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A Case of Poliomyelitis-like Syndrome

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Abstract A 30-month old boy presented with acute flaccid paralysis of the left leg. Electrodiagnostic studies revealed reduced amplitude of compound muscle action potential and prolonged F wave latencies. Magnetic resonance imaging of spine showed signal abnormality in the region of anterior horn cell in the spinal cord. Extensive virological tests in stool and blood samples were negative.

Key words Acute flaccid paralysis; Guillain Barre syndrome; MRI; Poliomyelitis-like syndrome

Introduction

Poliomyelitis is a viral infection involving the anterior horn cells of the spinal cord. With the extensive use of effective vaccine, it has become increasingly rare in developed countries. Most reported cases in developed countries are related to vaccination with live-accentuated polioviruses. We report a case with clinical and radiological presentation of poliomyelitis but negative virology studies.

Case Report

A 30-month old boy was assessed by the orthopaedic surgeon for a limping gait after slipping and falling while in the bath. He had had a mild cough and runny nose for 2 days before the presumed injury. During the initial assessment there was full range of movement of the joints and no definite swelling. Although he could still bear weight, he refused to walk on his left leg. The initial working diagnosis was a contusion of the left thigh. However over the following few days, he developed a swinging fever and refused to stand or move his left leg. He remained conscious but was tired. Further assessment on day 5 showed that he kept his neck in an extended posture and there was minimal spontaneous movement of the left leg, although no areas of local tenderness could be identified. The knee and ankle jerks of the left leg were hypoactive. Examination of the cranial nerves, sensation, upper limbs and right leg was normal. Sphincter function was unaffected. He had received full polio vaccination with the last booster dose being given approximately one year before and there was no history of contact with recently vaccinated infants or children. He had no history of recent travel and has been living in Hong Kong since birth.

Urgent computer tomography of the brain was normal. The cerebrospinal fluid (CSF) on day 5 showed raised white cell count of 42/mm^3 (60% lymphocytes and 40% polymorphs), raised protein level of 0.64 g/l (0.15-0.45 g/l), and normal glucose concentration. Electroencephalogram (EEG) revealed an asymmetrical background with abundant slow delta waves over the right occipital region. Magnetic resonance imaging (MRI) of brain and spine (without contrast) on the same day were normal.

Electrodiagnostic studies done on day 7 of the illness

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showed relatively lower amplitudes of compound muscle action potentials (CMAP) over the left leg (peroneal motor nerve: left versus right = 1.7 mV versus 7.1 mV; tibial motor nerve: left versus right = 5.7 mV versus 19 mV). Nerve conduction velocities and sensory nerve studies were within normal limit. F wave latencies of both tibial nerves were prolonged: 28 ms and 26 ms on left and right respectively (upper limit 20 ms). F waves were absent in both peroneal nerves. Intravenous immunoglobulin 2 gm per kg was given in view of the possibility of Guillain Barre syndrome (GBS). Considering the clinical picture of fever, lethargy, neck stiffness, abnormal EEG and raised CSF cell count, he was also treated with cefotaxime and acyclovir for possible meningoencephalomyelitis.

A second lumbar puncture performed on day 20 showed a normal white cell count 6/mm^3 and protein content of 0.28 g/L. No virus was detected in both samples. Polymerase chain reaction for Herpes viruses and enteroviruses were negative. CSF viral titres for herpes simplex viruses, varicella zoster viruses, measles and mumps viruses were also negative.

Serum antibody titres (acute and convalescent) for poliovirus (type 1, 2 and 3), enterovirus (Coxsackie virus A9, B1-6, Echoviruses 4, 6, 9, 14, 24 and 30), adenovirus, herpes virus, influenza, mumps, measles, mycoplasma pneumoniae and varicella zoster virus were all normal. Repeated stool cultures for polioviruses taken according to the WHO recommendation for acute flaccid paralysis were also negative. This patient was reported to the Department of Health as acute flaccid paralysis.

Repeated MRI brain and spine at week 3 showed hyperintense T2 signals in the dorsal aspect of the upper medulla/lower pons. Enlargement and T2 hyperintense signal was also present in the ventral surface of the cord at the upper margin of C4, and from T10/11 level to L1 level (Figure 1). On axial images, the T2 hyperintense areas correspond to the anatomic location of the ventral horns. No abnormal contrast enhancement was found after the administration of gadopentetate dimeglumine (Figure 2).

The patient continued to make slow progress with improvement in the left leg weakness. He could walk independently with a hyperextended knee five months after the onset of the illness. There was still minimal movement of the left hip. Power for knee extension and flexion was 3/5 (Medical Research Council Grading), ankle flexion was 3/5 and ankle extension was full. Reflexes on the left leg were still absent and there was pronounced muscle atrophy of the left leg.

