory, as is evaluation of the risk of swallowing dysfunction and aspiration. Another significant morbidity in these patients is deep vein thrombosis and pulmonary embolism such that these patients should receive an effective prophylaxis regimen when they are unable to walk.

Specific treatment aimed at the presumed autoimmune pathogenesis involves a variety of immunotherapies. Good experience and data indicate that plasma exchange and intravenous immunoglobulin (IVIG) are equally and significantly effective at hastening recovery when administered within 2 weeks of disease onset. Counterintuitively, the corticosteroids prednisolone and methylprednisolone do not hasten recovery or affect long-term outcome when used alone nor do they enhance efficacy if added to plasmapheresis or IVIG therapy. Thus, choice D is not correct.

PROGNOSIS

The usual course of Guillain-Barré syndrome is stabilization and recovery within 28 days of onset. The clinical course is often quite hectic: 66% of patients cannot walk independently at the point of maximum weakness and 25% receive mechanical ventilation. And, the intermediate prognosis is also more guarded than usually appreciated, with 20% of severely affected patients still unable to walk 6 months after disease onset. Thus, choice B is far too optimistic and not correct.

REFERENCES:


Figure – This photomicrograph depicts findings observed in a 48-hour culture of Campylobacter jejuni bacteria revealing characteristic “thin”, “comma-”, “S-” or “gull winged-shaped” forms displayed by this bacterium. (Courtesy of the CDC / Robert Weaver, PhD)