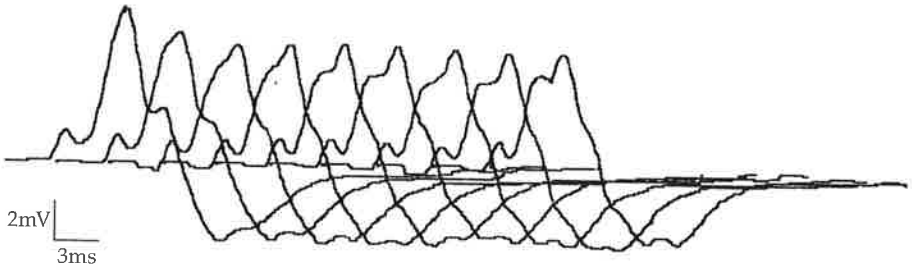
## CASE 9.12 DYSARTHRIA, PTOSIS, AND DECREASED EXERCISE TOLERANCE

**IMAGE 9.12 Decrement on Repetitive Stimulation**
Repetitive stimulation testing was performed by stimulating the ulnar nerve of the right arm and recording the CMAP over the right abductor digiti minimi muscle (see Figure 9.9; KCC 9.2). Stimulation was repeated at 3 per second for a total of 9 stimuli. Successive stimuli are shown displaced sequentially to the right (3 ms displacement per stimulus) to allow comparison of amplitudes of successive CMAPs: The first CMAP in the series is displayed farthest to the left, and the ninth CMAP is displayed farthest to the right. There was a decrement in CMAP amplitude of 23%.




## CASE 9.14 MYSTERIOUS WEAKNESS AFTER DINNER*

### MINICASE

One evening after dinner, a 58 year old woman began to have **swallowing difficulties** and **double vision.** Over the next few hours she developed **ptosis, facial weakness,** and **difficulty breathing** so her family brought her to the emergency room. Examination was notable for normal mental status, **severely limited horizontal and vertical eye movements with normally reactive pupils, facial diplegia, dysarthria, weakness of the bilateral arms and legs worse proximally than distally, 1+ patellar reflexes but otherwise undetectable deep tendon reflexes,** and a normal sensory exam. **Vital capacity was 600 cc** (normal is greater than ~3.5 L for an adult woman) so she was intubated.

### LOCALIZATION AND DIFFERENTIAL DIAGNOSIS

On the basis of the symptoms and signs shown in **bold** above, where is the lesion? What are some possibilities for the diagnosis?

*A description of this patient was published previously in Shapiro BE, Soto O, Shafqat S and Blumenfeld H. 1997. *Muscle and Nerve,* 20: 100–102.

## Discussion

The key symptoms and signs in this case are:

- **Diplopia, limited horizontal and vertical eye movements, ptosis, facial diplegia, dysarthria and dysphagia**
- **Proximal arm and leg weakness**
- **Breathing difficulty, with reduced vital capacity**
- **Diminished reflexes**

Generalized weakness without sensory loss can be caused by a brainstem lesion (see KCC 14.1) or by a widespread disorder of the lower motor neurons, peripheral nerves, neuromuscular junctions, or muscles (see KCC 6.3, Generalized Weakness or Paralysis). Proximal greater than distal weakness suggests a muscle or neuromuscular junction disorder, but the diminished reflexes suggest a possible acute polyneuropathy such as Guillain–Barré syndrome (see KCC 8.1). Another possibility is an acute upper motor neuron lesion, which can sometimes be associated with decrease rather than in-

creased reflexes (see Table 6.4). In summary, the differential diagnosis is large and includes an acute brainstem lesion such as infarct or hemorrhage, and rapidly progressive peripheral disorders such as Guillain–Barré syndrome, myasthenia gravis, heavy metal or organophosphate toxicity, botulism, porphyria, poliomyelitis, and tick paralysis (see KCC 8.1).

## Clinical Course

The patient was intubated and admitted to the intensive care unit. MRI with MRA did not reveal any brainstem abnormalities, routine blood tests and cerebrospinal fluid were normal. The family and patient repeatedly denied any possible toxin exposure. **Nerve conduction studies** demonstrated normal conduction velocities but markedly diminished compound motor action potential (CMAP) amplitudes (see KCC 9.2). In addition, fast repetitive stimulation or strong voluntary muscle contraction caused CMAP amplitudes to increase 2- to 3-fold. Increment of this kind is usually seen in presynaptic disorders of the neuromuscular junction, such as Lambert-Eaton myasthenic syndrome or botulism (see KCC 8.1; 9.2).

On further questioning, the family admitted to operating a home canning operation. They revealed that the patient had prepared a spaghetti dinner using their canned tomato sauce on the day of admission. Stool specimens and residual sauce brought in by the family were positive for botulinum toxin type B. She was treated with botulinum antitoxin, and had a prolonged course in the intensive care unit, but eventually recovered fully. When she was extubated and able to talk again, she explained that she had opened a dented can, and the sauce did not smell right, so she tried to feed it to the dog. The dog refused to eat it (wise choice), but the patient tasted some of the raw sauce which seemed alright, so she then cooked it and fed the whole family a spaghetti dinner. Fortunately, she cooked the sauce long enough to inactivate the toxin.

## Additional Cases

Other chapters describe related cases for the following topics: **radiculopathy** (Cases 8.1–8.11); **distal symmetric polyneuropathy** (Cases 6.5 and 10.3); and **cranial neuropathy** (Cases 12.2–12.7, 13.1–13.3, and 13.5). Other relevant cases can also be found using the **Case Index** located at the end of this book, and new cases are also available through the **Online Review and Study Guide**.



## Brief Anatomical Study Guide

1. The **brachial plexus** arises from **C5 through T1** (see Figure 9.2), while the **lumbosacral plexus** arises from **L1 through S4** (see Figure 9.4).
2. The most clinically important nerves in the upper extremity are the **radial, median, ulnar, axillary, and musculocutaneous nerves**; Table 9.1 summarizes the sensory and motor functions of these nerves.
3. The most important nerves in the lower extremity are the **femoral, obturator, sciatic, tibial, and peroneal nerves**; Table 9.3 summarizes the sensory and motor functions of these nerves.

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