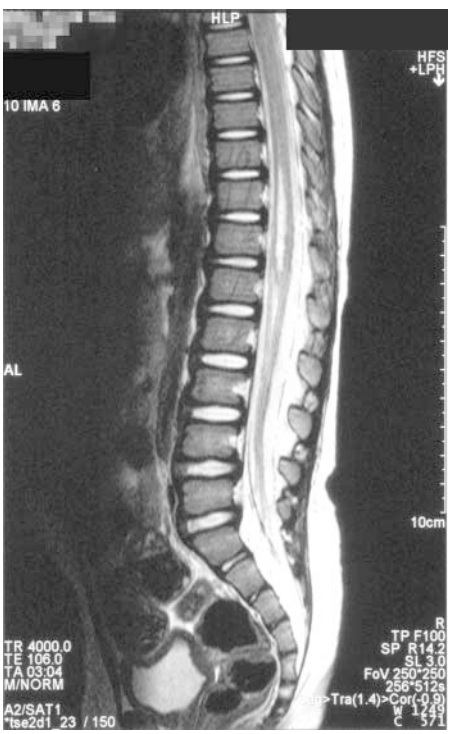

Figure 1 Enlargement and T2 hyperintense signal in the ventral surface of the cord from T10/11 level to L1 level.

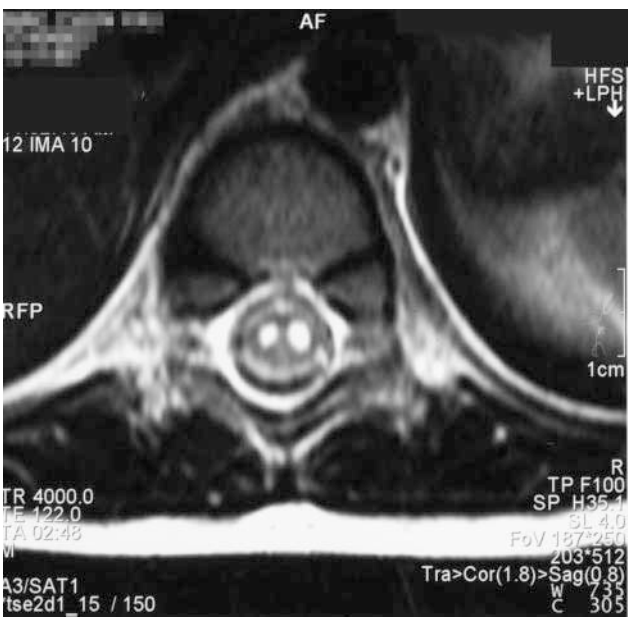

Figure 2 Axial image at T10 level demonstrating T2 hyperintense areas in the ventral horn regions.

Discussion

With the eradication of wild-type poliovirus in developed countries, most cases of acute flaccid paralysis are caused by oral polio vaccination or other RNA viruses, such as echovirus, Coxsackievirus and other enterovirus.[1-4] Vaccine-associated paralysis usually occurs within 30 days of immunisation with the relative risk of infection being estimated to be between 0.02-0.04 cases per 1 million doses of oral poliovirus vaccine. Most of these cases have been linked to poliovirus type 3.[5] The lethargy and neck stiffness in our patient was suggestive of aseptic meningitis. The acute flaccid paralysis produced by non-polioviruses can be indistinguishable from poliomyelitis clinically. In such cases, only a vigorous search for aetiological agents can help.[6] The clinical presentation of paralytic poliomyelitis can also be confused with that of GBS, which is the commonest cause of acute onset of weakness in developed countries. Both conditions can present with a prodrome of fever, upper respiratory or gastrointestinal symptoms, followed by asymmetric flaccid paralysis with hyporeflexia. Weakness typically affects proximal muscles more than distal. Pleocytosis in CSF is also a common finding. Patients with typical GBS demonstrate demyelinating neuropathy in electrodiagnostic studies. Reduced CMAP amplitudes are expected in patients with poliomyelitis, but these can also be found in patients with the axonal variant of Guillain Barre syndrome.[7,8] MRI spine is a useful tool in making this differentiation, as observed in our patient. T2-weighted signal hyperintensity and apparent enlargement of the ventral horns of the spinal cord, are fairly specific findings for poliomyelitis.[9] The MR features correlate with the pathological findings of severe inflammation, neuronophagia, active gliosis and destruction of the anterior horn cells in this group of patients.[10-12] The above findings contrast with those seen in patients with Guillain Barre syndrome who commonly have nerve enhancement seen on MRI.[13] The lack of significant clinical improvement in our patient also suggests irreversible damage of the anterior horn cells, as expected in poliomyelitis or poliomyelitis-like syndrome, whereas the prognosis for recovery in Guillain Barre syndrome is generally better.

Although poliomyelitis/poliomyelitis-like syndrome is uncommon in clinical practice, its signs and symptoms may not be easy to differentiate from Guillain Barre syndrome, especially if the virology tests are not informative. Characteristic finding of signal abnormalities in the anterior horn cell region of the spinal cord can then provide useful information for diagnosis and prognosis.

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