Acute flaccid myelitis

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1 Acute flaccid myelitis is a clinical syndrome characterized by rapid onset of muscle weakness

From January to November 2018, there were 90 confirmed cases of acute flaccid myelitis in the United States and 48 suspected cases in Canada.[1,2] Injury to the anterior horn cells of the spinal cord causes abrupt-onset limb weakness, pain, paresthesias and areflexia.[3] About 25% of patients develop cranial nerve or bulbar signs or both.[4] It may resemble Guillain–Barré Syndrome, but can be distinguished by its typical appearance on magnetic resonance imaging (see point 3 below) and its motor distribution (asymmetric weakness in acute flaccid myelitis v. symmetric and ascending in Guillain–Barré Syndrome).[3] Typically affecting children aged 4–15 years, it is usually preceded by a nonspecific viral prodrome[3–5] (Box 1[6]).

2 Enteroviruses are the predominant cause in children

Clusters of acute flaccid myelitis occurred in North America in 2014, 2016 and 2018 during the late summer–fall months, coincident with peak enterovirus season.[2,4] With the near eradication of polioviruses, EV-D68 and EV-A71 have emerged as the predominant viruses associated with acute flaccid myelitis.[4] EV-D68 was likely responsible for the 2014 and 2016 clusters in North America,[7] but no cause has been established for the 2018 cluster.

3 Infectious workup and neuroimaging should be performed urgently

Cerebrospinal fluid (CSF), respiratory and stool samples, and selectively blood, should be tested for enteroviruses. If infection is present, the CSF will show a lymphocytic pleocytosis and normal to mildly elevated protein.[4] Magnetic resonance imaging with contrast of the brain and spine typically shows T_2 hyperintensities in the grey matter of the spinal cord and, often, enhancement of the nerve roots. In Guillain–Barré Syndrome, nerve root enhancement is seen, but spinal cord grey matter is not involved. Electromyography and nerve conduction studies show acute motor axonal neuropathy.[3] Suspected cases of acute flaccid myelitis should be reported to the Public Health Agency of Canada.

4 Acute flaccid myelitis is a medical emergency

Patients should be referred urgently to a tertiary care centre, as 24%–52% require admission to the intensive care unit for ventilatory or nutritional support or both.[5,8] Treatment should be considered on a case-by-case basis and may include immunomodulatory therapy, such as high-dose intravenous corticosteroids, intravenous immunoglobulin and plasmapheresis.[4] A vaccine has been developed for EV-A71 but is not licensed in Canada.[4]

Box 1: CDC case definition of AFM*[6]

- **Confirmed AFM**: acute focal limb weakness and MRI findings of mainly grey matter lesions involving one or more spinal cord segments.
- **Probable AFM**: acute focal limb weakness and cerebrospinal fluid pleocytosis (> 5 cells/mm^3).

Note: AFM = acute flaccid myelitis, CDC = Centers for Disease Control and Prevention, MRI = magnetic resonance imaging.
*No age limitation.

5 Acute flaccid myelitis often results in residual impairment

Canadian and US studies have described persistent deficits in most children after 4–6 months' follow-up, with full recovery in only 8%–18%, and 8%–14% either requiring assistive devices for ambulation or complete dependence on caregivers.[5,8]

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